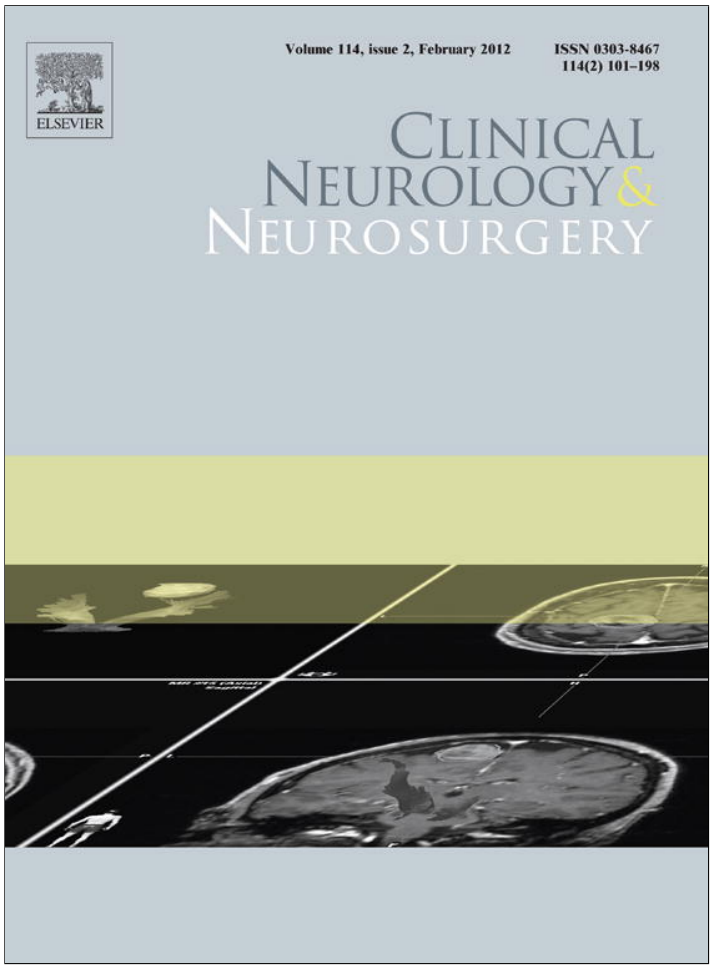

Clinical Neurology and Neurosurgery 114 (2012) 166–168

Contents lists available at SciVerse ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Case report

Slow progression and benign course of a primary malign melanoma of a lumbar nerve root

Ozan Ganiüsmen[a], Füsun Demirçivi Özer[a], Mesut Mete[b,*], Nail Özdemir[a], Ümit Bayol[c]

[a] Izmir Tepecik Training and Research Hospital Department of Neurosurgery, Izmir, Turkey
[b] Nevsehir State Hospital Department of Neurosurgery, Nevsehir, Turkey
[c] Izmir Tepecik Training and Research Hospital Department of Pathology, Izmir, Turkey

ARTICLE INFO

Article history:
Received 1 August 2011
Received in revised form 6 September 2011
Accepted 21 September 2011
Available online 20 October 2011

Keywords:
Lumbar spine
Nerve root
Primary malign melanoma

1. Introduction

Over 90% of malignant melanomas of the spine are metastatic. Rarely, however, a primary malignant melanoma may originate from the spine. In this setting, an intramedullary location is common. The remaining malignant melanomas in this region originate from the spinal cord or leptomeninges, which surround the nerve roots. The diagnosis is confirmed immunohistologically, as it is difficult to distinguish a malignant melanoma from melanotic schwannoma and meningeal melanocytoma [5]. Total excision of a spinal malignant melanoma can have a better course with adjuvant radiotherapy and chemotherapy [3]. This case report presents a patient who underwent surgery in 2007 and the pathological diagnosis was a primary spinal malignant melanoma originating from a nerve root. Despite metastasis, the patient was pain free and had no neurological deficits at the 4-year follow-up.

2. Case report

In 2007, a 49-year-old female with a history of heart problems was admitted to Izmir Tepecik Research and Training Hospital complaining of low back and leg pain that started 2 years earlier and had increased in severity over the last 6 months. There were no findings other than decreased left knee flexion and extension. Magnetic resonance imaging (MRI) of the lumbar spine showed a 4 to 5-cm mass that was derived from the left L3 nerve root and had destroyed the L3 pedicles (Figs. 1 and 2).

