

Aneurysmal Bone Cyst of the Sphenoid Bone Extending into the Ethmoid Sinus, Nasal Cavity and Orbita in a Child

Çocukta Etmoid Sinus, Nasal Kavite ve Orbitaya Uzanan Sfenoid Kemik Kisti

ABSTRACT

An aneurysmal bone cyst (ABC) typically involves the long bones of the extremities, thorax, pelvis, or vertebrae. Skull base involvement is rare. We describe the case of a 9-year-old girl with ABC of the skull base. The patient had presented with nasal obstruction and headache over a period of approximately 8 months. The patient had no history of trauma or surgery. Physical and neurological examination findings normal. Magnetic resonance imaging (MRI) showed a multicystic lesion arising from the sphenoid sinus and extending into ethmoid sinus, superior nasal cavity and medial walls of the orbit. The lesion contained thin internal septations that revealed high signal characteristics on all sequences. The lesion was resected via an extended frontal approach without any complications. Histological evaluation confirmed that the lesion was an ABC. The patient did not receive postoperative radiotherapy. No recurrence was observed after 22 months. ABC should be considered in the differential diagnosis of bone neoplasms in this region.

KEYWORDS: Aneurysmal bone cyst, Sphenoid sinus, Ethmoid sinus, Superior nasal cavity, Orbita, Skull base

ÖZ

Anevrizmal kemik kisti (ABC) tipik olarak ekstremitelerde uzun kemikleri, toraks, pelvis veya vertebrayı tutar. Kafa tabanı tutulumu nadirdir. Biz kafa tabanı ABC'si olan 9 yaşında bayan hastayı sunuyoruz. Hasta 8 aydır devam eden burun tıkanıklığı ve baş ağrısı şikayeti ile başvurdu. Hastada travma veya cerrahi öyküsü yoktu. Fiziksel ve nörolojik muayene normaldi. Manyetik rezonans görüntülemesinde etmoid sinüs, süperior nasal kavite ve orbitanın medial duvarlarına uzanan sfenoid kemikten kaynaklanan multikistik lezyon görüldü. Lezyon tüm sekanslarda yüksek sinyal veren ince septalar içermekte idi. Lezyon herhangi bir komplikasyon olmadan genişletilmiş frontal yaklaşım ile rezekt edildi. Histolojik değerlendirmede lezyon ABC olarak tanımlandı. Hasta postoperatif dönemde radyoterapi almadı. 22 aydır rekürrens görülmedi. ABC bu alanın kemik tümörlerinin ayırıcı tanısında düşünülmelidir.

ANAHTAR SÖZCÜKLER: Anevrizmal kemik kisti, Sfenoid sinüs, Etmoid sinüs, Süperior nasal kavite, Orbita, Kafa tabanı

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INTRODUCTION

The most common lesions of the anterior cranial base are encephalocele, fibrous dysplasia and esthesioneuroblastoma in children (18). Aneurysmal bone cyst (ABC) rarely seen, is a benign osseous lesion recognized as a distinct clinicopathological entity by Jaffe and Lichtenstein et al. in 1942 (9). Approximately, 36 to 50% of all ABCs are found at the end of long bones and 25% involve the vertebrae with only 3% located in the cranium (4). ABCs occurred at various localizations in the calvarium and the skull base, including the occipital bone, frontal bone, temporal bone, parietal bone, sphenoid bone, and ethmoid bone (4,13). ABC of the sphenoid bone can be localized at the sphenoid wing with expansion to adjacent areas such as the orbital cavity (5,8,17) and sphenoid sinus (1,3,6,10,14,19). We present a child with ABC of the sphenoid sinus extending to the ethmoid sinus, superior nasal cavity and medial walls of the orbit. The tumor was resected almost totally via an extended frontal approach without any complications.

CASE REPORT

A 9-year-old girl presented with nasal obstruction and headache over a period of approximately 8 months. She had no history of trauma or surgery. Physical and neurological examination were normal. Her serum prolactin, growth hormone, cortisol, free thyroid hormones (FT3 and FT4), and thyroid stimulating hormone levels were normal. Her visual acuity was 6/6 bilaterally and visual fields were full.

Radiology: Brain computed tomographic (CT) scan showed an extra-axial mass arising from the sphenoid sinus and extending to the ethmoid sinus, superior nasal cavity and medial walls of the orbit (Figure 1A). Brain CT also showed multiple fluid-fluid levels and trabeculae. Magnetic resonance imaging (MRI) showed a large, well defined extra-axial mass lesion. A mass with dimensions of 5 cm x 4 cm x 6 cm and a well-defined capsule was detected. The lesion arising from the sphenoid sinus extended to the ethmoid sinus, superior nasal cavity, the medial walls of the orbit, internal walls of the cavernous sinus, and the clivus. There was no longer a clear posterior border between the tumor and the sella turcica to the clivus (Figure 1B). The lesion contained thin internal septations that revealed high signal characteristics on all sequences. Cerebral

angiography demonstrated the mass was supplied exclusively by the left internal maxillary artery. The arterial feeder was occluded using the endovascular embolization technique (Figure 1C).

