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CASE REPORT

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Fibrous dysplasia of sphenoid wing with secondary aneurysmal bone cyst: a rare case report

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Abstract

Background An aneurysmal bone cyst is a locally destructive benign lesion affecting mostly the long bones. Aneurysmal bone cyst of the skull bones is a very rare phenomenon and the involvement of the sphenoid bone of the skull with extension into the orbit is even rarer. We present a case of 15-year-old adolescent with fibrous dysplasia of the sphenoid wing with secondary aneurysmal bone cyst.

Case presentation A 15-year-old male presented to us with chief complaints of headache with swelling in the left temporal region of the face and proptosis of the left eye associated with decreased vision for the past 2 months. NCCT showed a large heterogeneous mass in the left temporal region extending into left orbit. Gadolinium-enhanced MRI showed a well-defined multiloculated osteo-expansile lesion in the left middle cranial fossa extending into the anterior cranial fossa consistent with the fibrous dysplasia of the sphenoid bone with associated aneurysmal bone cyst. Digital subtraction angiography brain to look for any feeders to the lesion was done followed by microsurgical gross total excision of the tumor. The histopathology report confirmed it to be fibrous dysplasia secondary to aneurysmal bone cyst.

Conclusion Aneurysmal bone cyst is a rare entity, commonly affecting the long bones of the body. The involvement of sphenoid wing of skull is very rare occurrence. It can be primary or secondary to fibrous dysplasia, chondroblastoma, giant cell tumor, fibromyomas, etc. Fibrous dysplasia with secondary aneurysmal bone cyst should be kept in mind as one of the differential diagnoses while dealing with osteolytic bone lesions of skull.

Keywords Aneurysmal bone cyst, Sphenoid wing, Temporal, Fibrous dysplasia

Background

An aneurysmal bone cyst is a benign, locally destructive and highly vascular bony lesion which was first described by Jaffe and Lichenstein in 1942 [1, 2]. These lesions frequently arise in the long bones, followed by the spine and flat bones of the pelvis [3]. The aneurysmal bone cysts can be primary or secondary depending on the presence or absence of the associated bony lesion. Mean age of presentation is 18.4 years, with 80% patients usually under 20 years of age [4]. Fibrous dysplasia associated with secondary aneurysmal bone cyst involving the skull vault is a very rare entity which was first reported by Branch in 1986 [5]. It usually affects the metaphysis of long bones, but rarely affects the skull bones [6].

Herein through this article, we report a case of an adolescent male presented to us with headache, proptosis and gradually progressing vision loss. Radiography suggested the lesion to be an aneurysmal bone cyst of the

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sphenoid bone extending to the orbit. We treated the patient with complete excision of the tumor and the final histopathology reported it to be a fibrous dysplasia with secondary aneurysmal bone cyst which is a very rare entity to occur. Hence, we want to highlight that this rare entity should be kept in mind as one of the differential diagnoses while dealing with the osteolytic lesions of the skull.

Case presentation

A 15-year-old male presented to us with chief complaints of swelling over left temporal region of face associated with headache and decreased vision in left eye for 3 months. He also had proptosis in left eye for past 2 months. He had no significant medical history in the past and he presented to us in good general condition. On examination, he was fully conscious and well oriented to time, place and person. He had a visual acuity of 6/18 in right eye and finger counting at 1 m in left eye as assessed by Snellen's chart. There was also fullness of the left peri-orbital space associated with proptosis of the left eye. He had a non-tender swelling over left temporal region which was bony hard and fixed to the underlying bone and having a smooth overlying surface. The rest of the neurological examination was grossly normal. There was no other abnormality on systemic examination. After admitting the patient, we got a plain CT (computed tomography) head done which revealed a large multiloculated heterogeneous mass in left middle cranial fossa and extending into left orbit from posterior orbital wall. On bony window with three-dimensional reconstruction of the plain CT head, there was erosion of the greater and lesser wings of left sphenoid bone along with squamous part of the left temporal bone and posterior wall of the orbit. Contrast-enhanced magnetic resonance imaging (CEMRI) of brain and orbit showed a well-defined extra-axial, multiloculated, osteolytic expansile lesion, measuring 61.5×38×53.9 mm, in the left middle cranial fossa and extension into the orbit with multiple loculated cyst containing blood fluid level and enhancement of the septations. The osteolytic lesion seemed to involve greater and lesser wings of left sphenoid bone along with a part of the left squamous temporal bone. Anteriorly it was extending into extraconal space of left orbit, abutting and displacing the left lateral rectus causing proptosis of left globe. Antero-inferiorly, it was causing mass effect on posterior wall of left maxillary sinus; posteriorly, it was involving left temporal region and causing buckling of surrounding brain parenchyma without bone invasion, medially it was abutting the lateral wall of left cavernous sinus with no evidence of brain invasion, superiorly the lesion was causing buckling of left temporal lobe with hyperintensity and causing stretching of left middle cerebral artery and its branches, and inferiorly lesion was extending into the infra temporal fossa (Fig. 1).

