34 Primary Tumors of the Spine

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Primary tumors of the spine are very rare. The incidence is 0.4% of all tumors (77). Togetherness with myeloma, 1/4 of all primary bone tumors located to vertebrae. This number is increased by myeloma because nearly half of malignant bone tumors are myeloma. Hemangiomas are the second most common primary tumors of the spine (51).

The most commonly seen in the thoracic spine and at least in the cervical region. Primary tumors of the spine are derived from bone, cartilage and fibrous structures and all of them have benign and malignant forms. However, bone marrow-derived ones are always malignant and are usually the reflection of a systemic disease to the vertebrae (77).

Localization of tumors in the axial spine could give an idea about the underlying pathology. On the one hand, giant cell tumor, hemangioma, osteosarcoma and chondrosarcoma are usually located to vertebral corpus, on the other hand OO, osteoblastom and aneurysmal bone cyst are tend to localized to posterior elements (77).

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Benign Tumors and Tumor-like Lesions

Hemangioma

Vertebral hemangioma was firstly described by Perman in 1926, and in 1930 Bailey and Bucy (61). It is the most common primary benign osseous tumor (31,27,77). In post-mortem examinations, its ratio was 10-12% (18,27,31,51,61,67,77). Rarely it is symptomatic and it consist 1-3% of all symptomatic spinal tumors (51,77). Hemangioma is seen two times more common in women, and causes clinic symptoms in fourth and fifth decades (31,77). Pregnancy is a risk factor for revealing or exacerbating the symptoms. Aggressive hemangiomas are occur most often on T3-T9 vertebral segments (31).

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More than half of the bone hemangiomas are seen in the skull and vertebrae (5,27,51,67,77). The most frequently encountered in thoracic region (18,27,31,77). Hemangiomas are solitary lesion in 2/3 of patients, and had multiple spinal involvement in 1/3-1/4 of patients (27,31,51,77). Despite, the localization is mostly to vertebral corpus, it could spread pedicles, lamina, or other vertebral elements (31,51,77). Last plaques are usually preserved. Hemangioma is a type of arterio-venous shunt, which fed from small arteries that originating from the intercostal or lumbar arteries (77). It has three types (capillary, cavernous, and mixed). Cavernous type is more common (31,67,77). Hemangioma is usually silent and does not require treatment (18,27,77). Symptoms occur in 0,9 to 2% of the patients (27,31,61). Clinical symptoms may occur in the presence of neural compression, epidural hemorrhage or vertebra corpus compression fractures (27,31,51,77). Pain is the most common presenting symptom and also may cause radicular complaints and progressive paraparesis (51,67,77). Hemangiomas with aggressive progression and pathological fracture are rarely reported (27,31,77). Characteristics of aggressive hemangiomas are bone expansion, extra osseous extention and deterioration of local blood flow and these are rarely cause to compression fracture. In aggressive hemangiomas the incidence of neurological deterioration is 45%. The rest is characterized only by pain (61).

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Diagnosis is easy on plain radiographs. Rough vertical lines on vertebral corpus are characteristic diagnostic image (27,31,51,61,67,77). These radiological findings are seen because, horizontal trabeculae are destroyed by the tumor and compensatory hypertrophy is occur in vertical trabeculae (51,61,77). This view is more pronounced on lateral radiographs (51). Also due to the lytic areas "honeycomb" view is a characteristic finding on plan radiographs (27,31,61,67,77). Also "mottled" view which is called "polka-dot" is a characteristic finding on computed tomography (CT) (23,27,31,61,67,77). This "mottled" view is seen less in clinically silent hemangiomas (23). Silent hemangiomas have some characteristics on magnetic resonance imaging (MRI). They appear hyperintense on both T1 and T2 weighted images (5,23,27,31,51) (Figure 1,2).

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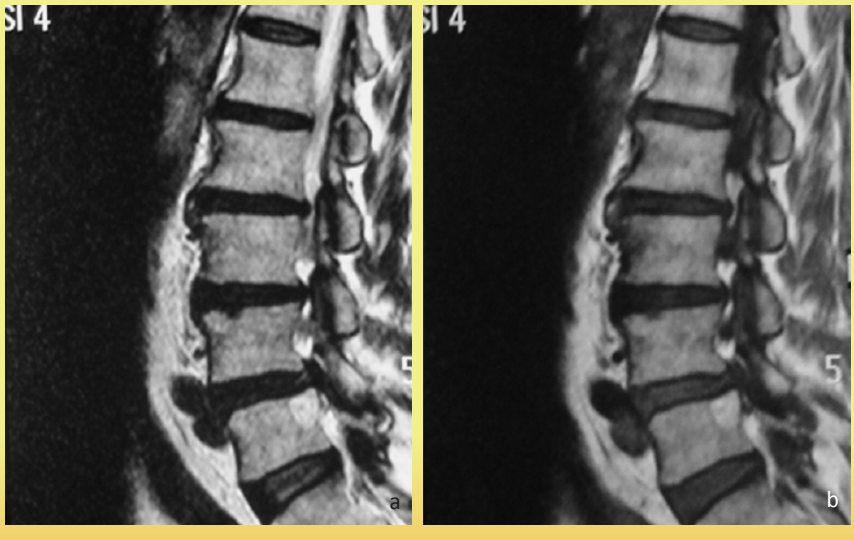

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Figure 1

L5 vertebral hemangioma is determined incidentally in lomber stenosis. Lesion is seen hyperintense both on T2 (a) and T1 (b) weighted MRIs (Yusuf Kurtuluş Duransoy's archieve).

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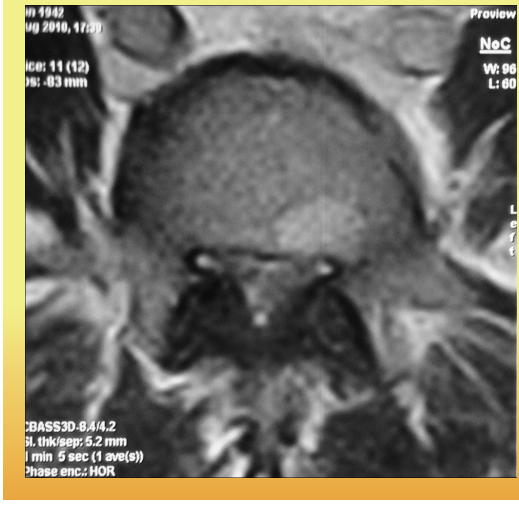

Figure 2

Hemangioma located to vertebra corpus is seen hyperintense on T1 weighted axial MRI (Yusuf Kurtuluş Duransoy's archieve).

Hemangiomas with clinical signs are appear less hyperintense due to less fatty content. This view leads to difficulty in metastatic tumors (23,61). If, only vertebral body is involved, the diagnosis is not difficult due to the characteristic radiological findings. However, if there is involvement outside the vertebral body diagnose is difficult. In the presence of pedicle involvement, it could differentiating with metastatic tumor (51,77).

Spreading of hemangioma from vertebrae corpus to soft tissue mimic the tuberculosis but lack of involvement in disc space is a remarkable feature in diagnosis (77). If the vertebra corpus is expansion or cortical thinning and rough trabeculae are occur similar images

like in Paget's disease may appear (51,61,77). However, involvement of other bones, increase in erythrocyte sedimentation rate and high value of alkaline phosphatase are favor findings for Paget's disease (51,77).

