



Case Report

Aneurysmal Bone Cyst (ABC) Involving Petrous Apex Treated by Microsurgical Removal: A Case Report

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Abstract

Aneurysmal bone cyst (ABC) is a benign progressive expansile bone lesion that usually involves often located in vertebrae, long tubular bones and flat bones. A small percentage of aneurysmal bone cysts arise from the skull base. Skull base involvement is very rare. Here, we describe a 22-year young man presented with diplopia, right sided facial paresis and right sided sensori-neural hearing loss. CT scan and MRI showed a right petrous apex skull base mass that was confirmed as ABC in histopathology after surgical removal. [*Journal of Science Foundation, January 2020;18(1):25-28*]

Keywords: Aneurysmal bone cysts; aneurysmal; petrous apex; skull base

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Introduction

Aneurysmal bone cysts (ABC) are benign, rapidly expansile, osteolytic lesions with well-known histology but obscure pathophysiology (Aghaghazvini et al., 2012; Gürsoy et al., 2015; Jaffe and Lichtenstein 1942). They are more commonly involved in posterior elements of vertebrae, long bones metaphysis & flat bones and are usually occur in the first two decades of life (De Silva et al., 2003; Gürsoy et al., 2015). ABC arising from the skull base is a rare condition with an incidence of 3.0 to 6.0% cases of all ABC cases and ABC located in the petrous apex is extremely rare (Lackmann and Tollner 1993). We had found a case of ABC located in the petrous apex of the temporal bone in a young man of 22 presented with 6th, 7th & 8th nerve dysfunction with dry eye.

Case Presentation

A young man of 22 presented with slowly progressing history of diplopia on looking toward right, facial asymmetry, dry right eye and right sided progressive hearing loss. There was no history of otitis media, trauma or surgery. On physical examination, he had right-sided sensorineural hearing loss, right sided lower

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motor type of facial paralysis (B & H grade-4) and right sided abducent nerve palsy. The remainder of nervous system including lower cranial nerves, head and neck examination and other systems were normal. On CT scan, an osteolytic expansile mass was detected on the right sided petrous apex (Figure I).

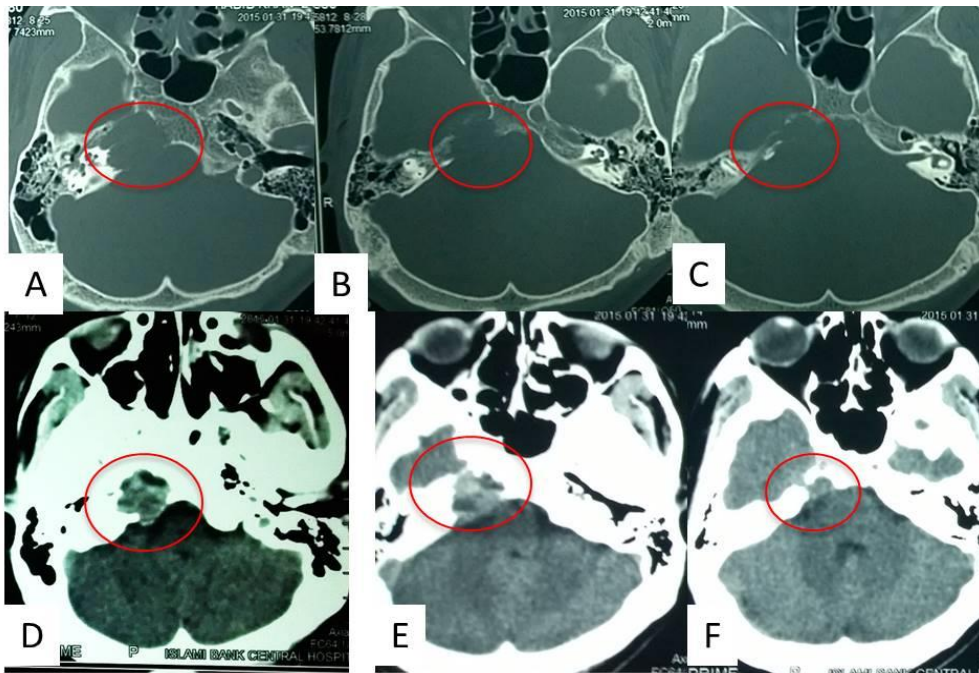


Figure I: CT scan of head axial images. A,B&C(bony window) showing destructive, expansile bony lesion in right sided petrous apex. D, E &F- mixed density bone destructive lesion in right petrous apex

On MRI, an iso intense lobulated well-defined mass in T1W sequence containing small high signal foci that was heterogeneous and iso to high signal in T2W (Figure II).

