

## Occipital Aneurysmal Bone Cysts in an Adult Patient: A Rare Case and Review of Literature

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### ABSTRACT

*This study aimed to report a rare case of occipital aneurysmal bone cysts (ABCs) in an adult patient. ABCs account for 1% of primary bone tumors. They are usually seen in pediatric age and rare in adults. They are benign fibro-osseous lesions, but they also tend to double in size rapidly and destruct the morphology of the bone. They have a tendency to extend to the long bones and vertebral column. The calvarial ABCs are 3%–6% of ABC. They usually affect the frontal and temporal bones of the skull, and cranial nerve findings are frequently seen. In the adulthood, occipital ABCs are extremely rare. Sixteen cases of occipital ABC, average age 15 years, have been reported in the literature. A 50-year-old female patient was admitted to the clinic with the history of an occipital mass lesion, which had been growing for 6 months. Magnetic resonance imaging and computed tomography of the cranium revealed an irregular-shaped destructive lesion of 55 × 18 × 28 mm<sup>3</sup> in the diploe part of the occipital bone. The mass with an approximately 6 cm radius, and 1 cm normal bone edge was completely excised. Because of total resection, no adjuvant therapy was performed. Neither recurrence nor residual lesion was observed in the postoperative 24 months.*

*Key words: Adult, aneurysmal bone cyst, occipital*

### INTRODUCTION

Aneurysmal bone cysts (ABCs), first described in 1942 by Jaffe and Lichtenstein, are uncommon, benign, rapidly growing osteolytic lesions consisting of two segments: an intraosseous, destructive tumor and an extraosseous, aneurysm-like cyst of the bone, which make up for 1%–2% of all primary osseous tumors (1). The true etiology and pathophysiology are not clear. It is believed that ABCs are a result of a vascular malformation that mostly affects the metaphysis of the long bones. They are benign fibro-osseous lesions but they also tend to double in size rapidly and destruct the morphology of the bone. They have a tendency to extend to the long bones and vertebral column (2–7). They are usually seen in pediatric age and rare in adults. Only 3%–6% of all ABCs present with extra- or intracranial extension found in the skull. They usually affect the frontal and temporal bones of the skull, and cranial nerve findings are frequently seen. In the adulthood, occipital ABC is extremely rare. This study reported a rare case of occipital ABC in an adult patient with clinical and radiological features and treatment strategy.

### CASE REPORT

A 50-year-old female patient was admitted to the clinic with the history of an occipital mass lesion, which had been growing for 6 months. Magnetic resonance imaging and computed tomography (CT) of the cranium revealed an irregular-shaped destructive lesion of 55 × 18 × 28 mm<sup>3</sup> in the diploe part of the occipital bone (Figs. 1 and 2). The mass with an approximately 6 cm radius, and 1 cm normal bone edge was completely excised, and cranioplasty with a titanium mesh was performed (Fig. 3). Immunohistochemical studies (SMA: Smooth Muscle Actin, EMA: Epithelial Membrane Antigen, CD34, ALK: Anaplastic Lymphoma Kinase, CD117, pancytokeratin, vimentin, desmin, and S100) revealed an ABC. Because

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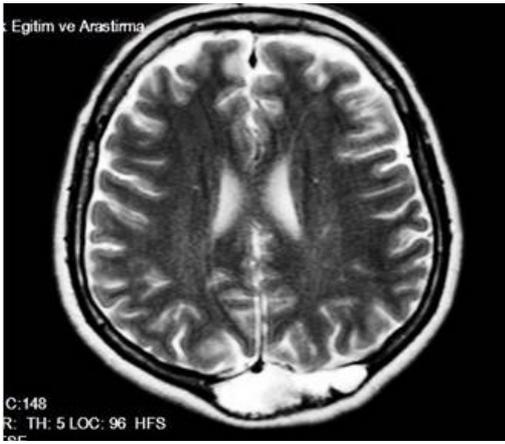


FIGURE 1: Axial MRI revealing a destructive lesion in the occipital bone.

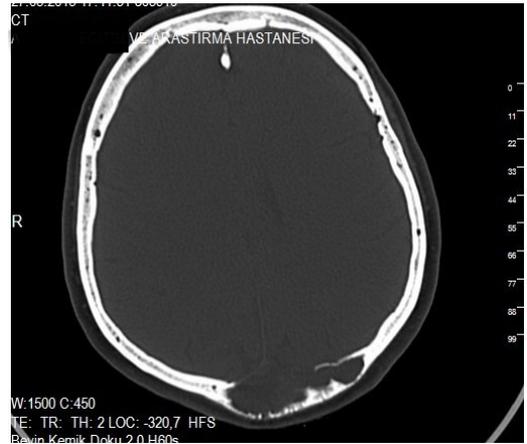


FIGURE 2: CT scan showing the occipital bone with a locally destructive mass.