Treatment is usually aimed at complaints (67). It is mostly diagnosed incidentally when plain radiography, CT or MRI examinations are taken for other reasons (5,31,51,77). Hemangiomas which determined in this way do not require treatment (31,51,77). Only patients with pain are followed by neurological and radiologic examination once a year. This follow-up is more important, especially in female patients with hemangioma of the thoracic vertebrae. In the presence of radiological findings such as bone trabeculation and cortical expansion into the spinal canal or pathological vascularization in angiography it is more likely to have neurological symptoms (77). Treatment options include surgical decompression, endovascular embolization, intralesional injection of absolute alcohol, vertebral kyphoplasty with polymethylmethacrylate (PMMA) and the radiotherapy (RT) (31,67). In the presence of progressive neurological deficit or spinal instability surgery is indicated (27,31,77). Surgical treatment is very difficult due to tendency to excessive bleeding. Patients with clinical symptoms and/or findings, there is often neural compression and kyphotic angulation. Therefore, if surgical intervention is planned anterior approach should be preferred (77). Although laminectomy may seems to remove compression (27) the results may not be satisfactory (31). Removing the posterior elements may result in progressive kyphosis, exacerbation of pain or neurologic deterioration (31). Spondilectomy or corpectomy should be preferred for hemangiomas which are located on the vertebrae corpus (27,31). Involvement of both corpus and posterior elements, two-stage surgical approach should be (anterior and posterior) planned (31). If there is no vertebral compression fracture, it does not lead to neurological deficits but can cause pain. RT, vertebroplasty, embolization or sclerotherapy can be applied in pain (61,77). RT is not doing significant improvement in complaints on the contrary can cause radiation myelopathy (51,77). RT has effects on pathological vascular structures, but not on bone tissue (77). RT has lower success rates in kyphotic deformity and neural compression (31,77). RT is not recommended as only treatment method unless surgery or other treatment methods can not be applied. RT can be applied to prevent recurrence if complete surgical removal can not performed (31,61,67). Embolization temporarily eliminates pain but can not correct pathological compression (18,31,67,77). Preoperative embolization may facilitate the surgery by reducing the amount of bleeding (18,27,31,51,77). Vertebroplasty or kyphoplasty are usually made with PMMA. It prevents vertebral collapse and provides mechanical stability as well as reduces pain. This is a palliative method only used to be in painful conditions however should not be used in root or cord compression (31,67,77). Percutaneous injections of absolute ethyl alcohol has been reported as an effective in the treatment (51,67).

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Aneurysmal Bone Cyst

Aneurysmal bone cyst (ABC) was firstly described in 1942 by Jaffe and Lichtenstein (4,28,40,47). These are benign, cystic, vascular, and non-neoplastic tumor-like lesions of bone (51). Although it is a benign lesion it is locally aggressive (4,38,77). Generally, women are more affected and quite common in the first two decades of life (4,9,28,38,40,47,51,78). ABC is the third most common benign bone tumor after OO and osteoblastoma (78). ABC creates 1-2% of all primary bone tumors (4,9,40,47,51,52,78) and 15% of all primary spinal tumors (4,9). Involvement of the spine is around 10-30% but contradictory rates were reported due to the scarcity of case series in literature (4,9,28,38,47,51,52,78). However, can be seen in all segments of the spine out of the coccyx (28). Usually occurs in posterior elements of the vertebrae (4,28,47,51,78). ABC can be classified as primary or secondary (9,47). Seventy percent of the cases are primary, and 30% are secondary to other tumors (4,51). Tumors are thought to originated and co-located with giant cell tumor, telangiectatic osteosarcoma, osteoblastoma and chondroblastoma (9,47). Also solid variation of ABC has been reported in the literature (9).

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The most common symptom is pain at night which located in the lesion site (47,78). The pain often begins suddenly, and gradually increases (51). Local swelling is a common finding (9,28). Neurological deterioration is rarely reported. Spinal cord compression, paraplegia or cauda equina syndrome may manifest due to pathological fractures (4,9,38). Although many theories have been suggested including vascular, traumatic and

genetic factors (4), the etiology is unknown (9,51,52,78). The prognosis is good even recurrence after the treatment. Rarely spontaneously regression of ABC has been reported (4,9,28,38,40,78). Malghem reported 3 cases of spontaneous improvement (40). Malignant transformation is rare and usually reported after RT (9,52).

Majority of cases are diagnosed by plain radiographs, CT and MRI. On plain radiographs sharply demarcated, osteolytic and expansile cavities in the posterior elements are characteristic. CT and MRI could determine multilobule lytic lesions or pathological vertebral body fracture. In MRI each lobular cavity could show different signal characteristics. In planning of surgical treatment, CT and MRI are important diagnostic tools (9,78). On plain radiography, CT and MRI, fluid-fluid levels in cystic areas can be seen. This finding is quite significant for the ABC but not pathognomonic because this appearance can be seen also in telangiectatic osteosarcoma, giant cell tumor, chondrosarcoma, secondary ABC and simple bone cysts (4,9,51,78). Also radiological determination of ABC as cystic (presence of fluid-fluid levels) or a solid character is important for the results of surgical treatment. In differential diagnosis giant cell tumor, chondroblastoma, chondromyxoid fibroma, fibrosarcoma, telangiectatic osteosarcoma, fibrous dysplasia, and plasmacytoma should be considered (78). Although CT and MRI are important diagnostic tools, biopsy is necessary to confirm the diagnosis because of the possibility of similar bone lesions.9,78

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Treatment of ABC is still controversial (4,40,78). Treatment options include intralesional curettage (with or without bone grafting), wide surgical removal (with or without adjuvant therapy), arterial embolization, intralesional injections of drugs (steroids and calcitonin), cryotherapy, sclerotherapy, radiofrequency ablation and RT or combinations all of these methods (4,9,47,78). The most important factors in the management of treatment are early diagnosis and appropriate surgical resection. En-bloc surgical resection (with or without instrumentation) is the best approach for reducing the recurrence. However, en-bloc surgical resection of large tumors may cause spinal instability (4,78). For this reason, implant requirement should be calculated in preoperative period. Injection of PMMA can be applied to strengthen the defect after intralesional curettage. Injections of methyl prednisolone or calcitonin into the cavity are safe procedures without adverse effects (78).

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Local progression was reported with biopsy followed by RT or RT alone. RT should not be considered in the presence of pathologic fracture, deformity, instability and neurological deficit (4). There are quite a lot, and serious adverse effects of RT4 (38,78), such as growth disturbances, radiation myelopathy, osteonecrosis, gonadal damage and the development of osteosarcoma (4,28,38,40,47,51,78). Papagelopoulos et al. reported a patient with osteosarcoma after 7 years of RT (52). However, RT is widely used as an adjuvant treatment after incomplete resection. As a result, RT provides supportive care in conditions such as incomplete surgical resection, aggressive recurrence or patients with a high risk for reoperation. However, RT is used as a primary treatment in very limited number of cases whom surgical treatment can not be applied (4,38,47,78).

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Selective arterial embolization is considered to be an alternative method to the standard surgical procedures because it is a less invasive, simplier, cheaper, more effective and repeated method (9,38,78). Recurrence rates comparable with intralesional surgery and can be applied as primary treatment in some region that can not be reached by surgery (8). RT should not be applied in the presence of pathologic fracture or neurological deficit (9). Also application of preoperative embolization reduced bleeding during surgery (4,38,52,78). In the thoracic or upper lumbar spine lesions, embolization should be applied very carefully otherwise inadvertently embolization of Adamkiewicz artery may lead to spinal cord ischemia (4,9,47).

The clinical course of ABC is unclear, but local recurrence is known to occur with different types of treatments (4,9). Recurrence rates have been reported about 10-44% and recurrence in en-bloc surgical resection is rarely (4,47,78). Ninety percent of recurrence occurs within the first 2 years (4,9,78). Hay et al. reported 7 of 28 patients (25%) with recurrence after incomplete surgical resection and no recurrence after en-bloc surgical resection (28) Boriani et al. reported recurrence in 1 of 5 patients with incomplete surgical resection and no recurrence in 13 patients with en-bloc resection without RT (9). Finally in 2013, Zileli et al. have reported 18 patients whom treated with surgical intervention. Thirteen of 18 treated by en-bloc resection and only 1 of 13 had recurrence. On the other hand 5 patients had treated by incomplete surgical resection and 4 of 5 had recurrence (78).

Osteoid Osteoma

Osteoid osteoma (OO) is a rare bone tumor and firstly described by Jaffe In 1935 (33,36,58). OO is a benign osteoblastic tumor and growth potential is limited (no more than 1 cm in diameter) (26,51,58). Histological features of OO and osteoblastoma are very similar. However, osteoblastom is greater, more aggressive and may have malignant transformation (26,33,58). OO creates 3% of all bone tumors and 11% of primary benign bone tumors. Ten percent of OO located in the vertebrae (26,33,51,58,77). Localization is mostly lumbar (56-60%), cervical (27%), thoracic (12%) and sacral (2%) vertebrae respectively (26,51,58,77). Usually located in posterior elements of the vertebrae (26,33,51,58,77). OO is seen in the young patients especially in the first decade (26,51). Most of the patients are under 30 years of age (33,51,77) and seems two times more in men than in women (26,33,51,77).

