Neuroimaging Highlight

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Left Eye Proptosis in an 11-Year-Old Child

Submitted by: Ahmed Alkhani

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Aneurysmal bone cysts (ABC) are uncommon non-neoplastic expansile lesions of the bone. They usually affect long bones and vertebrae in childhood and early adulthood.[1-7] Skull involvement is quite rare.[1,8,9] Only a few cases of orbital involvement were described in the literatures.[10,11] We are presenting a case of ABC that affects the orbit causing proptosis and external ophthalmoplegia.

An 11-year-old girl known to have a chronic seizure disorder and school performance difficulties presented with a four month history of rapidly progressing left orbital swelling and redness. This was associated with double vision, occasional throbbing headache and feelings of nausea. The patient and her family denied any history of trauma. Her past medical and family histories were otherwise non-contributory. Physical examination revealed a left eye proptosis with mild conjunctivitis (Figure 1). She had complete left external ophthalmoplegia with pupil sparing. The left orbit was non-tender with no detectable bruit. Visual acuity in both eyes was 20/20 (right) and 20/50 (left). Fundoscopic examination revealed mild papilledema in the left eye. The rest of the physical examination was essentially normal. Computerized tomography scan of the brain revealed a left fronto-orbital expansile bony lesion with multiple cystic compartments, distorting the left orbital roof (Figure 2). The lesion pushed the eye globe laterally and externally. Magnetic resonance imaging of the left orbit confirmed that the lesion was extradural, displacing rather than infiltrating the left frontal dura, causing mass effect on the left frontal lobe. Within the lesion, the cystic components exhibited fluid-fluid levels (Figure 3 a,b,c).

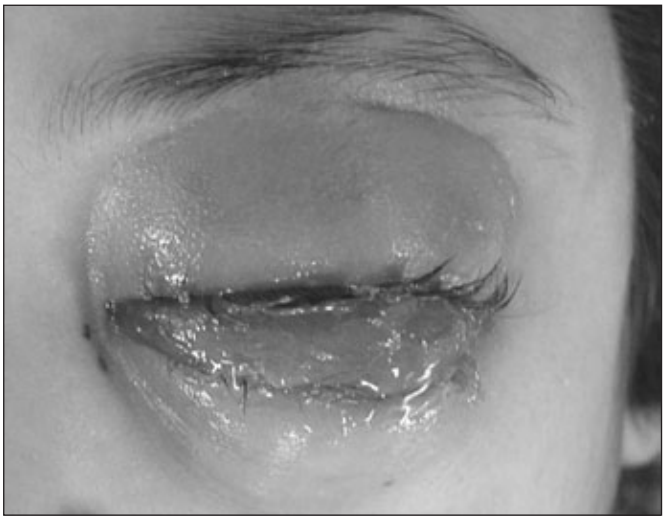

Figure 1: *Left orbital swelling and redness*

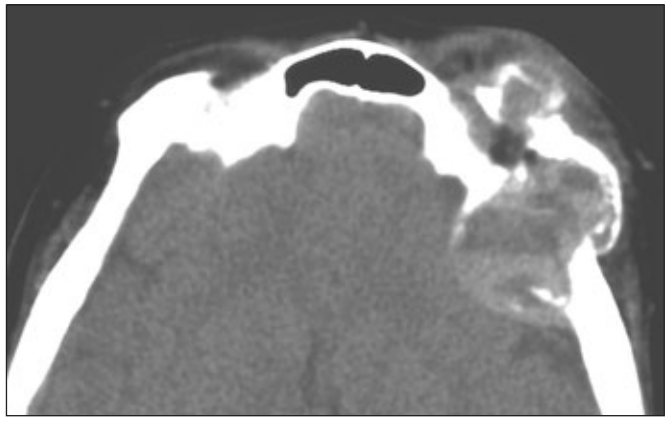

Figure 2: *Axial non-contrasted CT scan of the left orbital roof.*

From the Section of Neurosurgery, Department of Neurosciences, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia.

