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EC NEUROLOGY
Case Report

Aneurysmal Bone Cyst of the Occipital Bone

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Abstract

ABC is benign osteolytic lesion that is locally destructive. Calverial ABC is very uncommon. Among them occipital ABC is exceedingly rare. We describe a case of occipital ABC in a young girl presenting with headache and gait disturbances. We will also review the literature regarding its origin, clinical features, imaging characteristics, operative findings and treatment options.

Keywords: *Aneurysmal Bone Cyst; Occipital Bone; Posterior Fossa*

Introduction

Aneurysmal bone cyst (ABC) is benign, vascular, osteolytic lesion occurring commonly in metaphysis of long bone and spine in 50% and 20% cases respectively. ABC Of skull bone is very rare and represents only 3 - 6% of case. Most of the patients are female under age of < 20 years [1-4] very few cases of the occipital ABC has been described in literature. We will describe a case of occipital ABC in a 19 years newly married female presenting as severe throbbing headache in the back side of head. We will also focus on imaging features, treatment and intraoperative findings of this rare lesions.

Case Report

A 19 years female came to our hospital with the complain of occipital headache since 1 year which was gradually increasing in intensity since a month. She also complained of occasional vertigo, neck pain and tendency to fall on left side while walking. Examination of the patient revealed there were no obvious swelling over occipital region but there was tenderness on palpation. Tandem walking was not possible and there was tendency to fall on left side. Rest of the cerebellar and neurological examinations were normal. We did CT scan and MRI of brain with contrast including all investigations needed for undergoing surgery. CT scan revealed a contrast enhancing mass of size 52 x 35 x 16 mm located in posterior fossa, eroding occipital bone and compressing 4 the ventricle (Figure 1). The lesion was multiloculated expanding occipital squama in both intracranially and extracranially. Both inner and outer tables of skull were destroyed. There were multiple fluid levels seen. MRI of the brain with contrast revealed multiloculated and multilobulated extra-axial extradural lesion in midline posterior fossa protruding mostly inside the skull (Figure 2). There were multiple loculations with fluid- fluid and blood- fluid levels and multiple septae were separating the cystic cavity.it was hypointense on T1WI with few T1 iso and hyperintensities, hyperintense on T2WI,and areas of blooming with blood fluid level on GRE images. There Were peripheral wall and septal enhancement on post contrast images. There was no perilesional edema but there was mass effect over adjacent bilateral cerebellar hemisphere and brainstem with compression of 4th ventricles.

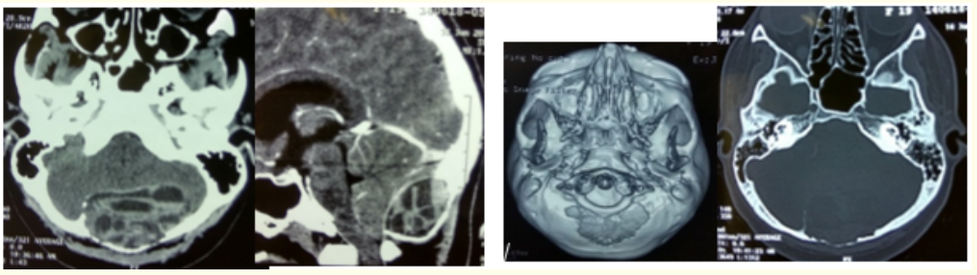

Figure 1: *Shows CT SCAN of brain showing aneurysmal bole cyst in occipital region with soap bubble appearance and bony destruction.*

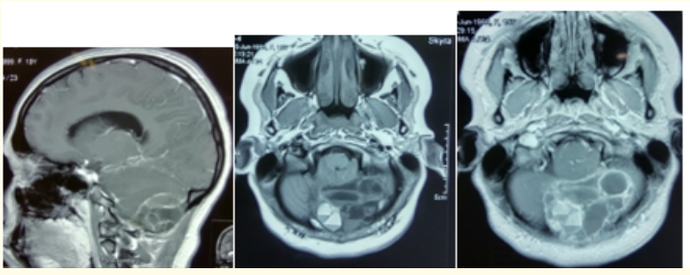

Figure 2: *Showing MRI with contrast image having multiple soap bubble within lesion. The main mass of lesion lies with posterior fossa.*

Because of the bloody nature of the ABC we arranged 2 unit of blood and patient was taken to operating room. We did suboccipital craniectomy. The craniectomy margin was extended upto 1 cm of the normal bone (Figure 3). The outer and inner tables of the bone were partially eroded. The dura was tightly adherent to the lesion which were carefully separated from the bone. The inside mass was dark, spongy and hemorrhagic. Microscopically sinusoids like areas filled with uncoagulated blood were seen. Continuous oozing of blood was noted intraoperatively till the whole of the lesions were removed. The involved posterior margin of foramen magnum was also removed. Hemostasis was secured and cranioplasty was done with titanium mess. Wound was closed in layers without putting the drains. The post-operative course was uneventful and the patient was discharged without having any complain. Postoperative image showed no trace of tumor with complete removal (Figure 4).