Operation and follow-up: A day after embolization, the mass was excised through an extended frontal approach. A bicoronal skin flap with bilateral osteotomy of orbital and zygomatic process was undertaken. The mass was entirely extradural in nature. Macroscopically, the tumor was cystic, hemorrhagic in some areas and whitish-grey in appearance. The inner surface was smooth with multiple bony compartments. Nearly total excision of lesion was performed. At the end of the resection, areas of normal bone of a firmer consistency could be seen along the periphery. The dura mater was intact underneath the lesion. A wide extended subgaleal fascia pericranial flap from both frontal regions and fat tissue was used to cover the region of the bone defect. Intraoperative bleeding was minimal (about 300 cc). Postoperative early MRI demonstrated removal of the tumor (Figure 1D). Olfactory function was not intact after surgery. She did not receive postoperative radiotherapy. No recurrence was observed after 22 months.

Histopathological examination: Histopathological examination showed cavernous spaces filled with blood. Large areas of extravasated blood were seen. The spaces were separated by collagenous tissue containing fibroblasts, focal collections of osteoclastic giant cells, and reactive bone formation (Figure 2). Histological evaluation confirmed that the lesion was an ABC.

DISCUSSION

ABCs may be primary or secondary. The primary ABC has no identifiable preexisting lesion or history of trauma. Periosteal trauma has been put forward as an important etiological factor (4,15). ABCs in the presence of another lesion are called secondary ABCs such as skeletal pathology, including fibrous dysplasia, giant cell tumors, chondroblastoma, chondromyxoid fibroma, nonossifying fibroma, fibrous histiocytoma, osteoblastoma, and osteosarcoma (4,15).

The ages of patients with ABC of sphenoid bone and extending into the adjacent areas, ranged from 4 years to 49 years (Table I). No significant difference was noted males and females with ABCs of skull (15). ABCs of the sphenoid bone have a predilection for the male sex. Symptoms include headache, ptosis, strabismus,

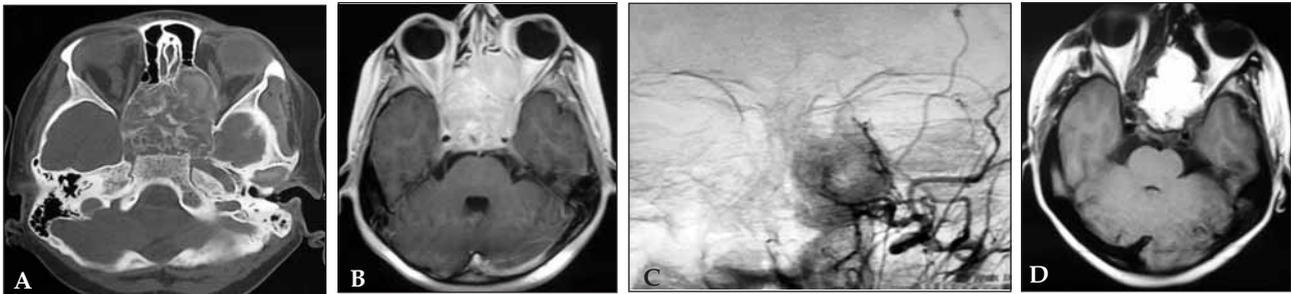


Figure 1: A- Axial CT showing the extradural extension of the tumor. There is a bony outer shell and the multiple, thin bony septae present a honeycomb appearance. B- MRI with contrast medium revealing the tumor as filling the sphenoid sinus and spreading the superior nasal cavity, medial walls of orbit, ethmoid sinus, and clivus. C- Left internal carotid artery subtraction angiogram showing tumor blush through the left internal maxillary arteries. D- T1-weighted axial MRI with contrast medium revealing the tumor was nearly total excised.

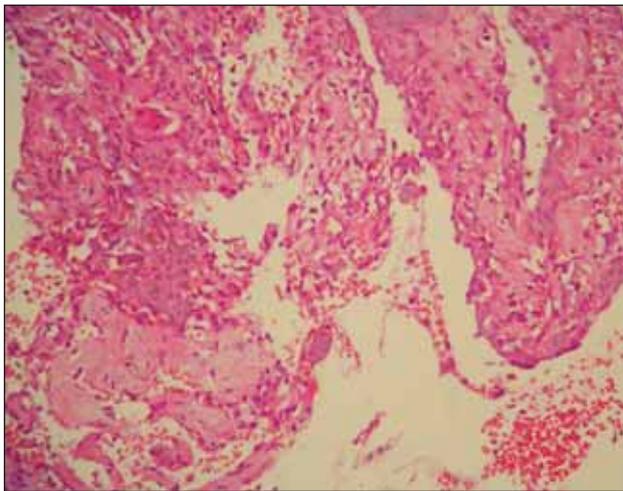


Figure 2: Photomicrograph showing the blood-containing cystic spaces. The cystic spaces were separated by collagenous tissue containing fibroblasts, focal collections of osteoclastic giant cells and reactive bone formation (Hematoxylin and eosin stain).

exophthalmus, double vision, swelling, visual loss, and nasal obstruction at ABC of sphenoid bone (Table I).