The most common feature of the OO is pain in the lesion site. Stiffness due to spinal muscular spasms is accompanied to pain (26,33,36). Pain shows slow progression for several months (36,51). Sometimes history of pain may be decades before the radiological findings (26,33,51). Characteristic of pain is change from intermittent and mild to constant and severe. Pain does not decrease with rest and exacerbated at night (26,33,51,77). The response to acetyl salicylic acid and other nonsteroidal anti-inflammatory (NSAI) drugs is very good (26,33,51). The most common physical finding is sensitivity of the lesion (51). Painful muscle spasm and scoliosis are the valuable findings. In a young patient with muscle spasm and suddenly appearance of scoliosis, tumor should be keep in mind (26,33,36,51,57). Often the first symptom is scoliosis. Tumor is typically located on the concave side, at the apex of the scoliotic curvature and at the posterior elements (26,36,77). The time to onset of symptoms to diagnosis is around 7-36 months (26,36,57,77). In patients with early recognized after surgical resection, scoliosis may resolve spontaneously. However, in patients with symptoms for more than 15 months, structural changes may also occur with muscle atrophy and scoliosis may become permanent (36,51,57,77). In this cases, surgical treatment may need for scoliosis (77).

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Plain radiographs and CT demonstrated sharply demarcated radiolucent nidus about 1 cm in diameter and sclerosis around the nidus (26,51,77) (Figure 3,4)

Figure 3

Axial, sagittal and coronal CT demonstrated typical appearance of osteoid osteoma located to C3 vertebra corpus (Nuri Erel's archieve permission).

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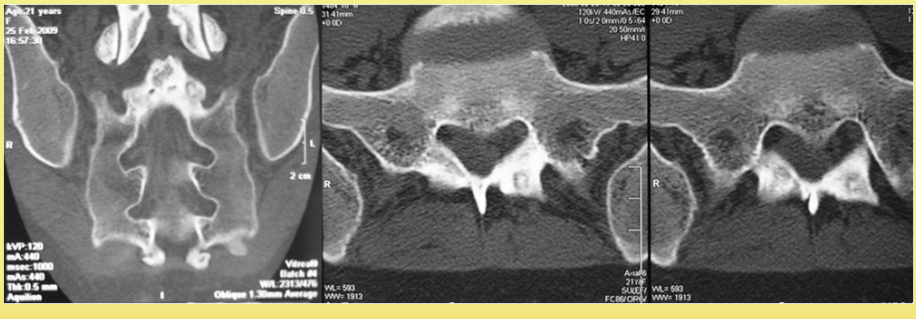

Figure 4

CT demonstrated typical appearance of osteoid osteoma located to posterior elements of sacrum (Nuri Erel's archieve permission).

This appearance is pathognomonic but not diagnostic. These radiological findings are similar to, chronic osteomyelitis such as "Brodie abscess" (51). On plain radiographs it is difficult to see lesion because OO located in posterior elements (77). These tumors are often nonspecific in MRI. However, the effects of the tumor on the spinal canal, spinal cord and spread to epidural space could be seen in MRI (58). Technetium bone scintigraphy is a useful tool that can demonstrate the location of the lesion (36,51,58,77). In scintigraphy, typically, hot sclerosis around the cold central nidus is seen (26,77) (Figure 5).

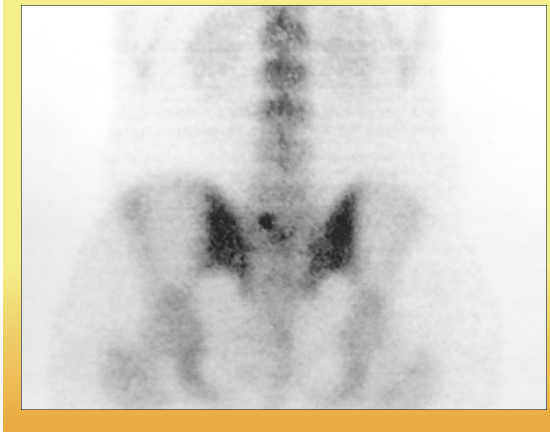

Figure 5

Sacral osteiod osteoma is seen as a hot lesion on scintigraphy (Nuri Erel's archieve permission)

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Until diagnosis, patients receive other diagnoses (51). In the differential diagnosis, osteoblastoma, osteosarcoma, osteomyelitis, Ewing sarcoma, eosinophilic granuloma, metastasis, fractures, aseptic necrosis, osteochondritis and ABC should be considered. OO is suspected from the characteristic clinical and radiological findings however histological examination of biopsy or surgical material confirms the diagnosis (51).

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Spinal OOs remain limited to 4-8 years in natural processes. However, if the symptoms persistence and spinal deformity progresses, surgical intervention is necessary (77). The most effective treatment is complete surgical resection of the lesion (26,33,51,58). If complete surgical resection is not be maintained removing a portion of the sclerotic bone which surrounds the nidus may alleviated symptoms (51). However, complaints never completely disappears if nidus is not removed completely (51). If the pain continues after surgery, it means incomplete resection (77). Recurrence rate is reported as 4,5% (33) but recurrence is not expected after complete resection (26,58,77). Spontaneous healing has been reported in rare cases (36,51). In recent decades, percutaneous radiofrequency ablation in the treatment of OO has been published (58).

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Osteoblastoma

Unaware of each other Jaffe and Lichtenstein firstly described benign osteoblastoma in 1956 (11,15,33,36,58). Osteoblastoma is a rare and histologically benign but aggressive bone tumor (51,59,77). Create less than 1% of all bone tumors (11,39,51,59,62) and 3% of benign bone tumors (11,51,62). OO is tend to placed to spine (51,15). Fourty percent of the reported cases have located to vertebrae (11,33,39,51,58,59,62,77). The rates for location vertebrae are lumbar, thoracic, cervical and sacral 35%, 35%, 25%, 5% respectively (51). Osteoblastoma usually located to posterior elements of the vertebrae such as pedicle, lamina, transverse and spinous process (11,19,33,36,39,51,58,59,77). Primary involvement of vertebrae corpus is very rare, it is more often seen in the spread of tumor from the posterior elements (15,59). While osteoblastoma is limited to only posterior elements in 50%, togetherness of posterior elements and vertebrae corpus is 42%. Involvement of vertebral corpus alone is 3% (39). About half of the spinal osteoblastoma, are located to lamina and/or pedicle of a vertebra (39,51). In 5% of cases, the tumor can invade the adjacent vertebrae (51). Right/left location rate is 2/1. Almost half of the reported cases had epidural extension (51). Involvement of paravertebral soft tissues are less (39). Large part of patients are adolescent. Approximately 80% of patients are under the age of 30 and 60% of patients are less than age of 20 years (15,33,39,51,58,59,77). More frequently seen in males (11,15,33,51,59). The tumor is larger than the OO (51,58,77). Diameter of the tumor is usually larger than 1,5-2 cm and shows aggressive behavior (26,39,51). There is no information about the pathogenesis (51).

The most common symptom is blunt local pain (33,39,51,58,59). Onset of pain is insidious and months or even years pass until the diagnosis (15,33,51,59). Characteristic of pain is different from OO (39,51). Pain is milder and not exacerbated at night (39,51,58,59,77), not reduced with acetyl salicylic acid and may increase with movement (15,51,58,59). More than half of the patients have radicular pain and it is another difference from OO (33,51,59). Scoliosis is detected in more than half of the patients (51,59,77). The leading causes for pain-related scoliosis are osteoblastoma or OO (19,36,57,51). There are non-specific physical findings in osteoblastoma. Local tenderness and swelling could detect on palpation of the tumor on the vertebrae (51). In the presence of neural compression, abnormal neural reflexes, muscle atrophy, muscle spasm, muscle weakness and neurological deficitis can be detected (33,51). Neurological disturbance are more in ostoblastoma than OO (26,33,51,59). Because the size of the tumor and neural compression are more in osteoblastoma (39,51,59,77). Spinal cord damage reported in 1/4 of cases (51).

Osteoblastoma develops faster, inflate and destructed the region (33,51). On plain radiographs characteristic lesion is seen as sharply demarcated and surrounded by a thin shell of reactive bone (19,51,59,62). On the contrary to OO, there is no bone formation around lesion (51,77). Middle of the lesion is often radiolucent but sometimes it may be radiopaque (19,39,51,59). CT is superior to MRI and plain radiography in the determination of these characteristic features of the tumor and local staging (62). In addition, CT and MRI demonstrate the location and extent of lesions in the exact format which can not seen in on plain radiographs (59) so CT and MRI provide great convenience for surgical intervention (36,51,58,62). Osteoblastoma is seen low or medium signal intensity on T1 weighted images and high signal intensity on T2 weighted images. Although, MRI provides good information about reactive changes in the adjacent bone and soft tissue, it has limited value in determining the characteristics of osteoblastoma (58,62). Bone scintigraphy help to determine the location of the lesion (19,51,58,59). In literature there is no false-negative result for Scintigraphy (19,51) so it is an important examination in terms of determining residue after surgery (19).