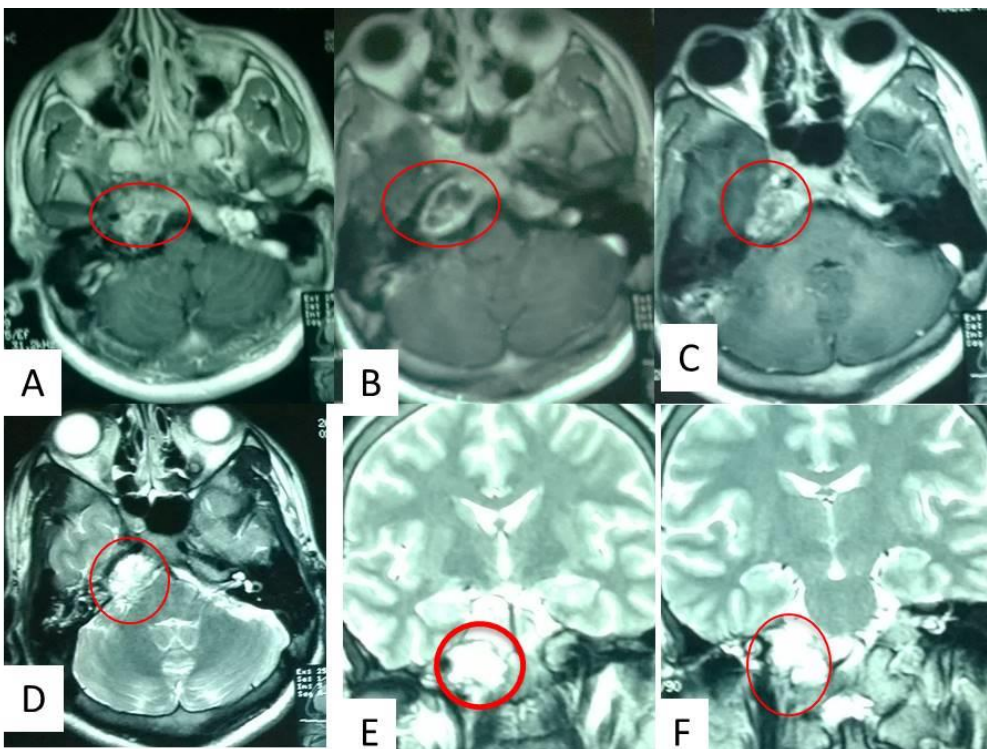


Figure II: MRI of head; A,B&C-contrast axial images showing irregularly contrast enhancing lesion in right petrous apex. D- axial, E&F coronal T2W images showing hyper intense bony lesion in right petrous apex (ABC)

There was also peripheral contrast enhancement. Through right sided temporal craniotomy petrous apex was reached extradurally. Then tumor was identified and removed completely by micro-drill and curated with preservation of facial and greater petrosal nerve. Tumor involved bony cochlea and labyrinthine part of facial canal. After resection, histopathology reported (multiple blood filled cystic cavities separated by thin fibrous septa and many multinucleated giant cells) ABC of petrous apex (Figure III). Post operatively his right sided 6th, 7th and 8th nerve functions were as preoperative status without improvement or deterioration. There was no recurrence till last follow up (21 months after operation).