ABCs show some characteristic features on radiological examination (7,13). CT images disclose erosion, thinning of cortex and ridges in the bony walls. Fluid levels on bone CT scan images are seen in only 30% of the lesions (7). The CT scan image demonstration of fluid levels did not correlate with the type of fluid found within the cyst. These cysts are most commonly filled with blood. Sometimes, it can be a serosanguineous, straw-colored fluid, thick fluid of the consistency of "crankcase oil" (7). MRI shows well-defined lesion with heterogeneous signal intensity on T1-weighted images, well-defined hypointense capsule, and a homogenous increase in signal intensity on T2-weighted images (13). The fluid-fluid levels in the mass

on the T2-weighted images are characteristic of, though not specific for, ABC. Digital subtraction angiography (DSA) can sometimes reveal the blood supply to these vascular lesions and may also occasionally reveal arteriovenous shunts. Some reports showed that the mass was a vascular lesion and blushed (13) while other reports showed an avascular lesion (11).

Treatment regimens for ABCs include sclerotherapy, embolization, radiotherapy, simple curettage, surgical excision, or some combination of these methods. Percutaneous sclerotherapy offers the least invasive initial therapeutic option when compared with surgery or endovascular strategies reserved for resistant lesions (4). When the surgical decompression is hazardous (possible damage to neural structures, massive hemorrhage) or in high-risk patients, sclerotherapy alone can be used at ABC of cranium (4,12). Curettage is successfully used to treat ABCs. But, recurrence rates after curettage are between 21% and 50% in cases with cranial ABCs treated (13). Surgical excision is the ideal treatment of choice for ABCs (3,10,17). Transcranial-transbasal approach, a transfacial approach or anterior craniofacial approach can be used for this purpose in ABCs of the sphenoid bone (3). The lesion in our case was resected with an extended frontal approach. The extended frontal approach includes bifrontal free bone flap and orbitoethmoidal osteotomy. This approach can be used for midline tumors of the whole clivus and with extension into the sphenoid region. Total surgical resection is the best treatment option in that case. Preoperative embolization of cranial ABC may make surgery easier in selected cases and can reduce the vascularity of the lesion, as well as decreasing intraoperative blood loss as a preoperative adjunct therapy (2,3). Radiotherapy is generally not

Table I. Cases of aneurysmal bone cyst involving the sphenoid bone.
(Ref.: reference)

Ref.	Age (years), sex	Signs & symptoms	Location	Pre-op embolization	Treatment	Follow-up
10	10/M	visual loss	sphenoid sinus	(-)	resection	6 years
19	10/M	visual loss	sphenoid & ethmoid sinuses	(-)	resection	1 year
16	49/F	headache, anosmia	sphenoid & ethmoid sinuses	(-)	resection	undefined
14	11/M	nasal obstruction	sphenoid sinus	(-)	resection	40 months
3	14/F	headache	sphenoid & ethmoid sinuses	(+)	resection	3 years
1	17/M	headache	sphenoid& ethmoid sinuses	(-)	resection	30 months
6	28/M	headache,diplopia	sphenoid sinus	(-)	resection	3 years
8	7/M	periorbital swelling, decreased visual acuity	sphenoid wing & orbital involvement	(-)	resection	1 years
4	4 (sex undefined)	ptosis, diplopia	sphenoid bone	undefined	resection and sclerotherapy	undefined
2	4/M	ptosis, strabismus	sphenoid wing	(+)	sclerotherapy	2 years
17	14/F	headache, ptosis, strabismus	sphenoid wing & orbital and temporal fossa involvement	(-)	resection	undefined
5	5/F	exophthalmus, double vision, swelling	sphenoid wing & orbital and temporal fossa involvement	(-)	resection	3 years
Our case	9/F	nasal obstruction, headache	sphenoid & ethmoid sinuses, superior nasal cavity	(+)	resection	22 months

used because of the risks of postradiation neoplasm development, the benign nature of the ABC lesion, and the high rate of cure with total excision. However, radiotherapy seems to be effective for recurrent cases of ABC and with satisfactory results (12).

Consequently, lesions of the sphenoid bone extending to the adjacent areas of the skull base is rarely seen. This should be kept in muding during the differential diagnosis of sphenoid bone lesions. Although the lesion is benign, it might be necessary to use the skull base approach when it settles in complex regions like the sphenoid bone.

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