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Definitive diagnosis is made by histological evaluation of the biopsy specimen (51). In the differential diagnosis osteosarcoma, OO, ABC, giant cell tumor, chondroblastoma should be considered (15,39,51,77). Clinical, radiological and histological, findings of osteosarcoma (especially osteoblastic type) can interfere with osteoblastoma (15,19,51). On plan radiographs, surrounding of lesion by reactive bone layer, histologically absence of anaplasia cells criteria and cartilage component help differentiation of osteoblastoma from osteosarcoma (51). However in recent years, an osteoblastic lesion which is seen rarely have been proposed for borderline form between osteoblastom and osteosarcoma. For this intermediate form the "malignant osteoblastom", "aggressive osteoblastoma", "low-grade malignant osteoblastoma" terms have been proposed (15,19,39). In some recurrent osteoblastoma, sarcomatous changes were determined in borderline lesions and suggested that they are borderline from the beginning and they initially misdiagnosed (19). In the differential diagnosis it is difficult to distinguish from OO histologically (19,33,51,77). Typical features of the pain (77), diameter of tumor (smaller than 1,5-2 cm) (33,39,58,59,77) not extend beyond the bone (19,33,59,77) as well as more regular and organized osteoid trabeculae ensures correct diagnosis for OO (19,51).

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Without neurological impairment or loss of function en-bloc removal of the lesion and the radical curettage of the surrounding normal bone should be the choice method for treatment (33,51,59,77). However, because of involvement of the posterior elements the opportunity to do this kind of surgery is low in osteoblastoma (51,59). A portion curettage of the lesion, allow to decrease complaints and can stop the tumor growth for a long time (51,77). The role of RT in osteoblastoma treatment is controversial. RT should be considered only when there is residual or if surgery can not be applied. RT is not a harmless method and has some risks such as malignant transformation, increasing the risk of spinal cord compression and necrosis of the spinal cord (51,59).

Although there is insufficient information about the prognosis of osteoblastoma (51) most of the cases have benign course (39,51). Generally, this behavior continues even when there is recurrence (51). Recurrence rate has been reported between 5-20%.39 (51,59), Initially, rare malignant transformation of benign osteoblastoma cases have been reported (39,51).

Giant Cell Tumor of Bone

Giant cell tumor of the bone (GCT) is a rare tumor with unknown origin (43,77). Although histologically benign, clinically shows aggressive behavior (51,69,74,75,77). Recurrence rate is high and can be done distant metastasis (69,75,77). The incidence is 5% in all bone tumors (14,43). Frequently localized to epiphyseal and metaphyseal part of the long bones (14,43,51,60,69,77). The sacrum is the fourth most common region and sacral involvement is reported between 1,7% to 8,2% (14,43,69). The incidence for localization to other part of spine is 2-4% (43,77). Involvement of the vertebrae (sacrum included) is 2,7 to 6,5% (60,64,74). In all localizations men and women effects equally and commonly seen between 20-45 years (43,51,74). In vertebrae involvement slightly female superiority has been reported (14,51,69,74). GCTs can benign metastases to lung (14,43,77). Yang et al. reported the development of lung metastases in 3 of 11 patients in their series (75). The most common symptoms are pain and neurological disturbance (43,74,75). Complaints are found for months or even years before diagnosis (75). Sensitivity can detected in tumor region (51). Progresses of tumor and neural compression caused to radicular pain and neurological deficits (51).

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In radiological examination expansile lytic lesion without a sclerotic margin can available on lateral of the sacrum (43). In vertebrae (excluded sacrum) equal involvement is seen. Lytic lesion with more involvement is seen in vertebra corpus (43,51,77). It does not show sclerotic margin and periosteal reaction (77). MRI is important because it shows the spread of tumor to epidural space (51,77). In radiologically, GCT may be confused with cyst-like lesions such as ABC and osteoblastoma (77). Also primary hyperparathyroidism, may cause similar radiographic findings in sacrum (51,77). This lesion is called "brown tumor" (77). CT-guided biopsy is a valuable tool for the diagnosis (43,77).

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The aim of treatment is resection of the tumor, reduce the local recurrence, elimination of the pain, protection of neurological functions and providing spinal alignment and stability. Arterial embolization, curettage, en-bloc resection, RT and cryotherapy are different treatment methods (43,60,69,75). Still ideal method for treatment continues to be a debate (43,60). Surgical resection has been reported the main method for treatment (75). Best results are usually obtained by en-bloc surgical removal. However, it is more difficult to achieve in vertebra lesions (60,64,75). Martin et al. recommended en-bloc surgical resection for appropriate cases. Also they recommended intralesional surgery after preoperative embolization in large tumor. Serial arterial embolization therapy is offered as a treatment option due to the low morbidity rates (43). Marcove et al. reported that they had obtained good results with cryotherapy. However Leggon et al. reported higher

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rates of recurrence after curettage and cryotherapy (69). In GCTs the use of RT after intralesional surgery is still controversial (43,60,64,69,75). RT has some risks such as myelitis, graft complications and sarcomatous transformation (14,64,75). Yang et al. reported malignant transformation in 1 of three patients after postoperative RT (75). Crew et al. reported a malignant transformation that resulted in death after RT for sacral GCT (14). The possibility of radiation-induced sarcoma progression was reported as 10% (60,69). If the RT dose is over 45 Grey this ratio will go up till to 29% (14,77). Therefore, RT should be used only as an adjuvant treatment if complete surgical resection can not be applied (21,43,75,77) or should be reserved for recurrences (21,75).

Local recurrence is the most common complication and its rate is reported as 11-50% after en-bloc surgical resection (21,43,60,69,77). In all cases, the average recurrence rate changes from 22,4 to 41,7% (60,74). Thangaraj et al. reported that RT provides high local control rate and radiation-induced malignancy rate is low (69). The rarity of the disease due to the localization to vertebrae, a few information has been published, and most of known has been obtained from small case series (60,74). Martin et al. implemented en-bloc surgical resection in 11 patients and reported only 2 recurrences (43). Although recurrence is noted after 10-year period of time as well (64) recurrence usually occurs within the first 6 years (21,60,64). Even local recurrence often occurs in the first three years after surgery (21,64).

It is difficult to predict the prognosis (77). The main determinant of prognosis is width of the surgical tumor resection (69,75). Clinical, radiological and histological criteria which shows the right treatment option to reduce the rate of recurrence is still undetermined

(75). Xu et al. reported 102 patients and identified that being under the age of 40 and en-bloc surgical resections are good prognostic factors for low recurrence rates (74). There is no correlation between histological staging and prognosis (51,77). Enneking staging which is made on the basis of clinical and radiological findings, still remains its validity for treatment selection and determining the prognosis (51,77). Protection from the recurrence is the most important issue in the management of treatment (74).

The malignant equivalent of this tumor is known as "giant cell sarcoma". This is a high-grade sarcoma which includes scattered giant cells, and usually develops in the same locations after RT (77).

Eosinophilic Granuloma

Eosinophilic granuloma (EG) is a benign tumor like lesion (54,77). Histologically, contains a large number of histiocytes and eosinophils (77). Solitary EG was described for the first time in 1940 by Lichtenstein and Jaffe (48). In childhood and adolescence, it is the most common benign lesion of the vertebrae. Mostly seen between 5 to 15 years (48,51) Eighty percent of the patients are under the age of 10 (12,48) and seen more common in women. Constitute less than 1% of all bone tumors. Placement of the vertebrae is around 7-17%12 and mostly localized to thoracic vertebrae (12,48,51,77) which followed by lumbar and cervical region respectively. Localiztion to sacrum is very rare 48,51 Lesions may be solitary or can be a part of systemic diseases such as histiocytosis-X, Letterer-Siwe, Hand-Schuller-Christian (12,48,51,54,77).

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The most common presenting symptom is pain. 12 Cause to local pain on acute onset (77). In the presence of neural compression, neurological complaint can occur (51,77). Compression on vertebral corpus is regular so rarely cause to deformity (51).