Since the lesion was appearing highly vascular, we planned for the digital subtraction angiography to look for any arterial feeder supplying the lesion and embolize it beforehand undergoing definitive surgery. But there were no significant arterial feeders to the tumor. The patient was then shifted to operation theater for microsurgical excision. Left temporal craniotomy was made, and gross total excision of tumor was done. En bloc resection of the osteolytic bony lesion including a satisfactory margin of healthy bone with curettage of the underlying soft tissue mass was done. The cavity developed after excision of the tumor was filled with fat harvested from lateral aspect of thigh, and the bony reconstruction of the skull base was done using bone cement. The overlying skull defect over the surface was reconstructed by cranioplasty using titanium mesh and fixed with mini plates and screws. The excised tumor tissue along with the excised lytic bony lesions was sent for histopathology.

The histopathological examination showed mixed cystic and solid components, fragments of bone with tumor comprising of solid areas with bland spindle-shaped cells in the collagenous matrix and cystic spaces filled with hemorrhage. Cystic spaces showed septations containing loosely arranged bland looking spindle to stellate cells. Based on these microscopic features, histopathology reported the lesion to be fibrous dysplasia with secondary aneurysmal bone cyst.

In the postoperative period, there was significant resolution of the proptosis of the left eye with complete relieve of the headache. There was significant improvement in the vision of the right eye with minimal improvement in the left eye vision from finger counting at 1 m to finger counting at 3 m at 1-month follow-up. There was no added neurological deficit postoperatively, and the patient was discharged with full Glasgow Coma Scale (GCS) score of 15/15. The patient is kept in close follow-up.

Discussion

An aneurysmal bone cyst is a benign locally destructive and highly vascular bony lesion which was first described by Jaffe and Lichenstein in 1942 [1]. These lesions frequently arise in the long bones, followed by the spine and flat bones of the pelvis [3]. The aneurysmal bone cysts can be primary or secondary depending on the presence or absence of the associated bony lesion. The commonly associated primary diseases with secondary aneurysmal cyst are chondroblastoma, osteoblastoma, osteosarcoma, giant cell tumor, hemangioma and fibromyxoma [7]. In the past literature, the incidence of the aneurysmal bone cyst involving the skull is reported to be less than 1% [8].

