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LETTER TO THE EDITOR

Giant occipital aneurysmal bone cyst caused to hydrocephalus in a child

Mesut Mete[1] · Yusuf Kurtulus Duransoy[1] · Celal Çinar[2] · Gülgün Ovali[3] · Peyker Temiz[4] · Mehmet Selcuki[1]

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Dear Editor:

This letter reports a case of hydrocephalus due to a giant occipital aneurysmal bone cyst (ABC) in a child. ABCs are benign, expansile, osteolytic lesions that represent 1–2% of all primary bone tumors, and are usually located in the metaphyses (50%) of long bones. Approximately 20% of these lesions occur in the spine and, thus, cranial ABCs, which represent 3–6% of such lesions [1], and occipital ABCs are extremely rare. A 12-year-old girl presented at our hospital with headache and vomiting; her history revealed head trauma 3 months prior and 2 months of headaches and intermittent vomiting with swelling in the left occipital region of her head. A physical examination revealed a non-painful mass in the left occipital region in the absence of neurological deficits. A computed tomography (CT) scan showed a partially cystic mass of approximately 6 cm with fluid–fluid levels arising from the left occipital calvarium that was surrounded by thin inner and outer bones (Fig. 1a). A magnetic resonance imaging (MRI) scan showed an expansile occipital bone that had fluid–fluid levels within it. There was significant cerebellar compression and the effacement of the fourth ventricle with obstructive hydrocephalus (Fig. 1b–c). A preoperative particular embolization using polyvinyl alcohol was performed to prevent unexpected intraoperative bleeding and the patient underwent a left sub-occipital craniectomy with a total excision of the lesion (Fig. 1d); there were no perioperative complications. The lesion was entirely extradural and was causing significant compression of the cerebellar hemispheres. The bulk of the tumor was removed in large pieces with the surrounding rim of the occipital bone. Macroscopic and histopathological evaluations of the tissue revealed an ABC without an associated lesion (Fig. 2a–b). The postoperative period was uneventful and the patient was discharged 4 days after the operation.

ABCs were first described by Jaffe and Lichtenstein in 1942 as tumor-like benign lesions with an unknown etiology. Currently, factors such as trauma, underlying neoplasms, and cytogenetic abnormalities are thought to be involved in the pathophysiology of ABCs [1] but the lesion could be secondary to hemodynamic disturbances associated with elevated venous pressure and hemorrhage induced by trauma [2]. In up to 30% of cases, ABCs are associated with other lesions including a giant-cell tumor, chondroblastoma, chondromyxoid fibroma, non-ossifying fibroma, osteoblastoma, fibrosarcoma, histiocytoma, and osteosarcoma [3]. Approximately, 50% of patients with ABCs show a chromosomal translocation [4] and these lesions can present with cranial nerve palsies, signs of raised intracranial pressure, epilepsy, and cerebellar symptoms [2, 5]. The lesions grow rapidly, with a mean symptom duration of 3 months after head trauma. Therefore, ABCs should be distinguished from leptomeningeal cysts because the latter are associated with continuous pulsatile pressure of the cerebrospinal fluid (CSF), and the expansion of the cyst causes the resorption of adjacent bone, which erodes bone edges and widens the skull fracture. However, the symptoms associated with this type of

✉ Mesut Mete
dr.mmete@hotmail.com

[1] School of Medicine Neurosurgery Department, Celal Bayar University, Yelken Evleri Sitesi B Blok k:5 d:16 Güzelyurt, Manisa, Turkey

[2] School of Medicine Department of Radiology, Ege University, İzmir, Turkey

[3] School of Medicine Department of Radiology, Celal Bayar University, Manisa, Turkey

[4] School of Medicine Department of Pathology, Celal Bayar University, Manisa, Turkey

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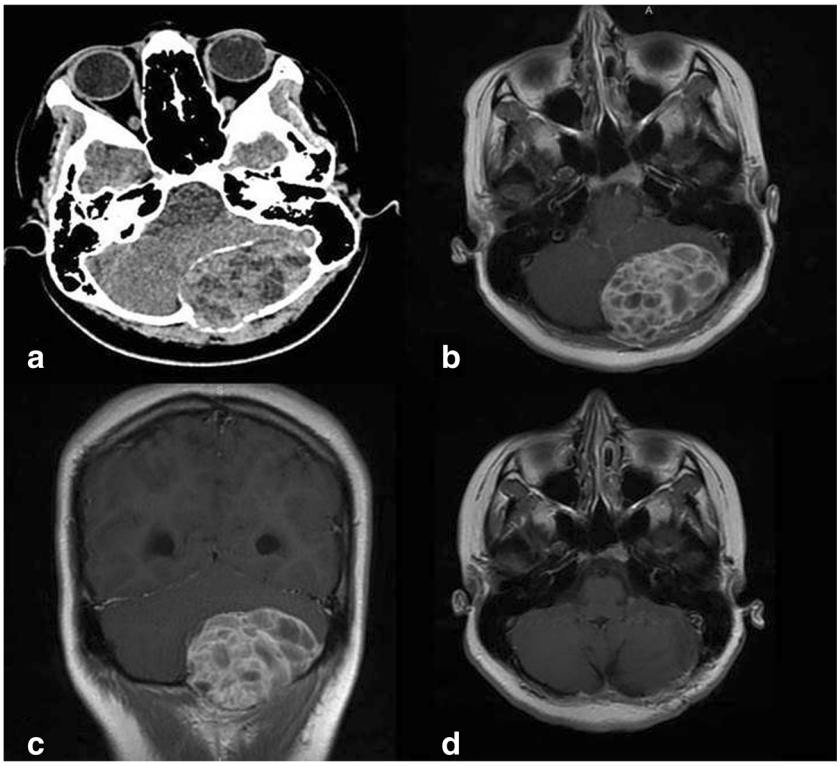