The most common radiographic appearance is lytic lesions on vertebral corpus. If compression fractures occur in children, "vertebra plana" appearance is seen (48,77). This appearance is pathognomonic for EG and seen in late stage of the disease (48). Vertebra plana is typically seen in EG and also can be seen in other pathologies. In addition, image "on top of money" is a typical radiographic appearance. Vertebral corpus collapsed and disc spaces are preserved (77). CT demonstrated the characteristics of lesion. MRI provides information about soft-tissue component and helps to evaluate neural compression. MRI is also a useful tool in the evaluation of response to treatment (12,51). In differential diagnosis osteoblastom, ABC, Ewing sarcoma, infectious discitis and Paget disease should be considered (48,51). Although scintigraphy is not a sensitive diagnostic method for these tumors, it is important for both differential diagnosis and to demonstrate single or multiple lesions (12,51). Although diagnosis is mostly made clinically and radiologically, in suspected cases biopsy and histological examination obtained for definitive diagnosis (12,48,51,54).

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The goal of treatment is to relieve pain, to preserve spinal stability and neurological function and eradicate the lesion (12). Some authors advocate the use of an orthosis to ensure the protection of the stability of the spine in patients who have solitary lesion without neurological involvement (12,48,77). Raab et al. reported that, they treated 14 children who have no neurologic deficits by orthosis immobilization and they provide the normal height of the vertebral corpus and vertebral remodeling (54). However, this treatment modality in patients with neurological deficit is controversial. In the presence of moderate neurologic deficit, on the one hand some authors defending immobilization and RT (12,48,77), on the other hand some believe RT and chemotherapy (ChT) after neural decompression and fusion (12). The systemic form can be treated with ChT. Solitary EGs are self-limiting and can resolve spontaneously (12,48,51,54,77). In the presence of more than one bone involvement without additional tissue, prognosis is very good (51). In children because of negative effects on the bone growth, secondary malignancy risk and radiation myelitis risk RT treatment is not included in the routine. RT is only reserved for progression (12,48). In certain histopathological diagnosis, CT-guided intralesional corticosteroid injections are used. After a few injections stagnating the lesion development, decreasing of the pain, and complete resolution is seen. Even children vertebral corpus which flattened, rotates back to its original size and shape (48,51,54).

Spot girilecek

Malignant Tumors

Multiple Myeloma and Solitary Plasmacytoma

Multipl myeloma (MM) is primary malignant bone tumor of plasma cells. In adults, it is the most common primary malignant tumor of bone (51). MM creates 50-55% percent of all malignant bone tumors (51,77). Solitary plasmacytoma (SP) creates only 3-5 % of malignant bone tumors (7,35,45,51,77). SP is most common localized to spine (68%) in all bones (45). MM is also the most common primary malignant tumor of the spine (77). MM is more over the 50 years of age and is seen equally in both gender (6,7,51,77). Chronic inflammatory, viral diseases and myeloproliferative disorders are considered to be the initiator factors. In MM vertebrae involvement is always seen however can not be symptomatic. Patients with symptomatic involvement of thoracic, lumbar and cervical vertebrae are 59%, 31% and 10% respectively (51).

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The first and most common symptom is pain (35,51). Thirty five percent of patients present with low back pain. Initially, the form of pain is mild and transient. The pain increases with movement and decreases with resting (51). Sometimes the patient may present with radicular symptoms (35,51). Twenty percent of patients usually presented with vertebral fracture with history of trauma (51,77). In most patients history of the complaints are shorter than 6 months (51). In solitary lesions, the mean onset of clinical history can range from 6-12 months (35,45,51). If vertebral collapse reaches to further dimension or tumor makes neural compression, neurological disorders can occur (6,35,51). Paraparesis or paraplegia may occurs more in SP and kyphosis can develop (45,51). Due to an abnormal increase in immunoglobulin, progressive renal failure, susceptibility to infection and amyloidosis may occur (51,77). If bone marrow involvement increases with disease progression, widespread bone pain, fever, pallor and purpura symptoms may become constant. Laboratory investigations revealed several important findings which may be helpful in the diagnosis (51). In complete blood count usually normochromic normocytic anemia is detected because of the diffuse bone marrow involvement. Erythrocyte sedimentation rate is high (6,77). Biochemical globulin fraction increased and dense bone involvement may cause to hypercalcemia. Alkaline phosphatase is usually normal or increased. Urea and creatinine levels are improved in kidney failure (6,51). Protein electrophoresis is also distorted in MM (6,77). Serum protein changes are also quite characteristic (51). A single antibody which made from the heavy and light chains are height in MM. Excessive light chain construction may lead to Bence-Jones proteins in the urine (6,51). Bence-Jones proteinuria is seen in 50% of the patients and symptomatic from time to time (77).

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Basic radiographic evidence is, the presence of large number staple holes or moth-eaten osteolytic foci (51,77). The absence of reactive sclerosis around the lytic lesions is a characteristic finding in MM. Involvement of vertebrae corpus without posterior elements helps for the differential diagnosis from other osteolytic lesions. Metastatic tumors are usually involve the vertebral corpus and the pedicles (51). Intensive osteolysis in bones may thought to be osteoporosis (51,77).

SP, can be seen in many organs, tissues and including the bones (35,51). Pathology can be seen in vertebral corpus, pelvis and long bones. Spine SPs have different radiographic features. It can be seen as an osteolytic lesion or as an expanded lesion included sclerotic trabeculae. The involved vertebral corpus may break or may disappear completely. Sometimes tumor invasion reach to adjacent vertebrae and this can mimic infection (51). Lesions including rough trabecule with striasyon may be confused with vertebral hemangioma (51). Bone scintigraphy determined cold or normal uptake because this technique does not show lytic lesions (6,51,77). Therefore scintigraphy in the diagnosis of MM is not a good screening method (51,77). CT may reveal involvement of bone lesions before reflection on plain radiographs (51). CT is also helpful to identify the degree of tumor penetration and soft tissue involvement also to investigate ektramedullary plasmacytoma (6,45,51) (Figure 6).

MRI is a useful method for determination early changes in bone structure (45,51) In MRI hypointense signals on T1 weighted images and hyperintense signals on T2 weighted images are seen based on normal bone marrow (Figure 7).

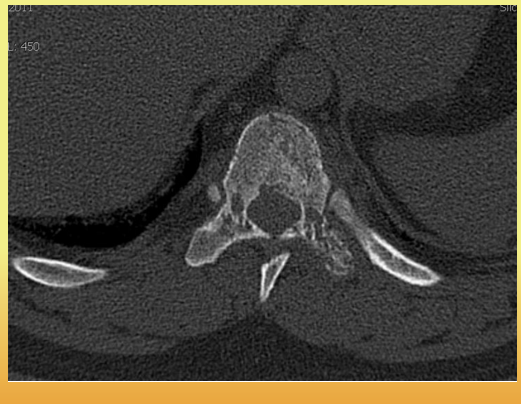

Figure 6

Typical appearance of solitary plasmocytoma on CT in thoracic region (Yusuf Kurtuluş Duransoy's archieve)

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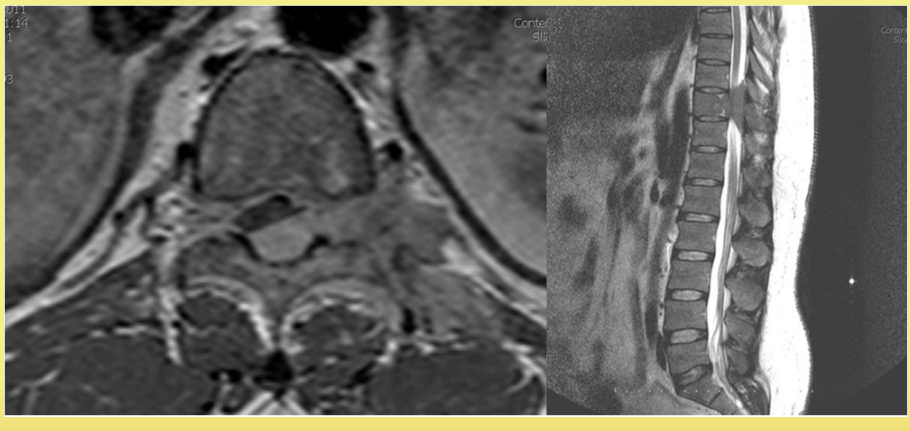

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Figure 7

Plasmacytoma cause to spinal cord compression in thoracic region. Tumor appears hyperintense according to bone both on T1 and T2 weighted MRIs (Yusuf Kurtuluş Duransoy's archieve)

Positron emission tomography (PET) does not distinguish myelomatosis lesions but useful in determining involvement regions (6).