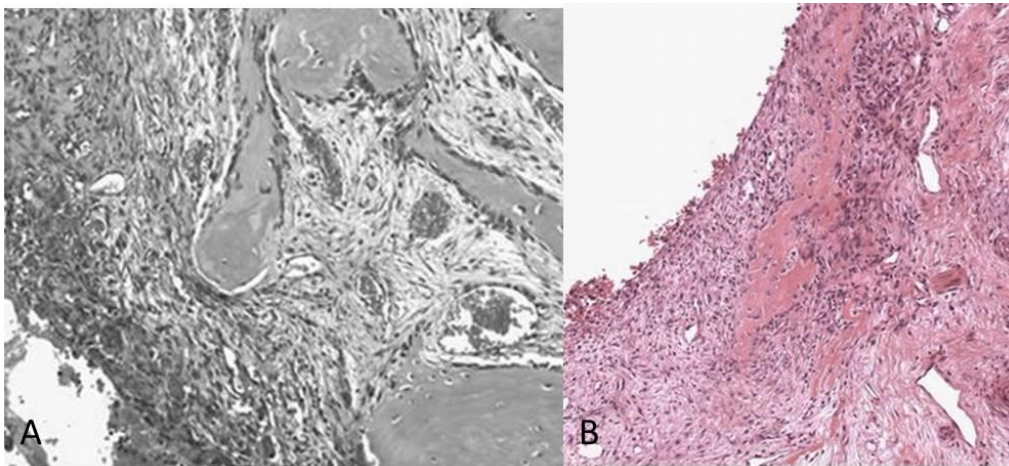


Figure III: Microphotograph (A&B) of Histopathological Slides of the Resected Specimen of Petrous Apex Tumor (Aneurysmal Bone Cyst)

Discussion

ABCs are benign, but locally aggressive bone tumors, consisting of thin-walled blood-filled cysts lined by connective tissue with giant cells and trabecular bone. They are typically found eccentrically in long bone metaphyses and posterior elements of vertebrae (Lackmann and Tollner 1993). ABCs are usually encountered in the first two decades of life (De Silva et al., 2003; Lackmann and Tollner 1993). However, age of our case was a little older. Temporal bone ABCs most commonly present as swelling in the temporal region (Tuna et al., 2003) and ABC located in the petrous portion of temporal bone is extremely rare. Symptoms of temporal ABCs are temporal swelling, hearing loss, 3rd, 6th and 7th cranial nerve paralysis, headache, decreased vision, ptosis and even seizure, intracranial hemorrhage and recurrent meningitis have been reported (Vergel de Dios et al., 1992; Connor et al., 2008). In our case clinical features were 6th cranial nerve palsy, diplopia, hearing loss, facial palsy and dry eye. The portion of the abducent nerve passing through Dorello's canal at the petrous apex was probably affected by the tumor. Other symptoms were due to facial nerve and cochlear involvement by the tumor.

Computed tomography shows that the lesion is intra-diploic with expansion of the bone and thinning of bony tables. MRI is the method of choice in the diagnosis showing an expansive, well defined multiloculated cystic mass with hypointense peripheral rim and internal septations. Fluid-fluid levels are characteristic but not specific findings for ABCs, which can also be seen in other lesions like giant cell tumor, osteoblastoma, telangiectatic osteosarcoma, chondroblastoma, solitary bone cyst and fibrous dysplasia (Tsai et al., 1990). The cysts have heterogenous signal due to blood products of varying ages. Heterogenous internal enhancement is noted on post-contrast scans. MRI findings were typical for ABC in our case. In MRI intensely enhancing tissue that is of low signal intensity on T2W images surrounding cystic spaces have been described as fibrous elements, which are thought to be more abundant in the more cellular or solid variant of ABCs (Buxi et al., 2004). Treatment of ABCs are complete surgical excision which usually results in cure (Gürsoy et al., 2015).

Recurrence of ABC of the calvaria is rare, whereas recurrence of ABC of other bones may be as high as 50.0% cases (Tsai et al., 1990; Buxi et al., 2004)). Recurrence has been related to younger age, larger tumor, presence of mitosis, incomplete surgical removal and dural involvement. The success of surgery is strongly

associated with the rate of recurrence; while total excision is curative in most cases, simple curettage and subtotal excision may exhibit high recurrence rates varying from 20% to 50% cases (Sayama and MacDonald 2010). ABC is either primary or secondary to a preexisting lesion like giant cell tumor, osteoblastoma, chondroblastoma, non-ossifying fibroma, angioma and fibrous dysplasia. Presence of a preexisting lesion has been reported to increase the likelihood of recurrence (Tsai et al., 1990; Gürsoy et al., 2015). Radiotherapy has not been used as a treatment option in the past because of risk of malignant transformation. It is now advocated for cases that are not suitable for surgery, and cases with recurrent or residual tumor (Sayama and MacDonald 2010).

Conclusion

Petrous apex ABC, although rare, should be considered in the differential diagnosis of a petrous apex mass lesion especially in first two decades of life isolated or coexistent sixth nerve palsy with the presence of typical MRI findings.

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