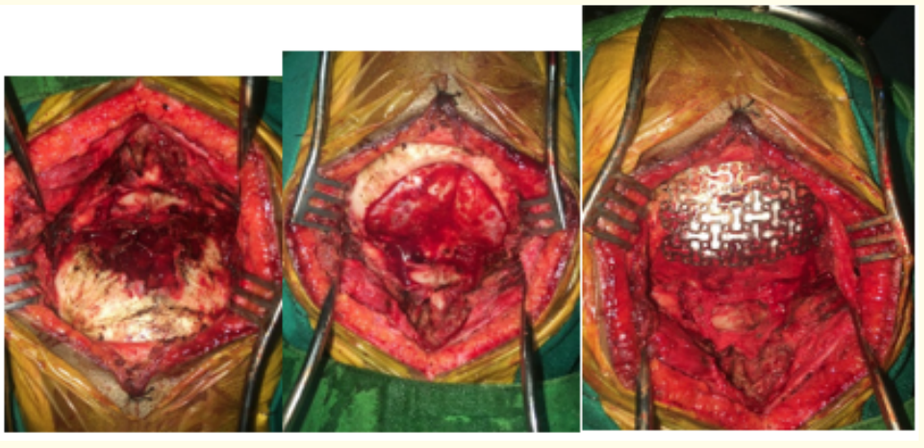

Figure 3: *Shows intraoperative picture having destruction of bone by ABC, our areas of craniectomy and cranioplasty by titanium mess plate.*

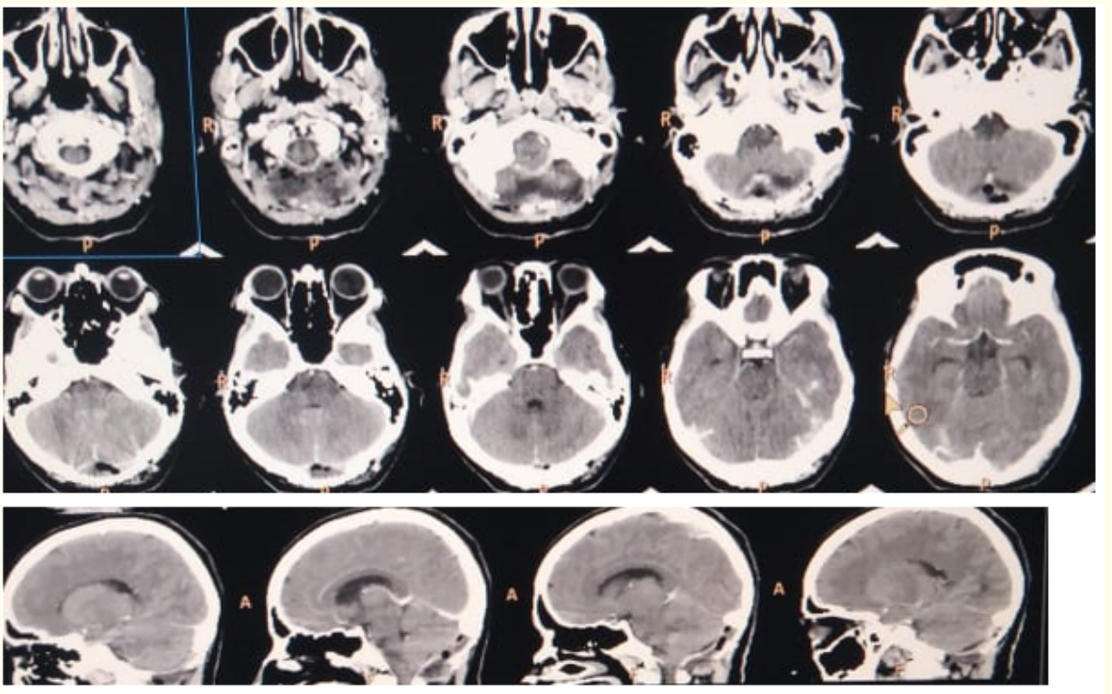

Figure 4: *Postoperative CT scan shows complete removal of tumor*

Histopathology revealed cavernous vascular spaces filled with blood and separated by fibrous septae. The thick fibrous septa are lined by flattened fibroblastic cells and osteoclast like giant cells. Focal areas show reactive new bone formations (Figure 5).