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Reprint requests to: Ahmed Alkhani, Division of Neurosurgery, Department of Neurosciences, MBC: 76, King Faisal Specialist Hospital and Research Centre, P.O. Box: 3354, Riyadh 11211, Saudi Arabia

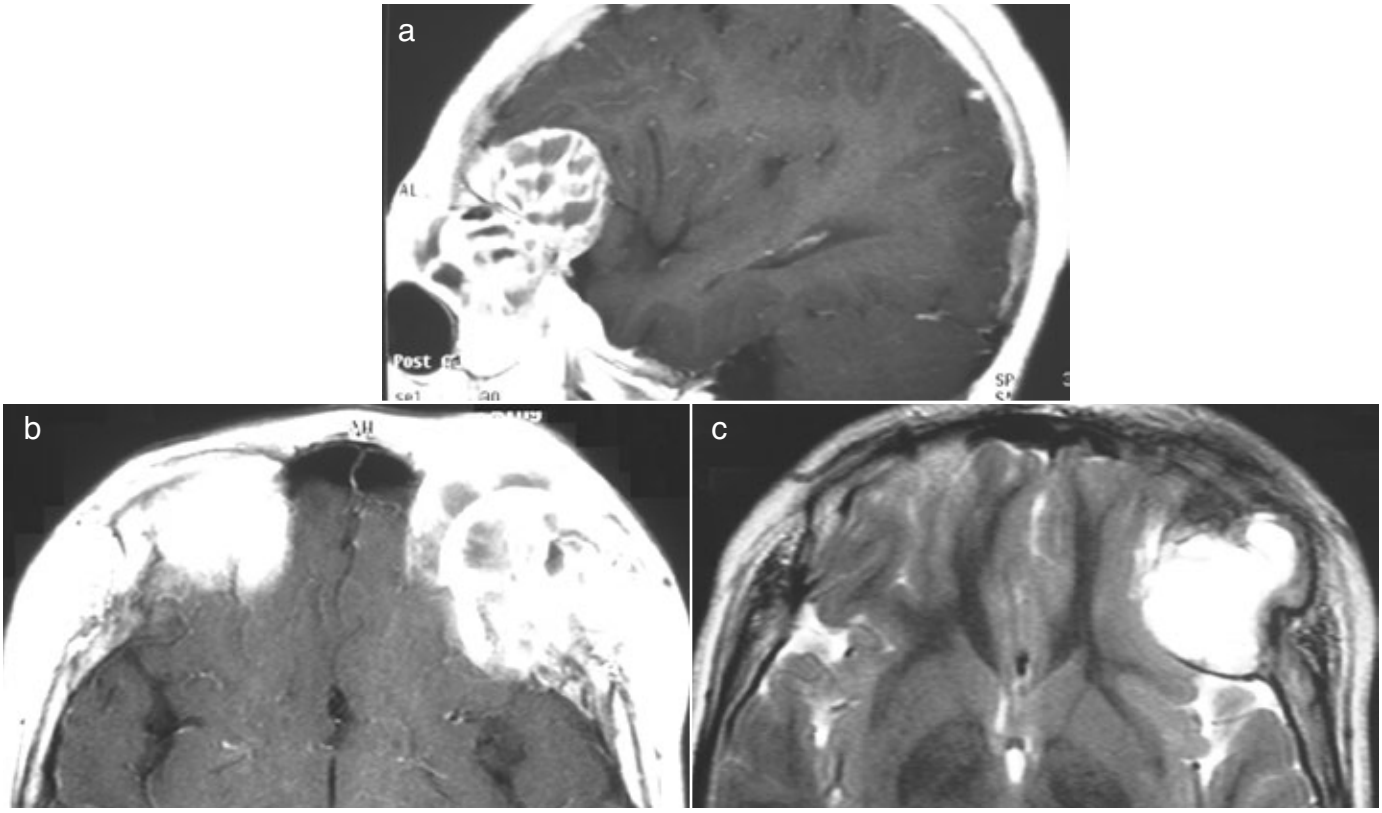

Figure 3: *T1 MRI after gadolinium injection, sagittal (a), axial (b), axial T2 MRI (c).*

At the referring hospital she was subjected to a limited left orbital incision and open biopsy. The procedure was complicated with massive intraoperative hemorrhage of more than 500cc of blood loss. The histopathological exam was non-conclusive. The patient was then referred to our institute. She was operated on for gross total resection of the lesion. The lesion involving part of the frontal bone and the roof of the orbit was completely excised surgically via a left fronto-temporal craniotomy. The lesion margins were treated with manual curettage and electrical cauterization using monopolar cautery. The bony defects resulting from the lesion removal were corrected using autologous bone graft supported with titanium mesh and screws. The patient had an uneventful intra-operative course. No postoperative complications were encountered. The histo-pathological studies confirmed the diagnosis of aneurysmal bone cyst. No other coexisting pathological tissue was identified. The patient had excellent results from her surgery. She was followed for more than three years in the outpatient clinic with no evidence of recurrence (Figure 4).