Fig. 1 CT scan showed a partially cystic mass of approximately 6 cm with fluid–fluid levels arising from the left occipital calvarium that was surrounded by thin inner and outer bones (a). MRI scan showed an expansile occipital bone that had fluid–fluid levels within it. There was significant cerebellar compression and the effacement of the fourth ventricle with obstructive hydrocephalus (b–c) and total excision of the lesion (d)

cyst, such as raised intracranial pressure and epilepsy, are similar to those of ABCs [5]. On CT scans, ABCs are well circumscribed, with multiloculated expansile changes in the diploic space [3] and cysts that are separated by septa and covered with thin cortical bone. MRI scans typically show a well-delineated expansile mass with enhancing septations that separate cystic components containing fluid–fluid levels with heterogeneous signals, which may be attributed to the various stages of hematoma. However, the signal intensity of a leptomeningeal cyst is equal to that of CSF on both T1- and T2-weighted MR images (5). In the present case, the patient suffered from headache, vomiting, and non-painful swelling that rapidly intensified in the 3 months after the head trauma. Radiological investigations revealed the presence of an ABC and, thus, the patient underwent surgery; histopathological evaluations of the lesion demonstrated the absence of an underlying tumor.

The treatment of choice for ABCs is an en-bloc resection to prevent recurrence [1]; recurrence rates vary from 20 to 70% with incomplete surgical removal. If the lesion is located in the skull base, orbit, or paranasal sinuses, it can be difficult to manage the tumor. In such cases, the partial excision or intralesional curettage of the lesion with adjunctive therapies, such as embolization, cryotherapy, or radiotherapy, should be considered [3]. In the present case, preoperative embolization was performed to prevent unexpected perioperative bleeding and the lesion was restricted to the occipital bone, which permitted total excision.

In conclusion, although cranial ABCs are rare, these lesions should be considered in the presence of rapidly growing

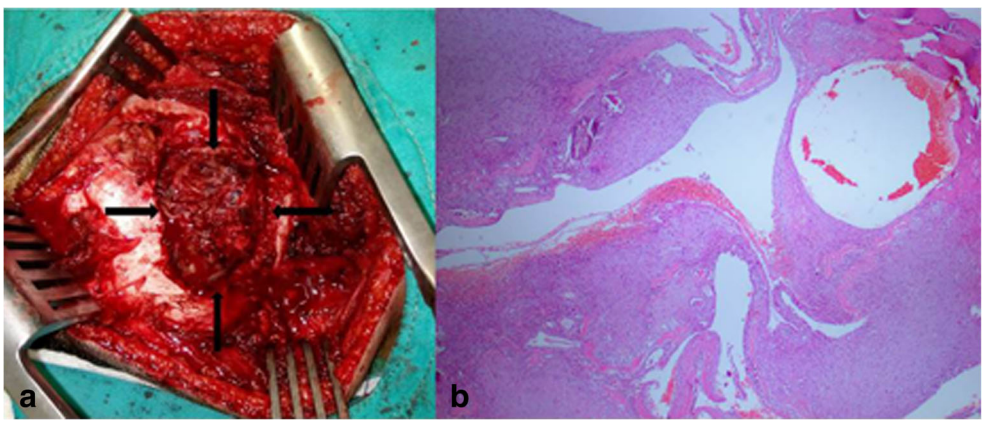

Fig. 2 Perioperative appearance of tumor tissue (black arrows) (a). Histopathological evaluations of the tissue revealed blood-filled cystic areas covered by fibroblasts, osteoclast-like giant cells and newly formed bone tissues (b)

soft tissue and swelling of the scalp following head trauma. If this diagnosis is confirmed via CT and MRI, treatment modalities should include preoperative embolization and total resection, as much as is possible, depending on the circumstances.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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