For correct diagnostic approach, clinical, radiological and laboratory data should be evaluated carefully. Histological diagnosis should be made if a lesion is detected (35,20,12,51). Bone marrow aspiration is usually sufficient for diagnosis but biopsy is rarely required (51,77).

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The diagnosis of SP is hard entity.51 Sometimes it may be the early stage of MM.35 Lesion can be accepted as SP in following cases 1)-If MM is diagnosed with biopsy, 2)-In the presence of a single focus on of skeletal scan, 3)-If there is no plasma cells in bone marrow smear, 4)-If Bence-Jones proteinuria or hyperglobulinemia was not detected, 5)-If the patient has followed for several years (51).

Metastatic tumors and other hematologic malignant diseases should be consider in the differential diagnosis. If appearance of the focal osteolysis detected on plain radiographs, infections and hemangioma should be considered in differential diagnosis. Also similarly hypercalcemia and bone changes can be seen in hyperparathyroidism (51).

The average survival is 4-5 years. The most important factor that reduces the systemic and spine complications is treatment method. Bortezomib (Velcade), thalidomide (Thalomid), and lenalidomide (Revlimid) are new generation chemotherapeutic drugs, which have positive effect on survival (6). ChT provides reduction in pain and slowing down of bone loss in 70% of patients. At the same time, intensity of abnormal protein is decreases, hematological disorders; urea, creatinine and calcium levels are begin to normalization. Laminectomy and/or RT is indicated for decompression in patients with spinal cord

compression (51). RT can also be used in painful SP (45,51,77). These lesions are very radio-sensitive (6,45,51,77). In MM surgical treatment should be considered in the presence of acute-progressive neurological deterioration or instability due vertebral compression fracture (6,77). Surgical indication is more in SP because delay in diagnosis cause to compression of neural tissues due to vertebral collapse. Timing of RT is important if surgery is considered. To avoid deterioration of osteoblastic activity in bone graft, application of RT is more useful after 4-6 weeks of surgery (45). In SP, aggressive treatment can provide long-term remission (77). Advances in the treatment of skeletal complications of MM (such as bisphosphonates treatment) decreases bone involvement (6). In addition, application of vertebro-kyphoplasty with PMMA improves to the quality of life of patients with MM (6,45).

Chordoma

Luschka described firstly clivus chordoma in 1856 (77). Chordoma is a malignant tumor originated from notochord remnants in skull base and the spine and usually symptomatic in elderly patients. It is a rare primary bone tumor (13,16,41,44,73,77). Chordomas are locally invasive tumors, and grow slowly (10,13,22,41,51,73). Presentation time to physician is about 1 year (32) and creates 1-5% of all malignant tumors (22,16,44,51,73). The most commonly seen between 40-70 ages (13,32,51,73,77). While placement to sacrococcygeal region is seen more in men, there is no significant difference for other spine regions for gender (44,51). Chordomas are completely localized to axial skeleton (13,51,73) which can be seen everywhere from skull base to coccyx (13,51,77). However, 85-90% of the cases reported in literature located to clivus and sacrococcygeal region (32,51,77). Localization to sacrum, clivus and the other vertebrae are in 50%, 35%, 15% respectively (13,22,16,41,44,51,77). In spine (excluding the sacrum), mostly located to cervical region. Cervical, lumbar and thoracic localizations are 6-10% 4-5% 1-2% respectively (13,51,77). Moreover, chordoma is the most common tumor of the sacrum and creates 40-50% of all primary tumors of the sacrum (22,73,77).

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Signs and symptoms may vary according to the location, size, and spreading of the tumor (13,51,73). In sacrococcygeal localization pain can be blunt or severe, temporary or permanent (51). Diagnosis is usually delayed because pain is insidious and symptoms are nonspecific (51,73,77). Patient can presented with severe constipation, frequent urination or inability to urination, urinary incontinence, dysuria, or with muscle weakness (51,73). Clivus chordomas can cause neurological symptoms due to neural structures compression (51,73). Cranial nerve dysfunctions or endocrine disorders due to spread the pituitary gland can occur (32,73). Neurological disorders are more likely occur (radicular pain, loss of sensation, etc.) in other spinal region involvement than sacral involvement (41,73) Neurological examination is very important to investigation these neurological symptoms (51). In pelvic localization tumor can detected with rectal examination (51,77). In lumbar and sacral localization, flaccidity of muscles in lower extremity is seen however spasticity may occur in upper level spine involvement (51,73).

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Chordoma creates a lytic region and soft tissue mass in bone (16,41,51,73). Lytic lesions can be easily seen on plain radiographs. Lytic lesion starts in a single vertebral corpus and is usually surrounded by sclerosis (16,41,51,73,77). Firstly disc spaces are intact but by time tumor spread to adjacent vertebral corpus (41,73,77). Disc space is collapsed and the facing surfaces of the adjacent vertebral corpus are gentled (16,51). In a further periods vertebra corpus is collapsed (41,77). Calcification is determined in 40-80% on CT or plain radiographs (16,51,77). Calcification is amorphous and usually localized to peripheral region (41,51,77). Especially prevertebral soft tissue mass in sacrum is typical (73,77). Soft tissue mass is greater than bone destruction (51,77). Soft tissue mass appear like muscle and has homogenous contrast enhancement (16,51,73). On MRIs hypo-or isointense signals on T1 and hyperintense signals on T2 weighted are seen (16,41,73). With CT and MRI paravertebral tumor size, both bone and soft tissue mass, invasion and neural compression can easily demonstrated without further investigation (16,41,51,77). Diagnosis of chordoma is easier because of its radiological features and location (51). A lytic lesion in the sacrum, primarily should be considered the chordoma (77). However, definitive diagnosis is made by biopsy (16,41,51). In the differential diagnosis, GCT, osteochondroma, chondrosarcoma, osteoblastoma, osteosarcoma, MM, metastatic tumors, ABC and sacrum bone cysts should be considered (22,41,51).

Primary method of treatment is tumor's en-bloc resection (13,41,51). Initially, the width of the surgical resection is the most important factor affecting the cure (10,13,16,22,73). However, large size of the tumor during diagnosis, invasive nature of the tumor and advanced age of patients made en-bloc surgical resection almost impossible (22,16,51,77).0 Therefore mostly intralesional approaches are preferred (16). However, should be careful to avoid spreading the tumor into surrounding tissues (13,16). In sacral region sacrectomy may allow en-bloc surgical resection of tumor but in the upper levels results of surgical treatments are worse (77). In most cases anterior intervention is required because of vertebral corpus involvement. Laminectomy is aimed to decompression however not preferred. Because after laminectomy, probability of recurrence rate is high and spinal instability can occur (16,51,41,77). RT treatment as a primary or adjuvant is still controversial (41,73). Chordoma relatively resistant to radiation (13,16,22,77), If en-bloc surgery can not be done RT can applied to shrink tumor and relieve the pain (10,13,16,77). RT should be preferred in tumor recurrence and if surgery should not be applied (10,13,51,77). Boriani et al. reported local recurrence in all patients within 17 to 20 months after palliative RT, RT alone, or intralesional intracapsular surgery. Also they reported local recurrence rate as 20% of patients within 56 to 94 months who underwent en-bloc surgical resection (10). The average life expectancy is 5-6 years (13,73,77). This period can be more with radical surgery and seems to be shorter if en-bloc surgical resection can not be applied (22,77). Chordomas may metastasize to lungs, bone, skin, brain, lymph nodes, muscle-skeletal system and other internal organs (10,16,73). Metastasis is not typical during diagnosis. Diagnosis is often delayed so distant metastasis is increased (22,73). During the diagnosis metastasis is seen in 5% patients (22,16,73). In the next period possibility of metastasis is up to 65% (16,73). Local recurrence indicates poor prognosis than distant metastasis (10,13,16,22,73). In chordomas sarcomatous transformation develops in 2-8% (13). ChT is not used in treatment of chordomas (13,22,41,77). Better results are expected in the future with new RT techniques and new generation ChT drugs (such as imatinib mesylate) for the treatment of recurrences and complaints (10,13).