During surgery under an operating microscope, a mass characterized by a black color, smooth borders, and internal necrosis was excised totally after detaching it from the radix. A perioperative frozen section of the tumor revealed “pigmented malignant melanoma.” On pathological examination of paraffin sections, the tumor was composed of heavily pigmented, solid epithelioid sheets. The pigment reacted negatively with Prussian Blue, which stains hemosiderin. Immunohistochemically, the tumor cells were negative for EMA and PANCK, and strongly positive for S100 and HMB45. The histological, histochemical, and immunohistochemical data were all diagnostic for “pigmented malignant melanoma” (Fig. 3).

The patient’s other organs were checked carefully, including the skin and eyes. Full-body computed tomography, MRI, and positron emission tomography did not show another focus. The patient was diagnosed with primary spinal malignant melanoma. A postoperative MR image was obtained 1 month later and revealed no residual tumor in the lumbar region (Fig. 4). Radiotherapy was administered postoperatively and temozolomide was started. On detecting lung nodules after 1 and 5 years, interferon was started, followed by chemotherapy with fotemustine. When examined in 2010, focal sclerotic lesions were found in the S1 bone. Nevertheless, the patient remains pain free and without neurological deficit and is being followed by the medical oncology and neurosurgery departments.

* Corresponding author. Tel.: +90 384 215 33 00; fax: +90 384 215 03 67.
E-mail address: dr.mmete@hotmail.com (M. Mete).

0303-8467/$ – see front matter
doi:10.1016/j.clineuro.2011.09.012

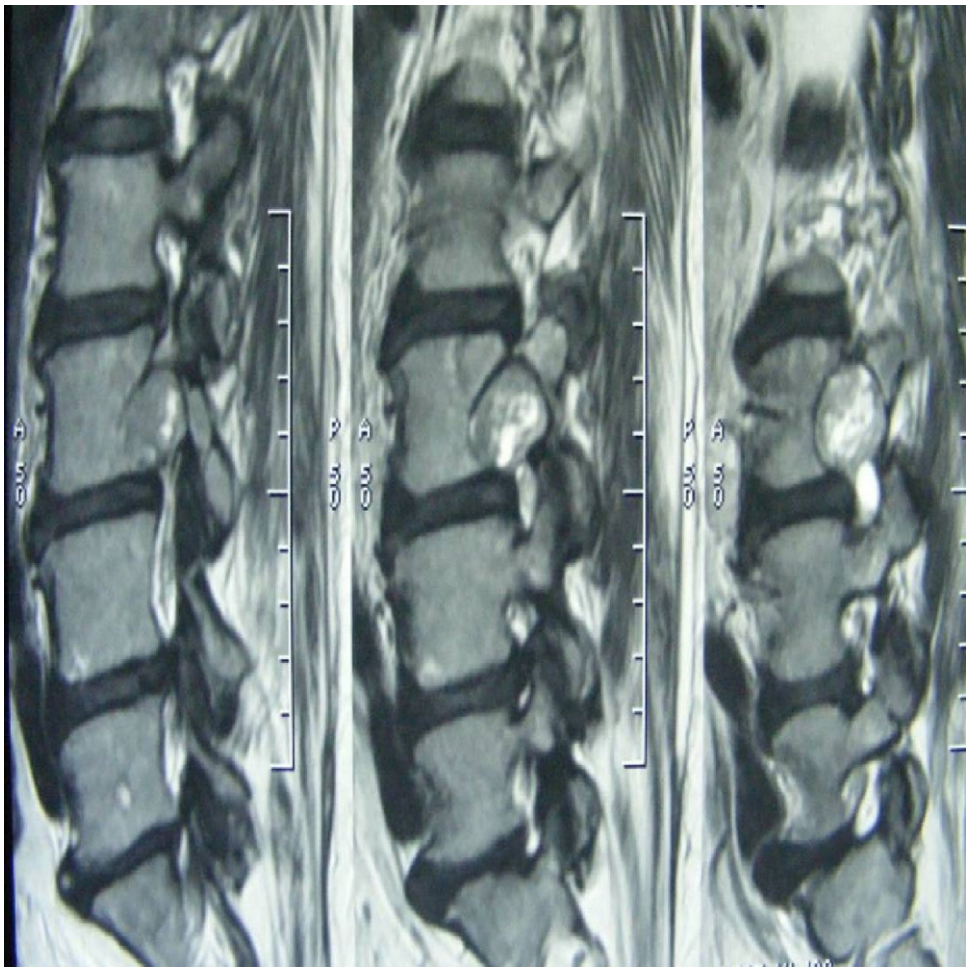

Fig. 1. T1-weighted MRI sagittal section of the lumbar spine shows the tumor exiting the foramen.

