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Greater wing of sphenoid, the home for aneurysmal bone cysts: a case report

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Abstract

Background: Finding an aneurysmal bone cyst in the skull is rare and for a neurosurgeon to come across such lesions in the sphenoid bone with orbital extension is even rarer.

Case presentation: We report a case of a 16-year female who presented with a three-month history of headache, proptosis, and deterioration of vision. Pre-operative imaging studies which included NCCT head and MRI brain, suggested the lesion to be an aneurysmal bone cyst of the greater wing of the sphenoid, with extension into the orbit. Intraoperative findings did corroborate with the preoperative imaging findings and were again confirmed later from the histopathology report.

Conclusion: Aneurysmal bone cysts of sphenoid bone with orbital extension, though rare, can be excised completely, without hampering the cosmesis. Being benign, patients can have a prolonged recurrence-free period if the lesion is completely excised.

Keywords: Aneurysmal bone cysts, Greater wing of sphenoid bone, Skull base

Background

Aneurysmal bone cysts, as a new clinicopathologic entity was first described by Jaffe and Lichtenstein in 1942 [1]. These lesions commonly affect the metaphysis of long bones. Involvement of skull bones is rare (3%) and sphenoid bone aneurysmal bone cysts with orbital extension are even rarer [2, 3]. These handful cases present mostly in their first three decades of life. We report a case of an aneurysmal bone cyst involving the greater wing of sphenoid bone with extension into the lateral orbital wall, which was totally excised without any cosmetic defect.

Case presentation

A 16-year girl presented with 3 months history of occasional headache and gradually progressive painless diminution in vision in the right eye. On examination, there was fullness in the right periorbital area with slight proptosis. It was nontender and the overlying skin was normal. Visual acuity was 6/12 in the right eye. There was no evidence of other neurologic deficits. NCCT showed a large heterogeneous lesion in the right anterior middle fossa and herniating anteriorly into the orbit through a defect in the posterior orbital wall. MRI showed a $4.3 \times 3.2 \times 3.4$ cm expansile lytic lesion arising from the greater wing of the sphenoid wing with multiple locules within, showing blood fluid level and enhancement of wall and septa (Fig. 1). Anteriorly it caused the obliteration of the right orbital space resulting in mild proptosis. Medially it compromises the optic nerve canal with compression of the intra-canalicular segment of the optic nerve.

We approached this lesion through right frontotemporal craniotomy. Tumour was well defined, encapsulated, involving the greater wing of the sphenoid with extension into the right orbit through the lateral wall. Posteriorly it extended up to the lateral wall of the sphenoid sinus. The lesion was solid cystic, with dark-coloured sanguineous spaces separated by fibrous septa. Gradually the whole lesion was curetted out with meticulous attention

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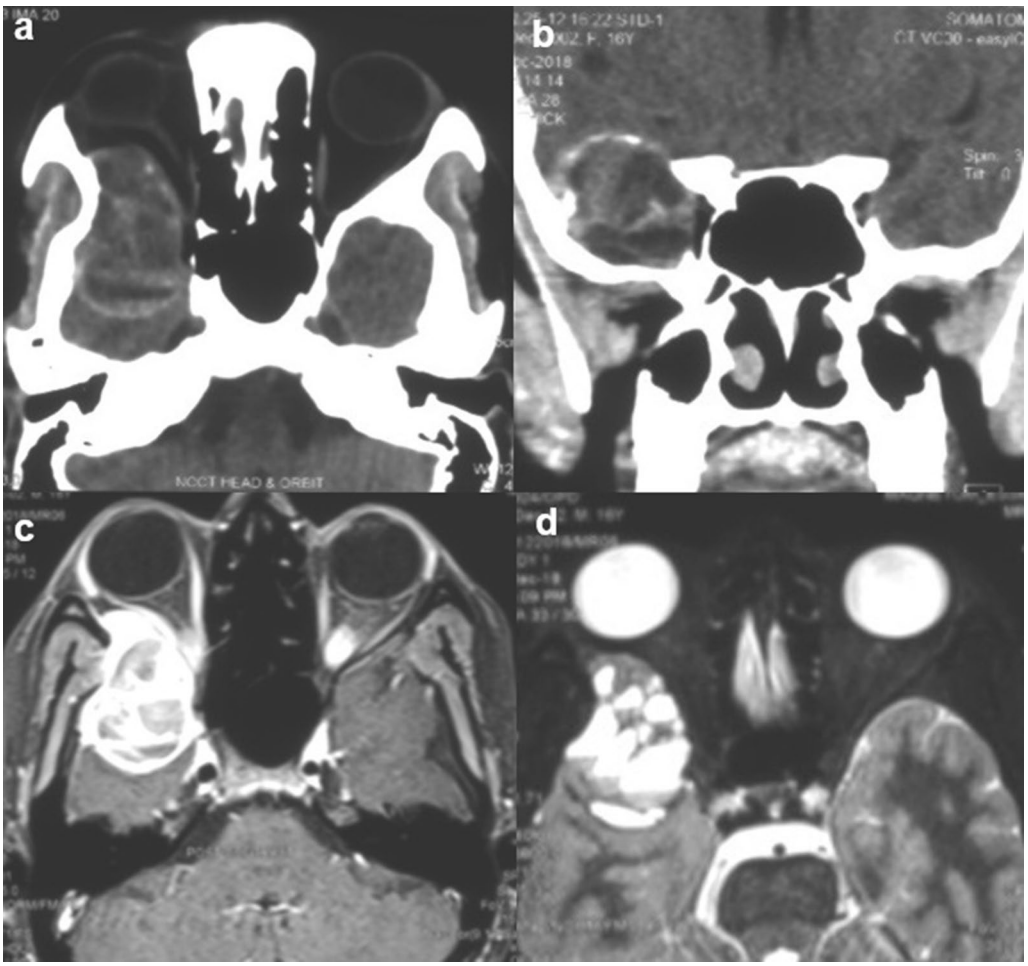