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Osteosarcoma

Osteosarcoma (OS) is a typical example for osteoblastic malignant tumors (25,70). It is the 2nd most common primary malignant bone tumor (20,70). Primary spinal osteosarcoma is very rare. Primary OS creates 3,6 to 14,5% of all primary tumors of the spine and 0,4 to 3% of all OS. Limb OS is seen frequently in children and adolescents but spine involvement is seen elderly (10-30 years old) (20,25,37,50,77). Increasing incidence in elderly is due to RT and development secondary to Paget's disease (25,77). Rare Osteoblastoma cases have been reported whom had malignant transformation into the OS (39). OS is two times more common in men and mostly lumbar, sacral, thoracic and cervical vertebrae are affected respectively. The most commonly located to vertebrae corpus. Both vertebral corpus and posterior element involvement are also common. However, the only posterior elements localization is rare. OS may metastasize to lungs, pleura, liver and other internal organs of the abdomen and to other bones. Spread to mediastinal lymph nodes can occur secondary to lung involvement (25).

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Frequently pain and neurological disorders occur (25,77). Typically lytic and destructive regions are formed on vertebral corpus and posterior elements (25,50,77). Osteoblastic shadow is a common radiological appearance (50). CT demonstrated destructive tumor and soft tissue involvement (25,77). MRI is an essential diagnostic method in the determination of neural compression and bone invasion especially in sacrum (50).

Differential diagnosis is required from solitary lytic, sclerotic and mixing lesions. OS should be differentiation from benign lytic lesions such as ABC, GCT and osteoblastom. Osteosarcoma is a malignant tumor and can be easily separated from benign osteoblastic lesions by clinical, radiological and histological findings (15). Also in differential diagnosis malignant lesions such as Ewing's sarcoma, metastasis, lymphoma, myeloma and chordoma should be considered (25).

CT or fluoroscopy-guided biopsy is not caused to the spread of the tumor so it is an important tool for the diagnosis. In spine involvement the diagnosis is often delayed (25,50). During the diagnosis metastasis was often determined (25).

The most important treatment is surgical resection (20). In the presence of neural compression or spinal instability surgical treatment is required to reduce tumor mass, relieve neural compression and provide the stabilization. However, complete surgical resection without damage to neural tissues is difficult (25,50,77). Ideal surgical treatment is en-bloc surgical resection without spreading to surrounding area (20,50,70). Feng et al. reported that, en-bloc sypondilektomy which applied to thoracolumbar OS provide, better local control and longer survival rates (20). Ozaki et al operated 22 patients who have spinal OS and reported that complete surgical resection is better than incomplete resection (50). It is well known that OS is sensitive to ChT and improves the prognosis (20). ChT should be applied prior to surgery (neoadjuvant) and postoperative (adjuvant) period to prevent systemic spread, to facilitate excision of the tumor by reducing the size and to reduce risk of local recurrence (25,20,50,70). In patients with surgery alone (without ChT) local recurrence is determined in high rate (50,70). Because of radiation-resistant RT is not used in the primary treatment (20,70). RT is used after incomplete surgical resection and in patients who are not appropriate for surgery (20,77). However, some authors reported that RT in routine treatment has positive effects (25,50). Ozaki et al. reported longer lifetime in OS patients with postoperative RT (50).

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OS is a high-grade malignant tumor (20,77). and has high mortality rate. In addition, because of its locally aggressive feature, it has high local recurrence and metastasis rates after appropriate surgical resection (20). Spine imvolvement has worse prognosis than limb involvement (25,20,50,77). The median survival is 9-12 months (25,50,77). Recently improvements with adjuvant and neoadjuvant therapy with ChT are reported (50).

Chondrosarcoma

Chondrosarcoma (CS) is malignant tumor that makes cartilage tissue. CS creates 10-20% of all bone tumors (51) and is the third most common bone sarcoma followed to OS and Ewing's sarcoma (8,34). Spinal involvement is ranged from 2-12% in different series (8,34,51,66). While the involvement of the lumbosacral rate is 50-68%, thoracic and cervical involvement rates are 23-32% and 9-18% respectively (8,34,51). Lesions are located to vertebral corpus, posterior elements and both in 5%, 40%, and 45% respectively (34). Often occurs between the ages of 30-70 (8,34,51,66,77). Very rarely seen under the age of 20 (8). Males are more frequent (8,51,55). Eighty five percent of CS is primary (34,51,77). Others may develop as a result of secondary to malignant degeneration of pre-existing cartilage tumor such as chondroma, enchondroma or osteochondroma (34,51,55,66,77). Secondary ones tend to be low-grade and has better prognosis than primary (34).

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It can reveal different biological behavior (77), and the findings are not specific to the tumor (51). Usually grows slowly and is not symptomatic until advanced periods (8,51,66,77). Sometimes, can behave like an aggressive metastatic tumor (77). Local or radicular pain is the most common presenting symptoms (34,66). Nearly half of the patients have palpable mass (8,34,51,66), and neurological disorder (34,51).

Radiological appearance of CS depends on the histological degree. On plain radiography it appears as well-circumscribed calcified lesions. High-grade tumors show intermittent periosteal reaction and moth-eaten lytic lesion plain radiographs. Initially, differented the lysis defect from enchondroma is difficult (34). Tumor appears such as cloudy, snowflakes or corn-like calcification (51). Sometimes it can be seen as lytic lesion which located to vertebral corpus and cause to compression fracture (34). Plain radiographs may be useful to reveal calcific tumors (51). However, CT and MRI are more useful to evaluate the extention of tumors to soft tissue (8,34,51). In tumor mass calcifications which are called "ring and arcs" appears are seen on CT.34 Also soft tissue mass is a common finding (8,34). On MRI tumor appears in low signal on T1 weighted images and heterogeneous low high and mixture signals on T2 weighted images (34). In addition, MRI determines an epidural or neural foraminal exceed and neural compression better than CT (34).

The differential diagnosis depends on the presence of calcifications. In the presence of calcification main reason is generally enchondroma otherwise metastasis, malignant fibrous histiocytoma, and fibrosarcoma should be considered. Deep endosteal scalloping (cortical thickness), cortical disruption, periosteal reaction, soft tissue mass and intense

radionuclide uptake are favor criteria for CS. In differential diagnosis angioblastic meningioma, osteosarcoma, Ewing sarcoma and hemangiopericytoma should be considered. Also pathological fractures and osteoporosis should be ruled out (34).

Before starting treatment histological diagnosis should be clarified with CT guided biopsy (34,66). Histological grading is an indicator to predict the biological behavior of the tumor. However, the prognosis determines by treatment (34). Nowadays, surgery is still the most appropriate treatment method (8,51,55). Surgical treatment is aim to provide the functionality of the spine, relieve pain, and provide control of local recurrence with long survival (34). The ideal surgical method is en-bloc resection of the mass with healthy boundaries without spread tumor cells to surrounding tissues. However, it is often difficult to achieve it in the presence of spine involvement (8,34,55,66,77). While local recurrence rate is 3-8% in en-bloc surgical resection (8,34,66), this rate is 100% in intralesional surgical resection (8,34,51,66) Similarly, survival times affected positively from the en-bloc surgical resection (8,34). Tumor-related mortality rates are of 12% and 42-61% in en-bloc resection and incomplete resection respectively (8,34) CS are resistant to RT and ChT protocols (8,34,51,55,66,77). Both RT and ChT used after complete surgical resection. However, their positive effects on survival and local control are not demonstrated in literature (34,55). ChT is used as an adjuvant therapy in incomplete resection (34,77) and as therapy in metastatic patients (34). RT is used as palliation therapy in patients who are not appropriate for surgery (51). RT has no role in the primary treatment. Although RT applications were used after incomplete resection, beneficial effects have not been reported (34).

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Prognosis depends tumor's histological grade and width size of the surgical resection (8,34,66). High-grade tumors grow quickly and metastasis early . Low-grade CS grows slowly, shows local recurrence and late metastasis. Low-grade CS has higher survival rate than high-grade and the time between treatment and recurrence time is longer (51). En-bloc surgical resection is an independent prognostic factor which has positive impact on local tumor control and survival (34,55). If en-bloc resection can not be performed local tumor recurrence is inevitable (8,34,51,66). In a clinical series with 69 patients including 12 spine CS, Bergh et al. reported 5, 10 and 15-year survival rates as%72, %67 and %63 respectively (34). In the same study, older age, higher tumor grade, inappropriate surgical margins, and local recurrence were reported as negative factors which have affecting the survival (34,55). York et al. reported 5 and 10-year survival rates as %64 and %40 respectively in their clinical series which include 21 patients treated surgically (34). In the same study, tumor-related death in the local recurrence rate is reported as %61 (34). Distant metastases are rarely reported in the literature and are associated with high tumor grade and local recurrences (34,55). In recurrence, increasing rate of histological grade is seen about 13% (34,51).