3. Discussion

Melanocytes are melanin-producing cells that arise from the neural crest during embryogenesis and migrate to the skin, mucous membranes, and central nervous system (CNS). In the CNS, melanocytes are most numerous in the leptomeninges at the anterior and lateral surfaces of the spinal cord, brainstem, and base of the brain. Therefore, benign and malignant melanomas derived from melanocytes develop anteriorly or laterally. The pia mater also

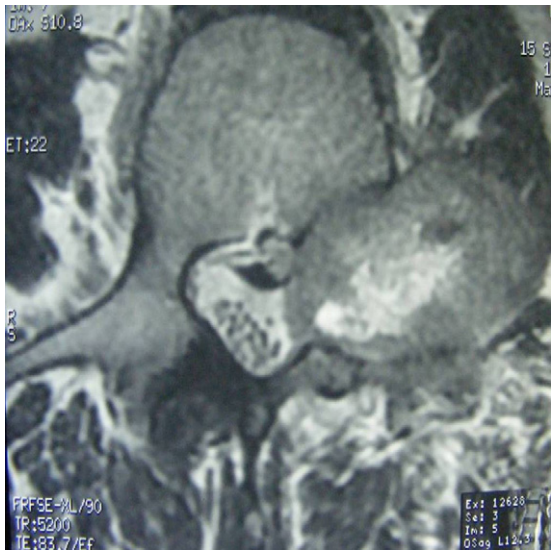

Fig. 2. T2-weighted axial MRI shows areas of hemorrhage within the tumor.

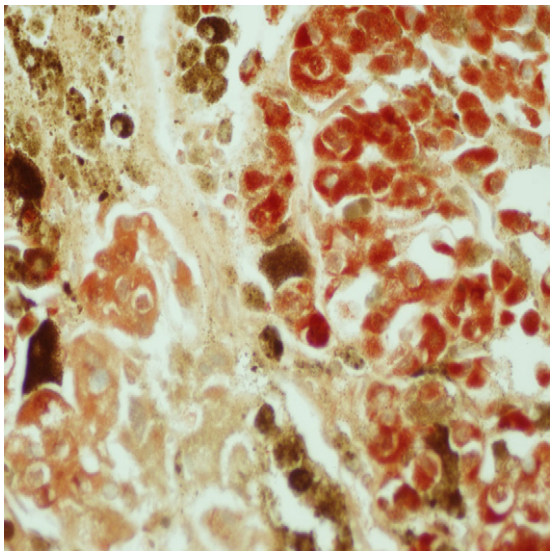

Fig. 3. Pathological evaluation based on immunohistochemical staining. HMB45 positive tumor cells (HMB45-AEC).