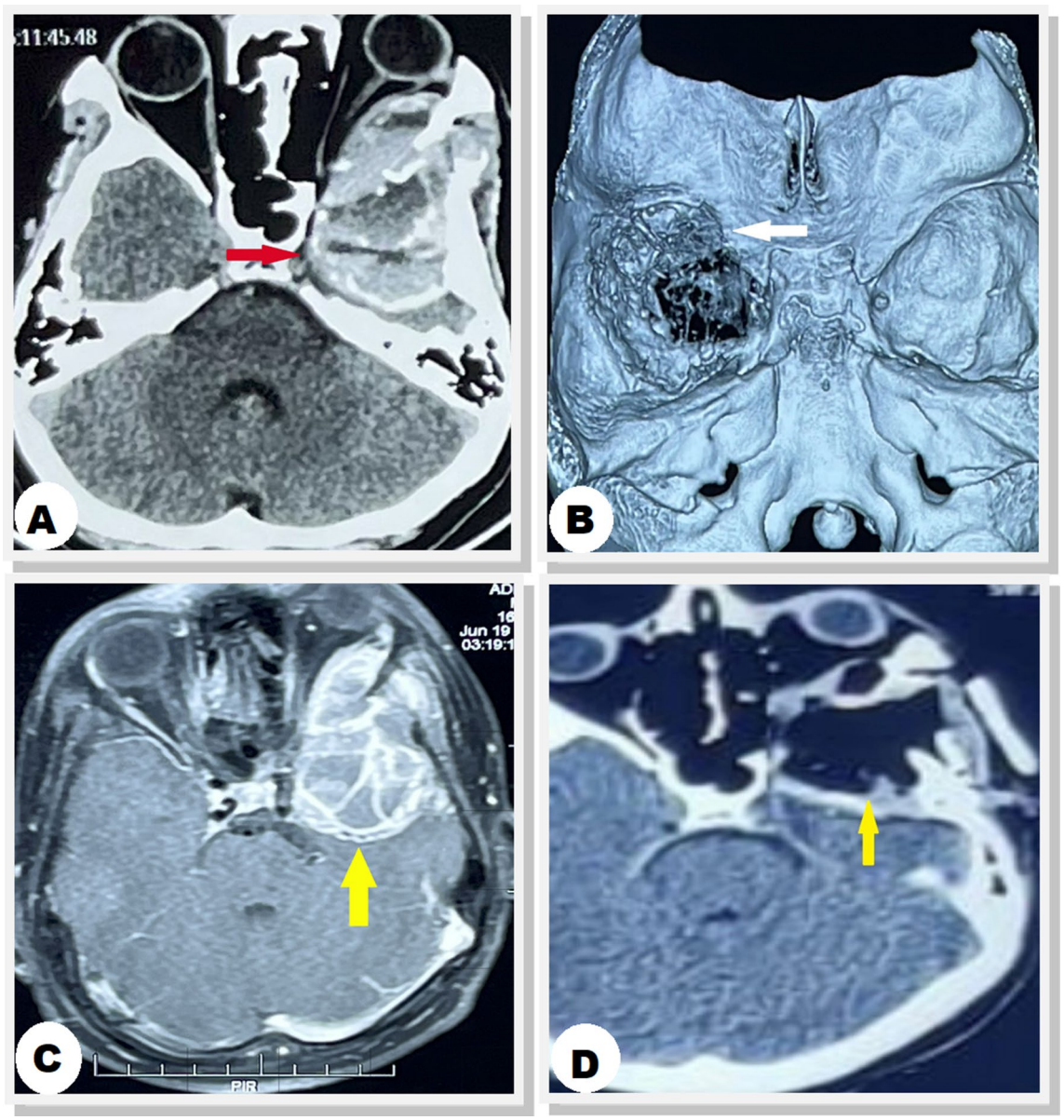

Fig. 1 **A** Preoperative axial image of NCCT head showing a large heterogeneous mass in left middle cranial fossa extending into left orbit (marked with arrow); **B** three-dimensional (3-D) reconstructed image of the skull showing the erosion of the base of the middle cranial fossa along with involvement of the left orbit and left temporal bone; **C** preoperative axial T1 contrast image showing multiloculated soft tissue lesion in left middle cranial fossa (marked with arrow); **D** postoperative NCCT head showing the complete excision of the osteo-expansile lesion

Fibrous dysplasia associated with secondary aneurysmal bone cyst involving the skull vault is a very rare entity which was first reported by Branch in 1986 [5]. In his case study, Martinez et al. found one case of aneurysmal bone cyst (ABC) in 42 patients of fibrous dysplasia [9].

The exact etiopathogenesis of the occurrence of ABC is not well known, and several hypotheses have been put forward in the past explaining their occurrence. One of the most important and widely accepted theory is the occurrence of ABC secondary to local trauma to the bone [10]. Lichenstein et al. suggested ABC can occur due to local circulatory disturbances secondary to sudden occlusion of the venous drainage leading to increased venous pressure locally resulting in formation of blood-filled spaces in the bone, and ultimately, this leads to the destructive lesions of the bone [11].

The fibrous dysplasia with aneurysmal bone cyst of the skull can present most commonly with headache, visual

loss, cranial nerve palsies, proptosis, nausea and vomiting and scalp swellings [10, 12, 13]. Our patient presented with headache associated with proptosis and gradually progressing visual loss.

The ABCs have a classic radiological finding that are highly suggestive of these lesions. On plain X-rays, these typically have a cystic and expansile osteolytic lesion with a "soap bubble appearance." CT scan is far more superior to plain X-rays in defining the extent of involvement of the bone and shows a heterogeneous mass with bone erosion and thinned cortex with multiple air fluid levels representing the sedimentation of the red blood cells within the blood-filled cystic cavities [14]. MRI shows multiple air fluid level and hypointense peripheral rim with multiple septations [15, 16]. The definitive diagnosis of the fibrous dysplasia with aneurysmal bone cyst can only be made by the histopathology report. Fibrous dysplasia with secondary aneurysmal bone cyst typically has two components on histopathology. The fibrous dysplasia component has irregular bony trabeculae with varying number of fibroblasts, and the ABC component has blood-filled cavernous spaces surrounded by multinucleated giant cells [17, 18].

Various treatment modalities are used for the treatment of aneurysmal bone cysts including simple curettage, gross total excision, radiotherapy, cryosurgery, sclerotherapy and endovascular embolization. Out of these, the treatment of choice for fibrous dysplasia with secondary aneurysmal bone cyst is en bloc resection of the involved bony lesion with gross total excision of the soft tissue mass followed by cranioplasty. In our case also, we followed the similar treatment protocol [19–21]. Preoperative embolization can be used to decrease intraoperative blood loss by reducing the vascularity of lesion [22, 23]. Endovascular embolization can also be used in cases where the lesion is located deeply in areas difficult for surgical excision [24]. Radiotherapy has been recommended in recurrent cases, but is reported to cause malignant transformation of the benign lesion [25].

Conclusion

Fibrous dysplasia with secondary aneurysmal bone cyst of the sphenoid wing is a very rare entity. It should be kept in mind as a differential diagnosis while dealing with the osteolytic lesions of the skull. En bloc resection of the osteolytic bony lesion with gross total excision of the soft tissue component followed by cranioplasty is the ideal and the most effective treatment modality. Endovascular embolization can be an adjunct to decrease the vascularity of the lesion.

Abbreviations

CT Computed tomography
CEMRI Contrast-enhanced magnetic resonance imaging
GCS Glasgow Coma Scale
ABC Aneurysmal bone cyst

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Not applicable.

Author contributions
All authors have contributed to the manuscript. The idea of the manuscript was given by MK, and the planning of the manuscript was done by PKS. Literature search and manuscript preparation were done by KKV and PKS. Data collection was done by VKM and PNS. The final editing and correction of the manuscript were done by PKS.

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Competing interests
The authors declare no competing interests.

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