Fig. 1 The axial (**a**) and coronal (**b**) views of preoperative NCCT head showing a heterogenous lesion, extending anteriorly into the orbit and causing proptosis. Preoperative axial CEMRI (**c**) and T2 sequence (**d**) showing an expansile lytic lesion arising from the greater wing of sphenoid wing with multiple locules within, showing blood fluid level and enhancement of wall and septa

to hemostasis. Gross total excision was achieved and without leaving any residual cosmetic defect. In the postoperative period, there was the resolution of proptosis (Fig. 2). Her vision was intact at the time of discharge. Histopathology report did confirm the lesion to be an aneurysmal bone cyst (Fig. 3).

Discussion

Aneurysmal bone cysts of the skull constitute merely 3% of all the ABC cases. They mostly affect the cranial vault and present as palpable mass [4]. Involvement of cranial base is rarely seen and only a handful of spheno-orbital ABC have been reported to date [5]. ABCs arise either as a primary pathology or as a reactionary lesion secondary to various pathologies such as chondroblastoma, fibrous dysplasia, osteoblastoma, giant cell tumor, fibromyxoma, etc. [6].

Bone erosion thinned out cortex and heterogenous mass with multiple fluid levels are frequently seen on computed tomography. MRI demonstrates heterogeneous signal intensities on both T1 and T2 sequences, with multiple fluid levels suggesting hemorrhage with sedimentation. Hypointense peripheral rim with multiple internal septations that show contrast enhancement is a peculiar MRI finding [7, 8]. Though these imaging features are consistent, but not pathognomonic for ABC. Confirmatory diagnosis mandates evidence of histopathologic features like multiple septated fluid-filled spaces, without any endothelial lining. These septations are in fact lined by multinucleated giant cells, myofibroblasts, fibroblasts, and histiocytes [9].

There are various modalities of treatment and each is associated with a different degree of recurrence. En bloc excision and intralesional resection have good outcomes