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Ewing Sarcoma

Ewing Sarcoma (ES) is described by James Ewing in 1921 (1,30). ES is the second most common primary malignant bone tumor in children and adolescents (1,24,42,71,72). Often seen in the second decade (1,24,30,42,72) and located to metaphyses of long bones (24,30). The most common primary involvements are to pelvis, femur, tibia and fibulae (1,24,30,42,71). Primary spinal involvement is 3,5 to 10% (3,42,63,71,72). In terms of response to therapy and survival rates primary spine ESs are can be divided into two separate groups such as sacral and nonsacral involvement (17,24,63). Sacral involvement is more than nonsacral (24). Primary nonsacral spinal involvement is 0,9% (24,63). Nonsacral spine tumor often located to posterior elements then spread to vertebral corpus. In sacrum most commonly located to sacral ala (30). ES often makes lung metastasis (63). Tumor often spreads to the neural canal and surrounding soft tissues (1,30).

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Local and/or radicular pain is the most common complaint (1,17,24). A palpable mass is detected. In nonsacral spinal involvement pain is usually intermittent and not worse at night. This intermittent feature may cause a delay in diagnosis (24). In patients with lumbar radicular pain it may be confused with lumbar disc herniation (24,63). Although neurological deficitis may be caused by compression to spinal cord, this process usually occurs in later stage of the disease (1,24,71).

X-rays do not help in early stages (17,30,63) but can visible after the occurrence of neurological deficitis (17,63). Lytic lesions are usual (1), however sclerotic lesions are rarely seen (1,17). In some cases, there is only a soft tissue mass, and can not seen on X-rays (63). CT is a valuable diagnostic method that can demonstrate the extention of the lesion on vertebral corpus (17,63). Plain radiographs and CT may be useful in determining lung metastasis (63). MRI is a valuable tool for early diagnosis. MRI is important in evaluation of response to adjuvant ChT, planning of surgical treatment for the determination of tumor margins (showing spread of epidural and bone marrow) and in the post-treatment follow-up (17,63). ES is often multifocal disease and whether or not the primary tumor or to determine the dissemination before treatment planning and in follow-up period, whole body bone scintigraphy should be done (17,24,63). In nonsacral spinal involvement in order to diagnose, seriously doubt is required. To reduce the delayed diagnosis a detailed story and careful physical and radiologic examination is required (17,24). Treatment results are much better in early diagnosis (24). ES, is a small round cell tumor of bone (1,17,24) In differential diagnosis small round cell tumors such as neuroblastoma, primitive neuroectodermal tumors of bone, malignant lymphoma, and rhabdomyosarcoma should be considered (17,24,63). For definitive diagnosis CT-guided needle biopsy is made (17,63,42).

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The most important determining factor in deciding ES treatment is the presence of neurological deficitis (17,24,63). Patients with rapidly progressive neurological deterioration early decompressive surgery should be the first preferred method (1,17,24,63). Surgical approach is preferred according to involvement. In the presence of anterior spinal cord compression due to tumor extantion anterior decompression is preferred. ES generally tend to spread from paravertebral soft tissues to neural foramen and invade all around the spinal canal. In these patients, laminectomy is an effective approach (17,24,63). Laminectomy without stabilization increased the possibility of post-laminectomy kyphosis (1,17). ChT and RT are should be done to control micrometastases and local recurrences respectively (17,24). After definitive diagnosis with needle biopsy, neoadjuvant ChT with combine chemotherapeutic drugs is recommended. Neoadjuvant ChT shrinks the tumor according to the sensitivity of the tumor and increases the probability of total resection and reduces the possibility of micrometastases (17,24,63) and histological grade (17). In literature there are published articles which shows improvement in neurological deficitis after ChT application (17,63). After neoadjuvant ChT, surgery or RT, or both should be applied. In spine ES, primary RT is not recommended (24,63). Because RT will create post-treatment edema and it can cause deterioration in neurological deficit or formation of new neurological deficitis (24,63).

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ES is an aggressive tumor and the prognosis is poor (63). While sacral involvement has the worst prognosis, other bone (excluding spine) involvements has the best prognosis (17,63,72). Factors affecting the prognosis are dissemination, tumor size, severity of neurological deficit, tumor location (being nonsacral or sacral), the patient's age and the response to neoadjuvant ChT (63,71).

Lymphoma

Primary bone non-Hodgkin's lymphoma is a rare disease (53,56,65,68) and has been described by Parker and Jackson in 1939 (53,56,68). Also called as primary bone lymphoma (53,68). The incidence of primary bone lymphoma in all primary malignant bone tumors is 2.8-7% (2,29,49,53,56). Primary placement of the spine is very rare (46,53,65,68). Primary spine lymphoma creates 0.1-6.5% of all lymphoma cases (65,53,68) and 1-9% of all primary bone lymphomas (2,29,53,65,68). The most common locations are lumbar and lower thoracic vertebra respectively (29,49,65).

Spine lymphoma is initially very insidious and can be asymptomatic until neurological complaints. Once patient is symptomatic, findings progress rapidly (65). Patients usually refers to a physician with pathological fracture or neurological symptoms (56).

Lymphoma is usually characterized with irregular osteolytic bone defects on X-rays. However, plain radiographs may be overlooked (49,65). CT and MRI of the spine demonstrate the tumor, but there is no specific findings for lymphoma (76). Tumor appears iso or hypointense on T1 weighted images and iso or hyperintense on T2 weighted images (76) (Figure 8)

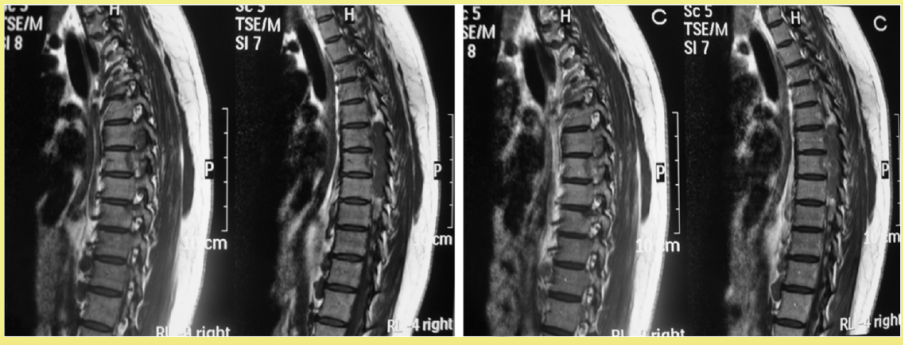

Figure 8

T1 weighted MRI demonstrated primary lymphoma in thoracic region. Lymphoma appears; A)-Hypointense on precontrast T1 weighted MRI. B)-Post-gadalinyum images demonstrated a slight contrast enhancement (Yusuf Kurtuluş Duransoy's archieve)

Spot girilecek

PET is useful tool in determining the tumor is whether primary or metastatic. None of the other imaging methods can not help in the differential diagnosis (68) therefore a needle biopsy is required for histologic diagnosis (68,76). The main purpose of treatment is extending the quality lifetime (53,68). ChT, RT, surgical treatment or combination treatments are well known for bone involving lymphomas (53,65,68,76). Lymphoma is quite sensitive to ChT and RT (46,53,68). Surgical treatment is aimed to eliminate neural compression, remove the tumor mass, and stabilization of the spine and to provide a definitive histologic diagnosis (29,46,53,65,68). In the presence of neurological deficitis the ideal treatment is very controversial (53,65,68). There is no sufficient case series in the literature so treatment modality is still unclear (53,68). Peng et al. published 13 primary lymphoma cases who are presented with neurological complaint and they reported that ChT and RT improved neurological deficitis rapidly (53). In an another study Tang et al. reported 40 primary spinal lymphoma cases who had neurological deficit. Authors advocated that in severe neurological deficit, surgical treatment should be preferred primarily. According to these authors, in moderate and mild neurological deficit without instability, ChT and RT can be applied as a first-line treatment. However, despite this treatment if neurological deficit does not improve within 2 weeks or mass does not shrink radiological or if neurological deficit is worsened, surgical treatment should be applied (68).

Spot girilecek

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