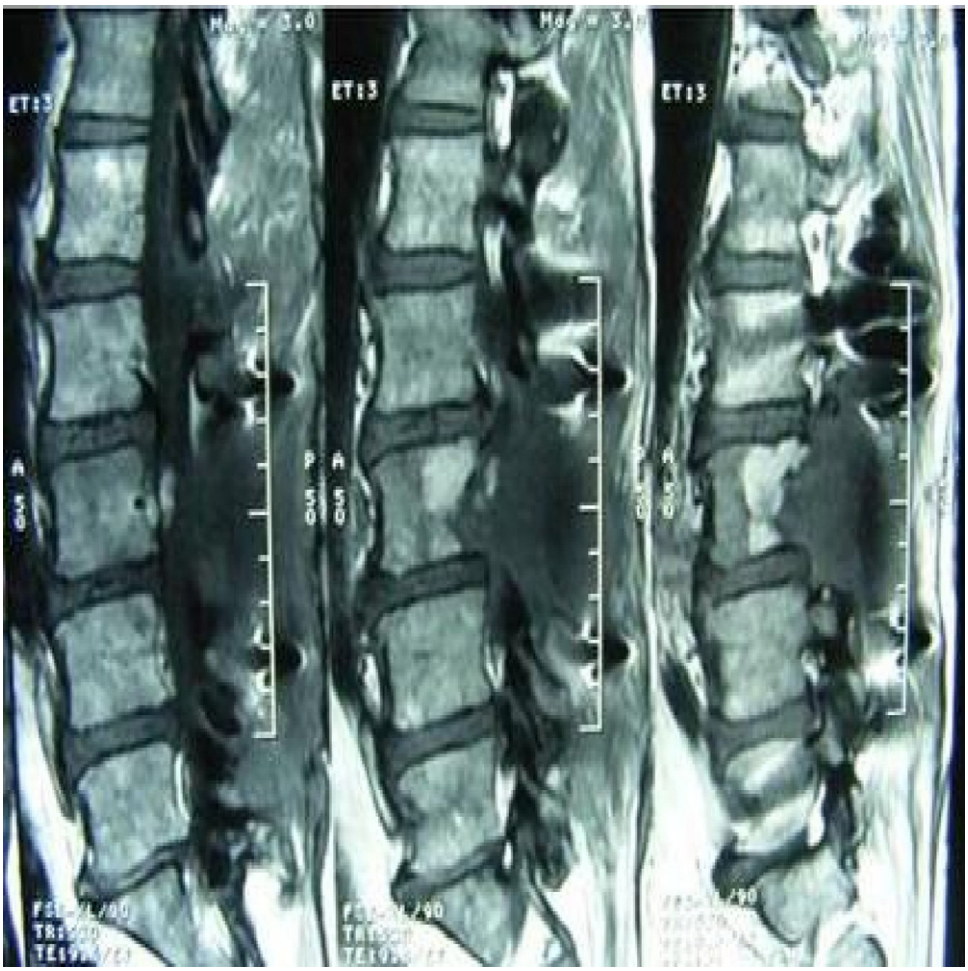

Fig. 4. A postoperative MR image was obtained 1 month later and revealed no residual tumor in the lumbar region.

contains melanocytes and covers the brain, brainstem, spinal cord, and nerves, and malignant melanoma can occur in these areas. Such seeding of the pia may also represent spread via the cerebrospinal fluid (CSF) from a primary CNS tumor [2].

Of the malignant melanomas found in the CNS, 90% are metastases, which grow rapidly and usually lead to a fatal outcome in less than 6 months. By contrast, primary CNS malignant melanoma has a better prognosis. Some authors believe that the presence of the blood–brain barrier in the spinal cord prevents hematogenous spread and makes metastasis more difficult [5]. In our patient, melanotic meningioma of the spinal cord was excluded because the specimen was EMA negative and metastatic carcinoma with melanocytic differentiation was excluded because the tissue was PANCK and EMA negative.

Such a slow progression for a tumor that appears malignant pathologically is not even found in histologically benign melanocytoma [4]. Soo Kim et al. explained this using Callender's classification of uveal melanomas [5]. Callender classified uveal melanomas into three groups according to the shape and differentiation of the cells: spindle A cells (fine chromatin and no distinct nucleoli); spindle B cells (plumper nuclei, more nucleoli, and coarser chromatin); and epithelioid cells (pleomorphic) [1]. There are reports that the best prognosis is seen with pure spindle cell tumors, regardless of the type. A worse prognosis is seen in mixed tumors, and the worst prognosis is seen in epithelioid cell tumors. Some findings support this hypothesis using Callender's classification for primary CNS tumors. Nevertheless, our patient falls into the group with the worst prognosis, but she has had prolonged survival. Considering the slow progression of primary malignant melanoma of the spine, patients should undergo total excision, which can eliminate any neurological deficits and reduce pain. If this is impossible, the literature suggests that radiation therapy reduces pain [3]. With slow tumor progression, the physician should adopt a multidisciplinary approach that includes stabilizing the spine, radiotherapy, and chemotherapy with tight control, as this can result in a comfortable life without pain or neurological deficits in these cases.

References

[1] Brat DJ, Giannini C, Scheithauer BW, Burger PC. Primary melanocytic neoplasms of the central nervous system. Am J Surg Pathol 1999;23:745–54.
[2] Hayward RD. Malignant melanoma and the central nervous system. A guide for classification based on the clinical findings. J Neurol Neurosurg Psychiatry 1976;39:526–30.
[3] Ku A, Henry A, Tunkel R. Lumbosacral radiculopathy secondary to L5 metastatic melanoma of unknown primary. Arch Phys Med Rehabil 1996;77:307–9.
[4] Skarli SO, Wolf AL, Kristt DA, Numaguchi Y. Melanoma arising in a cervical spinal nerve root: report of a case with a benign course and malignant features. Neurosurgery 1994;34:533–7.
[5] Soo Kim M, Heum Yoon D, Ah Shin D. Primary spinal cord melanoma. J Korean Neurosurg Soc 2010;48:157–61.