Aneurysmal Bone Cyst of the Orbit

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Aneurysmal bone cyst (ABC) is an uncommon benign lesion with a reported incidence rate of 0.14 cases for every 1,000,000 people. ABC can occur in any part of the skeletal system but is mainly detected in the long bones. Orbit involvement is rare (<1% of all ABCs). In this article, we presented two female patients aged 49 and 33 years old respectively who suffered from ABC. After successful surgical removal of the cyst via frontal craniotomy and simple curettage, the patients recovered well and have been in good health throughout the 2.5–4.5 years of follow-up.

A 49-year-old woman with a 5 years history of painless swelling of the right orbital rim was admitted to Fourth Hospital of Hebei Medical University in March 2009. She had no prior injury and no history of cranial tumors. Upon physical examination, her vision tested 0.12 in the right eye and 0.6 in the left eye. A 5 cm large swelling was seen around the right orbital rim. A 4 mm proptosis of the right eye was observed, but eye movement was not restricted. No retinal or optic nerve change was detected. Results of examinations on the left eye as well as of a systemic examination were within the normal range. Coronal and axial computed tomography (CT) scans showed an expansive 3.5 cm × 4.5 cm pyramidal tumor. Bone destruction was present in the superior and posterior walls of the right orbit [Figure 1a and 1b]. The average attenuation of the mass was 16.5 Hounsfield unit (HU). A cystic mass approximately 4.5 cm × 3.5 cm × 3.0 cm in size and containing hemorrhagic fluid was surgically removed on March 20, 2009. Histopathological analysis showed that the lesion was confined within a fibrous capsule, which consisted of numerous blood-filled sinusoidal spaces devoid of endothelial linings [Figure 1c]. The fibrous capsule and septations contained a number of multinucleate giant cells as well as hemosiderin-laden macrophages and cellular fibrous tissue containing some bony trabeculae. This structural composition confirmed the diagnosis of ABC. The postsurgical course was uneventful, and no evidence of recurrence was detected after 54 months of surgery.

A 33-year-old woman with a 7 years history of progressive, painless proptosis of the left eye was presented to Fourth Hospital of Hebei Medical University in May 2012. She did not have any history of injury. Systemic examination results were within normal limits. Visual acuity in both eyes was 0.8. CT of the orbit revealed an expansile lytic lesion with a thinned-out cortex and a bone window showing an osteolytic lesion with a bony enlargement and cortical thinning [Figure 1d]. A left frontal craniotomy was performed. A cystic bony enlargement involving the orbital roof was observed. The solid cystic lesion was completely removed. The diagnosis of ABC was confirmed via histopathological examination. The patient was discharged on the 7th day postoperation. No evidence of recurrence was observed after 30 months of surgery.

In 1942, ABCs were first described by Jaffeand and Lichtenstein as "peculiar blood-containing cysts of large size." ABC is a rare benign lesion with a complicated pathogenesis. Orbital ABCs have been described in <0.25% of ABC cases.[1] ABCs of the orbit have no significant sex preference because both sexes are nearly equally affected by the lesion. The ages of the patients mostly range from 10 to 30 years old, with 80% of the patients under the age of 20.[2] However, the present cases were not within the expected age range for orbital ABCs.

ABCs may be primary or secondary, and most cases of ABCs originate from lesions in the long bones and vertebrae. The exact cause of ABCs remains unclear; however, various hypotheses have been proposed to explain the etiology of ABC. The development of focal dynamic changes with secondary venous hypertension may cause a slow expansion of the cortex. Dabezies *et al*.[3] studied cases of ABC secondary to trauma and concluded that the disease represented an

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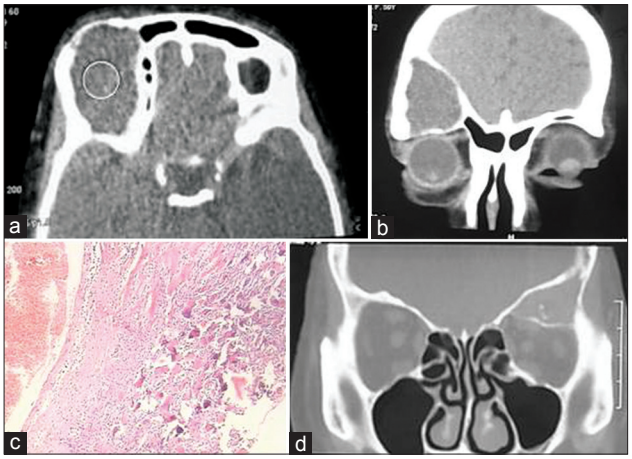

Figure 1: For the 49-year-old woman, the horizontal section (a) and coronal section (b) of computed tomography (CT) scan indicating a cystic lesion in the orbital roof; and histopathological assessment of the aneurysmal bone cyst (c) revealing numerous blood-filled sinusoidal spaces without endothelial linings and multiple giant cells within the walls of fibrous capsule (H and E, original magnification ×200). For the 33-year-old woman, CT bone window (d) showing an osteolytic lesion with bony enlargement and cortical thinning.

osseous manifestation of a posttraumatic arteriovenous fistula. A number of studies reported a history of local trauma prior to the development of ABCs; however, this association was less frequent with orbital ABCs. Our two patients had no history of trauma. The hypothesis that proposes the existence of a basic underlying arteriovenous anomaly that results in dilated vascular spaces appears to explain the course of ABCs sufficiently. Other authors proposed that genetic or chromosomal changes are involved in ABCs.[4]

CT is useful in the evaluation of ABCs. This technique shows an expansile interdiploic lesion that may be multiloculated and harbors areas with different densities. Fluid levels are observed in 35% of the cases. In the scans, the peripheral capsule and internal septations of the lesion are strongly enhanced with intravenous administration of iodinated contrast materials. However, the enhancing, heterogeneous, multilocular, fluid-containing lesion found in CT scans is not specific for ABCs. In our report, a well-defined expansile mass that caused cortical interruption was detected in the CT scans. The diagnosis of ABC can be made only by CT findings; however, an accurate histopathological evaluation is imperative to obtain a definitive diagnosis.

Although ABCs are known to occur in any part of the skeletal system, the long bones, such as the femur, tibia, humerus, pelvis, and fibula, are most frequently affected. Sheikh[5] observed that a large number of ABCs occurred in the temporal and occipital bones. Orbital ABCs have rarely been reported. ABCs occur mostly as primary lesions; only one-third of all cases involve secondary lesions. The exact cause of primary ABCs remains unclear. Meanwhile, secondary ABCs are formed from preexisting bone lesions such as angioma, nonossifying fibroma, osteoblastoma, giant cell tumors, chondroblastoma, and fibrous dysplasia. In this report, the two patients diagnosed with primary ABC did not show any preexisting bone lesions. On the basis of the clinical signs and histopathological examination results for our patients, a diagnosis of orbital ABC was highly plausible.

The ideal treatment for ABCs is total excision and repair of the bone defect in an appropriate manner. Other ABC therapy methods include radiotherapy, embolization, cryotherapy, and experimental percutaneous scierotherapy. A number of researchers has used a combination of the aforementioned techniques and had varying degrees of success. ABC lesions are very bloody; thus, preoperative angiography and embolization facilitate the surgery. A high recurrence rate of 10% − 30% has been reported. This finding underlines the need to follow-up these patients at regular intervals even after thorough curettage.

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