Aneurysmal bone cysts are uncommon, rapidly growing, destructive, expansile non-neoplastic lesions. They usually occur in long bones and vertebrae.[1-5] They rarely occur in the skull.[8,9] Orbital involvement has been reported in a few case reports in the literatures.[10,11] Other skull locations include temporal and occipital bones and skull base.[8,12-15] Aneurysmal bone cysts usually develop during childhood or early adulthood.[1,6,16] No sex predilection was suggested in the literatures.[1,7,16]

Aneurysmal bone cysts may develop *de novo* or in conjunction with other coexisting tumors (secondary) such as giant cell tumor, chondroblastoma, non-ossifying fibroma, osteoblastoma, fibrous histocytoma, osteosarcoma and fibrous dysplasia.[1,3,7,16-19] Histological examination of ABC reveals lesions composed of large separated sinusoidal cavities, frequently filled with blood and lined by endothelium and multinucleated giant cells. Hemorrhages within these cystic cavities are of variable ages. Normal bony trabeculae are usually seen at the periphery of the lesion.[10-12,15,17,20]

Radiologically, ABCs are described as well-defined osteolytic and destructive lesions. Typically, they show expanded, remodeled and ballooned contour of the bone. The fluid-fluid levels frequently encountered are secondary to hemorrhagic components. Computerized tomography scan with bone window can illustrate the expansile multi-cystic features of the lesion. Magnetic resonance image is extremely helpful to rule out dural involvement. Aneurysmal bone cyst on MRI usually exhibit as multi-cystic cavities with different intensity and fluid-fluid levels within these cystic cavities. These radiological findings are almost diagnostic but not pathognomonic of ABC.[1-3,7,16,17] Aneurysmal bone cysts secondary to other possible underlying lesions cannot be ruled out radiologically.

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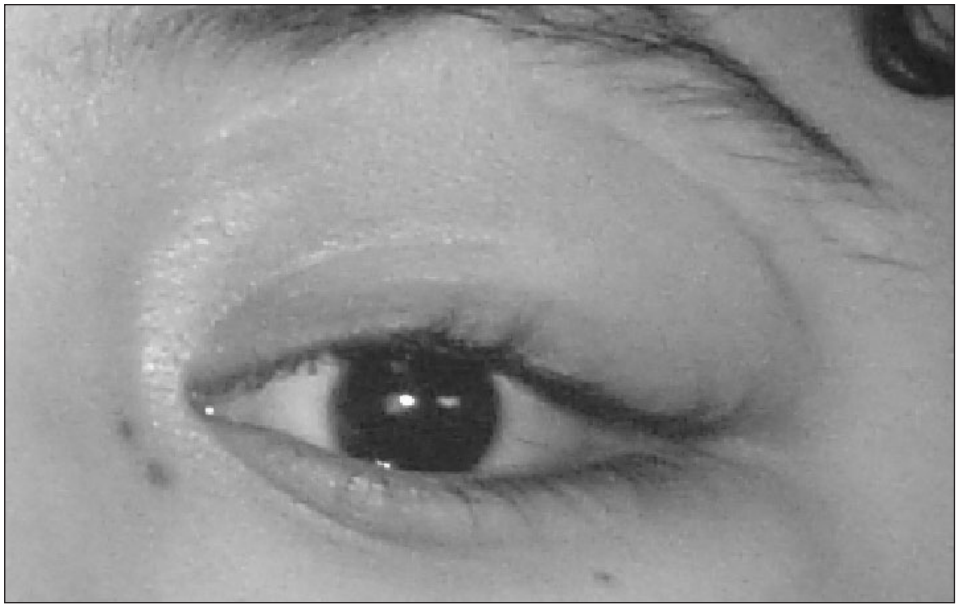

Figure 4: *Left orbit, six months post operatively.*

Total surgical resection, when feasible, is a curative procedure; recurrence is not usual after total surgical resection. Only simple curettage is associated with a high incidence of recurrence.[1,3-6] Radiotherapy for unresectable or residual lesion is of unproven value. In addition it may carry the risk of possible malignant transformation of pre-existing lesions such as fibrous dysplasia.[5,6,18,19] In many cases cosmetic repair of the bony defect is needed. Limited biopsy is not recommended, as it may not result in adequate diagnostic material and carries the risk of extensive intra-operative bleeding.

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