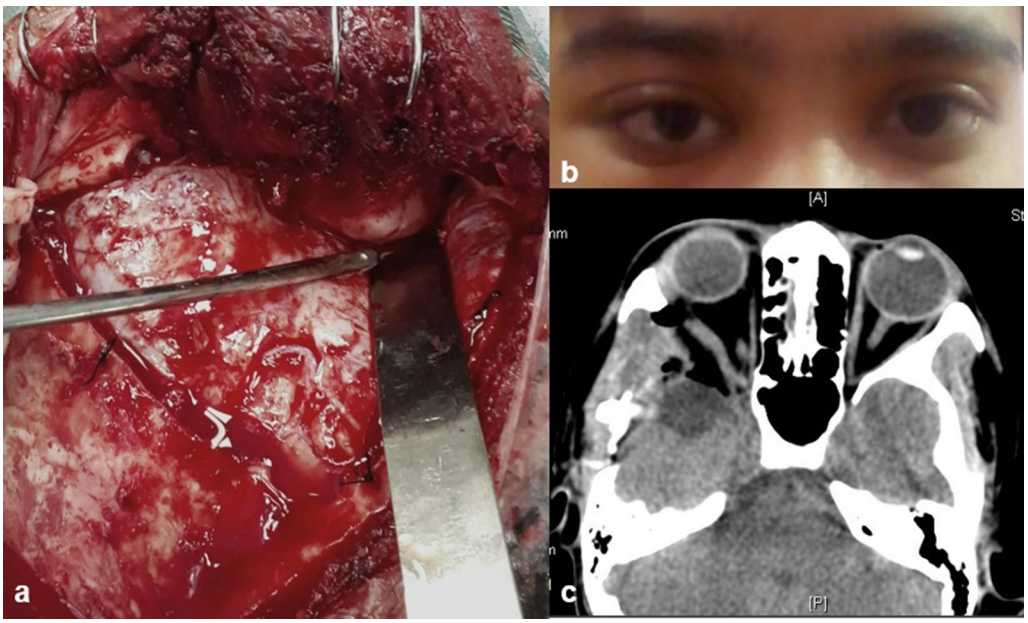

Fig. 2 **a** Intraoperative image showing an expansile lesion originating from the greater wing of sphenoid. **b** Clinical image showing resolution of proptosis in postoperative period. **c** NCCT head showing no evidence of recurrence after a follow-up period of 15 months

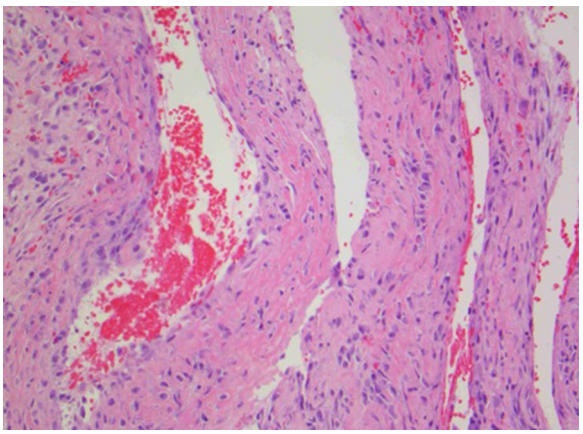

Fig. 3 Microscopically, it showed cystic spaces that are filled by blood and are separated by cellular septae, that consisted of giant cells and fibroblasts

concerning the rate of recurrence. Though en bloc excision achieves the least risk of recurrence, it is associated with the highest degree of morbidity [10]. Intralesional resection with high-speed burr mechanically disrupts the lesion up to the circumscribing healthy bone, thus achieves a local control rate similar to that of en bloc excision. Curettage alone carries a recurrence rate of 20–50% [12]. Although curettage with adjuvant therapies like sclerotherapy, cryotherapy, radiotherapy have been used for peripheral ABC, they can't be applied for spheno-orbital cases. Preoperative embolization can be used as a surgical strategy to decrease intraoperative blood loss by reducing the vascularity of lesions [13]. Although osteogenic sarcoma is a known complication in patients undergoing adjuvant radiotherapy for extracranial ABC, it has rarely been reported in cranial cases, thus radiotherapy has its role in recurrent cases [14]. In this case, we gained access to the lesion through the right frontotemporal corridor and achieved total excision through intralesional resection. At 15 months of follow-up, she had no recurrence.

Conclusions

Sphenoidal aneurysmal bone cysts constitute a rare clinicopathologic subgroup of ABC of the skull. Though benign, these lesions are locally aggressive and should be considered as a differential diagnosis in younger patients presenting with rapid onset painless proptosis. Intralesional resection with high-speed burr provides a viable option in achieving total resection, without leaving any cosmetic deformity and with a good degree of local disease control.

Abbreviations

ABC: Aneurysmal bone cysts; NCCT: Non-contrast computed tomography; MRI: Magnetic resonance imaging.

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Authors' contributions
DJM has designed, acquired, interpreted the data and then drafted the work. PS did the literature search, designed the work and revised the manuscript. All the authors have read and approved the manuscript and have agreed to be accountable for their contributions. All authors read and approved the final manuscript.

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Availability of data and material
All the information and data that are available, have been shared in the article and are genuine.

Declarations

Ethics approval and consent to participate
Ethical committee clearance was taken before including this case for study.

Consent for publication
Valid informed written consent of the guardian was taken to publish this case report, as patient was just 16 years old. They were informed that the details of patient will not be disclosed.

Competing interests
There are no financial and non-financial competing interests associated with this case report.

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