

MR Imaging in Aneurysmal Bone Cyst of the Orbit

Orbita Yerleşimli Anevrizmal Kemik Kistinin Manyetik Rezonans Görüntülemesi

ABSTRACT

Aneurysmal bone cyst is a benign fibro-osseous lesion usually described in the long bones. Intraorbital aneurysmal bone cyst is a rare clinical entity with an entirely different outcome and prognosis as compared to the usual pediatric orbital tumors. The authors report magnetic resonance imaging (MRI) features of an orbital mass lesion in a 4-year-old child who presented with painless proptosis of right eye. Characteristic MRI findings led to a suspicion of an aneurysmal bone cyst that was totally excised via a right frontotemporal craniotomy. Appreciation of characteristic MRI features can be of extreme help for appropriate diagnosis and management of this rare entity.

KEY WORDS: Aneurysmal bone cyst, Orbital, MR imaging, Radiology

ÖZ

Anevrizmal kemik kistleri genellikle uzun kemiklere yerleşen iyi huylu fibroosseos bir lezyondur. Orbita içine yerleşmiş anevrizmal kemik kisti, daha sık görülen çocukluk çağı orbita tümörlerinden tamamen farklı klinik seyir gösterirler. Yazarlar; 4 yaşındaki çocukta sağ gözde ağrısız propitoz ile bulgu veren orbital kitle lezyonunu rapor etmektedirler. MR görüntüleme bulgusu ile anevrizmal kemik kisti özelliği gösteren lezyon sağ frontotemporal kraniyotomi yapılarak çıkarılmıştır. Orbita yerleşimi nadir olarak görülen anevrizmal kemik kistinin MR bulgularının özelliklerinin tanımlanması bu lezyonun tanı ve tedavisinin planlanmasında yardımcı bir faktör olacaktır.

ANAHTAR SÖZCÜKLER: Anevrizmal kemik kisti, Orbita, MR görüntüleme, Radyoloji

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INTRODUCTION

Aneurysmal bone cyst (ABC) is a rare benign lesion of unknown etiology that most commonly occurs in the long bones and vertebrae. [4,8] ABC arising in the orbit constitutes less than 0.25% of all reported cases. Natural history of ABC is obscure. Characteristic MRI features may help to diagnose this rare lesion pre-operatively justifying aggressive attempt at total excision of this benign tumor. We describe a rare case of ABC of the orbital roof in a 4-year-old child that was suspected on MRI and treated by total excision.

CASE REPORT

A 4-year-old boy presented to our outpatient clinic with a three-month history of progressive painless proptosis of right eye. The patient had history of a fall from height 1 year back. Visual acuity was 6/36 in the right eye with 6/9 in the left on Snellen chart. Fundus examination was within normal limits. On local examination, there was non-pulsatile, non-axial proptosis involving the right eye.

Neuroimaging:

Computerized tomography (CT) of head revealed an expansile lytic lesion with thinned out cortex in the region of right lesser wing of sphenoid (Figure 1) and a bone window showing an osteolytic lesion with bony enlargement and cortical thinning (Figure 2). On magnetic resonance imaging (MRI), there was a lobulated extra-axial mass lesion iso-intense on T1WI (Figure 3A) and hyperintense on T2WI with dependent hypointensity, likely to be due to blood products (Figure 3B).

Multiple internal septations were seen which showed enhancement after injection of intravenous gadolinium (Figure 3C). The lesion extended into the orbit, displacing the globe anteriorly. Diagnosis of aneurysmal bone cyst was entertained and total excision was planned in view of the benign nature of the lesion.

Operative procedure:

A right frontotemporal craniotomy was performed. There was cystic bony enlargement involving the orbital roof and greater wing of sphenoid with thin lining of cyst filled with turbid fluid and moderately vascular, friable greyish white tumor tissue. Gross total excision of the solid cystic lesion was performed. The diagnosis was confirmed



Figure 1: Computerized tomography (CT) of the head showing an expansile lytic lesion with thinned out cortex in the region of right lesser wing of sphenoid.



Figure 2: CT bone window showing an osteolytic lesion with bony enlargement and cortical thinning with no periosteal reaction.



Figure 3: Axial T1W (3A), Axial T2W(3B) and contrast-enhanced sagittal(3C) MRI of the brain showing lobulated extraaxial orbital lesion isointense on T1WI with dependent hypointensity on T2W MRI.

on histopathological examination. The patient was discharged on postoperative day 5 but was lost to further follow up.

DISCUSSION

Aneurysmal bone cyst (ABC) is a rare benign non-neoplastic lesion that expands and destroys the bone. Orbital aneurysmal bone cyst represents less than 0.25 % cases of ABC.[4,8]

Various hypotheses have been put forth to explain the etiology of ABC. Some authors have postulated that local circulatory disturbance in the bone such as thrombosis or arteriovenous malformation leads to dilated vascular bed in the affected area with formation of ABC.[5] The relationship of preceding trauma and ABC is usually incidental, but it has been proposed that post-traumatic ABC could be a manifestation of occlusion of venous drainage of affected bone with subsequent distension and expansion of bone.[4] This may well be the most likely pathogenesis in our case and also in post-traumatic ABCs reported previously in a soccer and a hockey player.[1,9]

Plain radiographical and CT bone window findings of ABC are suggestive, yet not diagnostic. [4] MR imaging features of ABC are quite characteristic and show fluid-fluid levels which correspond to different intensities of supernatant fluid and sediment secondary to hemorrhage within the bony cavities.[5] Degradation byproducts of hemoglobin have a different relaxation time which

enable fluid levels to be seen clearly on T2W MR images.[2] MR imaging can thus be of help to diagnose ABC preoperatively and aid in prognostication as ABC is a benign non-neoplastic lesion which has an excellent prognosis following complete excision as compared to other pediatric tumors in this location, some of which can be malignant mandating a different treatment approach as compared to aneurysmal bone cyst.[3,6,10]

Management of orbital ABC includes complete surgical excision of the lesion (4). Simple curettage of the lesion is associated with a high recurrence rate of up to 50 % as reported by Purohit et al. [7] The recurrence rate can be reduced by grounding the bone substance with high speed burr.[10] Other modalities include radiotherapy, cryotherapy and experimental sclerotherapy with embolization either alone or in combination.[4] Autologous or synthetic bone grafts can be used to reconstruct the bony defect caused by surgical excision.[4]

CONCLUSION

The present case highlights the importance of MR imaging to diagnose this rare benign lesion of the orbit.

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