FIGURE 3: Postoperative CT scan showing total resection of the lesion and cranioplasty with a titanium mesh.

of total resection, no adjuvant therapy was performed. Neither recurrence nor residual lesion was observed in the postoperative 24 months.

## DISCUSSION

Accounting for 1%–2% of all osseous tumors, ABCs are rare and benign lesions, but they tend to behave destructively locally. Their characteristic blown-out appearance arises from a blood-filled fibrous tumor-like cyst that expands the surrounding bone. They are most commonly found in the vertebrae and metaphysis of the long bones. (2,3,6,8–10). Only a small percentage of ABCs are found in the skull and mandible, ranging from 3 to 6%. They are mostly diagnosed during an inspection where a scalp mass is observed but may also present as an intracranial mass or cerebral

hemorrhage. Most ABCs that are found in the skull tend to lie in the temporal bone and very rarely in the occipital bone (6,10–12). Only eight cases of adult occipital ABCs have been reported in the literature till date; the present case is the ninth case (Table 1). Although the specific pathogenesis of ABC is unexplained, the most commonly accepted concept is the formation of aneurysmal cysts following benign or malignant lesions. However, only one-third of the cases have an identifiable initial lesion. Giant cell tumor (19%–39%) is the leading precursor followed by osteoblastoma, angioma, and chondroblastoma. Fibrous dysplasia, unicameral bone cyst, nonossifying fibroma, chondromyxoid fibroma, fibrous histiocytoma, eosinophilic granuloma, and osteosarcoma are the least common causes (6,11). In 1942, ABCs were defined by Jaffe and Lichtenstein

TABLE 1: Summary of cases of adult occipital aneurysmal bone cysts reported in the English literature

Age/Sex	Location	Pathology	Clinical Presentation	Author	Year
19/Female	Occipital	Aneurysmal bone cysts	Headache and palpable mass	Luccarelli et al.	1980
18/Male	Occipital	Aneurysmal bone cysts	Palpable mass	Bilge et al.	1983
21/Male	Occipital	Aneurysmal bone cysts	Focal tenderness	David et al.	1993
19/Male	Occipital	Aneurysmal bone cysts + Fibrous dysplasia	Focal tenderness	Itshayek et al.	2002
35/Male	Temporal + Occipital	Aneurysmal bone cysts + Fibrous dysplasia	Headache	Iseri et al.	2005
19/Female	Parietal + Occipital	Aneurysmal bone cysts + Fibrous dysplasia	Headache	Mattei et al.	2005
54/Female	Occipital	Aneurysmal bone cysts	Focal tenderness	Lin et al.	2007
20/Male	Occipital	Aneurysmal bone cysts + Osteoblastoma	Headache	Han et al.	2008
50/Female	Occipital	Aneurysmal bone cysts	Palpable mass	Present case	

as lesions with distinct radiological finding of distended and ballooned-out periosteum (1). Although benign, ABCs are locally destructive causing elevation of the periosteum above and eroding the cortical bone to a very thin border, resulting in the typical “blown-out” or “soap bubble” appearance. In the vault bone, the lesion originates within the diploic space expanding the distance between the inner and outer tables of the cranium. CT and MRI are useful in establishing the diagnosis where multiloculation of the cyst demonstrates the characteristic fluid levels within the cyst. These levels are caused by partly cystic and partly solid content of the lesion where the solid part is clearly visible with contrast enhancement. Due to the breakdown of blood contents within the cyst, septa are formed and a heterogenous lesion can be observed in the MRI, which would also confirm the multiple fluid levels. Histological examination usually reveals cysts lined with spindle-shaped fibroblasts surrounding the venous blood-filled lesion. Multinucleated giant cells along with stromal cells may also be scattered within the lesion (6,8,9,13,14).

Treatment modalities range from simple surgery to a combination of methods. Although curettage followed by bone grafting is the most commonly employed modality, it has a high recurrence rate of 20%–40% (7,11,15,16). In the present case, the cyst was completely and aggressively scraped out to decrease the chance of recurrence. The remaining cavity was filled with donor bone tissue (allograft), bone chips (autograft), or other materials (2,3,6,7,8).

A marginal or wide excision of the surrounding bone is the

suggested surgical modality to decrease the chances of recurrence. If the lesion is found in an expandable bone such as the rib or fibula, aggressive wide excision is recommended (6,12,13,17). In the present case, an extensive excision of the mass with 1-cm normal bone edge was performed. Adjuvant therapy such as cryotherapy may be employed to decrease the chances of recurrence. If other modes of treatment prove inefficient, adjuvant radiotherapy following aggressive excision may also be performed.

## CONCLUSION

Adult occipital ABCs are extremely rare tumors. It is believed that total resection with a 1-cm normal bone margin of ABC does not need any adjuvant therapy. Routine controls must be done, and adjuvant therapies should be advised for the partially resected and recurrent lesions.

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