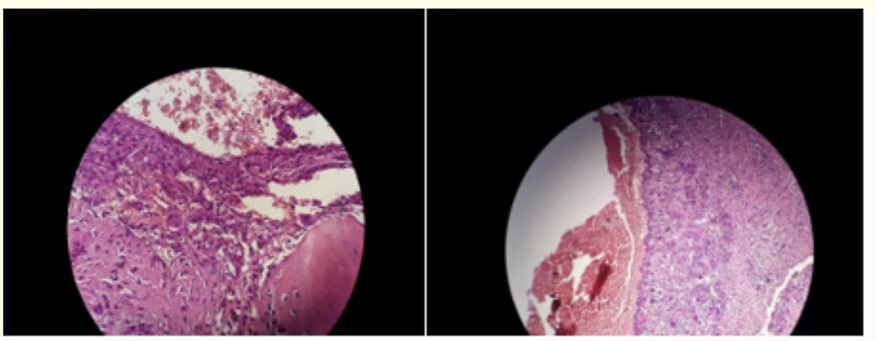

Figure 5: *Histopathology shows cavernous vascular spaces filled with blood and separated by fibrous septae. The thick fibrous septa are lined by flattened fibroblastic cells and osteoclast like giant cells. Focal areas show reactive new bone formations.*

Discussion

ABC is a benign and expansible osteolytic lesions occurring mostly before the age of 20 years [1,5,6]. Calverial ABC are very rare location and only 20 cases have been described in this region (Table 1 and table 2). Of those only 13 were pure occipital ABC (Table 1). Some of the cases have also co-existing lesions like fibrous dysplasia, eosinophilic granuloma, and osteoblastoma [4,8] (Table 2). Since 2000 (in span of 18 years) only 13 cases of ABC has been described so far which shows the rarity of our case. Several etiology has been described for the origin of ABC including post traumatic cause. other suggests that it results from underlying neoplasm where local and persistent alteration in hemodynamics occurs leading to increased venous pressure and development of dilated and engorged vascular bed within the affected boney area [3,7,8].

Serial No/Authors/Years	Age/sex	Clinical presentation
1. Lucarelli., *et a*l. 1980	19 f	Headache and palpable mass
2. Bilge., *et al*. 1983	18 m	Palpable mass
3. Bilge., *et al*. 1983	3 f	Palpable mass
4. David., *et al*. 1993	21 m	Focal tenderness
5. Braun., *et al*. 1987	4.5 f	Palpable mass
6. Arthur., *et al*. 1988	9 f	Palpable mass
7. Chateil., *et al*. 1997	9 m	Headache and vomiting
8. Petro and Lancon 2001	7 f	headache
9. Gan and Hockley 2007	8 m	exophthalmos
10. Lin., *et al*. 2007	54 f	Focal tenderness
11. Genizi., *et al*. 2011	2 yrs m	headache
12. Garber and Cambrin 2015 [3]	3 yrs f	Rupture cyst due to head injury
13. Kalina and Wetjen 2015 [8]	9 yrs m	Headache and focal tenderness
14. Chowdhury and Chaurasia 2018 (current case)	19 yrs f	Headache

Table 1: *Some of the previously published (only)occipital aneurysmal bone cyst [4].*

M: Male; F: Female.

Author	Age/sex	Associated pathology	Clinical presentation	Location
1. Roncaroli., *et al*. 2001	2 m	Eosinophilic granuloma	Focal tenderness	O
2. Itshayek., *et al*. 2002	19 m	Fibrous dysplasia	Focal tenderness	O
3. Iseri., *et al*. 2005	35 f	Fibrous dysplasia	headache	T+O
4. Mattei., *et al*. 2005	19 f	Fibrous dysplasia	headache	P+O
5. Han., *et al*. 2008	20 m	Osteoblastoma	headache	O
6. Lee., *et al*. 2010 [5]	40 m	-	proptosis	Petrous bone
7 . Valsangkar., *et al*. 2016 [1]	22 f	-	Headache, gait disturbance	Middle and posterior fossa

Table 2: *Some of the previously published occipital aneurysmal bone cyst with associated pathology [4].*

O: Occipital; p: Parietal; T: Temporal.

Most of the presenting complains are headache localized to the lesion, tenderness and palpable mass. Others may be due to traumatic rupture of cyst, focal compression by cyst, cerebellar symptoms and raised intracranial pressure depending on the locations [1,4,8].

CT is usually diagnostic which shows soap bubble appearance and multi lobulated multi cystic mass having thinning of boney cortex. It may show hemorrhage within the mass and blood fluid levels. MRI typically shows well demarcated mass having enhancing septations separating cystic lesions with fluid levels [3-5,8,9].

Total removal of the lesion which could be curative is the treatment of choice whenever possible as malignant transformation and recurrence has been observed in some cases. The case that are not amenable to gross total resection can be managed by partial resection, intra lesional curettage, with adjuvant therapy, Including preoperative embolization, postoperative radiotherapy and cryotherapy [1,4-6,8]. For resistant lesion and when surgical decompression is hazardous percutaneous sclerotherapy can be another option. We did gross total removal of our tumor without having any additional adjuvant therapy.

Conclusions

Occipital ABC is very rare lesions and can present with gait disturbance like our case. It can be managed surgically by gross total removal of tumor which could be curative. Early diagnosis and prompt treatment favors good outcome.

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