Aneurysmal Bone Cyst of Skull Base: a case report and review of literature

Yu-Chang Lee1 Jhi-Tsun Ho2 Wei-Che Lin3 Chun Chung Lui3

Department of Diagnostic Radiology1, E-Da Hospital / I-Shou University
Department of Neurosurgery2, Department of Diagnostic Radiology3,
Chang Gung Memorial Hospital - Kaohsiung Medical Center, Chang Gung University College of Medicine

Aneurysmal bone cyst (ABC) infrequently involves the calvarium and is even more unusual at the skull base. The sporadic reported cases of ABC occur primarily in the pediatric or adolescent group. We hereby present a huge aneurysmal bone cyst at the skull base in a 40 year-old male presenting with proptosis, decreased visual acuity and visual field with review of skull base aneurysmal bone cyst from the recent English literature.

CASE REPORT

A 40 year-old male presented to the ophthalmologist with proptosis of the right eye and decreased visual acuity. The patient has been well until he felt a gradually enlarging lump near the right temporomandibular joint since two years ago. On physical examination, tenderness and limitation of the range of motion at the right temporomandibular joint was noted; decreased visual acuity and narrowing of visual field of the right eye were noted. All the routine laboratory data were within the normal range. A computed tomography was performed which showed an expansile osteolytic tumor-like mass with the mass predominantly located at the right petrous bone. The mass had a bubbly appearance composed of numerous small hypodense cysts with septa (Fig. 1). The solid component, peripheral portion and septa enhanced after contrast administration. The mass resulted in thinning of temporal bone laterally and remodeling of lateral orbital rim medially. On MR imaging, the lesion exhibited multiple hyperintense fluid-fluid levels on T2WI with hypointensity at the dependant parts due to hemorrhage (Fig. 2a). The bulk of tumor-like mass was in the middle cranial fossa abutting the cavernous sinus medially and with inferior extension...
to the infratemporal fossa (Fig. 2b). Cerebral angiographic study demonstrated faint hypervascular blush over the right temporal region with blood supplies from internal maxillary, middle meningeal, posterior auricular and ascending pharyngeal branches on the selective ECA injection (Fig. 3). A right frontotemporal craniotomy with extirpation of the tumor was attempted by the neurosurgeon. At surgery, hemorrhagic multi-cystic lesion measuring up 10cm x 6cm x 6cm was discovered at right middle cranial fossa with erosion of the adjacent cortices of right sphenoid and maxillary sinuses. Yellowish fluid gushed out from the cystic components of the mass. The microscopic section showed blood clots and tissue fragments with diffuse infiltrates of heavily pigmented spindles, plumps of polygonal cells admixed with foamy histiocytes and scattered multinucleated giant cells in fibrotic stroma, consistent with aneurysmal bone cyst.

**DISCUSSION**

Aneurysmal bone cyst is a benign, non-neoplastic lesion, occurs most commonly before the age of twenty [1]. ABC usually involves the metaphysis of the long bones (60%); the vertebrae (20%). Only a few sporadic cases have been reported to have the lesion at the skull base [2]. After reviewing the recent English literature and excluding those ABC lesions originating from the calvarium, we have found that eleven cases of skull base ABCs (aged 1 to 19 years old, 6 males, 5 females) have been reported, with one report each associated with concomitant giant cell tumor, osteoblastoma or fibrous dysplasia (Table 1). Of the eleven cases [1-9], six tumors were found in the anterior cranial fossa with primary involvement of the frontal base or sphenoid bones; two tumors were found in the posterior cranial fossa with involvement of the clivus and occipital bones; the other three tumors were in the middle cranial fossa with erosion either to the squamous, petrous or mastoid portion of temporal bone, a similar location to the demonstrated case. The age, sex, location, presentation and image characteristics of these cases were listed in Table 1.

The pathogenesis of ABC may be linked to the development of local hemodynamic alteration, local trauma, or a secondary phenomenon superimposed

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**Figure 1.** Contrast enhanced CT reveals **a.** well defined thick-walled mass at right middle cranial fossa with enhancement over the peripheral margin and internal septa, and with remodeling and erosion of the greater sphenoid wing and the temporal calvarium. **b.** extracranial extension to the infratemporal fossa, with irregular enhancement of the solid component, expansion and erosion of the sphenoid sinus and displacement of posterior maxillary sinus wall.
Table 1. Reported cases of skull base aneurysmal bone cyst

<table>
<thead>
<tr>
<th>Author/Year</th>
<th>Age/sex</th>
<th>Location</th>
<th>Presentation</th>
<th>Other pathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sawin et al., 1995</td>
<td>10/F</td>
<td>Petrous and squamous portion of temporal bones</td>
<td>Progressive hearing loss and headache</td>
<td>None</td>
</tr>
<tr>
<td>Haddad et al., 1998</td>
<td>6/M</td>
<td>Squamous temporal bone and floor of middle cranial fossa</td>
<td>Slowly progressive non-tender mass</td>
<td>Fibrous dysplasia</td>
</tr>
<tr>
<td>Sheikh et al., 1999</td>
<td>6/M</td>
<td>Middle cranial fossa</td>
<td>Slowly progressive non-tender mass</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>1/M</td>
<td>Floor of anterior cranial fossa with erosion to orbital and nasal cavity</td>
<td>Mouth breather</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>10/F</td>
<td>Occipital and suboccipital</td>
<td>Slowly enlarged retroauricular mass</td>
<td>None</td>
</tr>
<tr>
<td>De Minteguiga et al., 2001</td>
<td>14/F</td>
<td>Sphenoid bone</td>
<td>Frontal headache, bouts of nausea and vomiting</td>
<td>None</td>
</tr>
<tr>
<td>Wang et al., 2001</td>
<td>16/F</td>
<td>Parasellar, subfrontal, sphenoid and ethmoid sinuses</td>
<td>Blurred vision and then loss of vision</td>
<td>Osteoblastoma</td>
</tr>
<tr>
<td>Nakdami et al., 2001</td>
<td>19/M</td>
<td>Anterior cranial fossa mass with extension to paranasal sinuses and nasal cavity</td>
<td>Slowly enlarging nontender mass over midface, loss of olfactory and visual acuity</td>
<td>None</td>
</tr>
<tr>
<td>Cansiz et al., 2002</td>
<td>17/M</td>
<td>Sphenoid sinus, ethmoid sinus, superior nasal sinus and clivus</td>
<td>Headache, diplopia, loss of vision</td>
<td>None</td>
</tr>
<tr>
<td>Ito et al., 2003</td>
<td>20/M</td>
<td>Clivus, petrous and occipital bones</td>
<td>Headache, nausea, vomiting</td>
<td>Giant-cell tumor</td>
</tr>
<tr>
<td>Theron et al., 2006</td>
<td>14/F</td>
<td>Anterior cranial fossa with extension to ethmoid and clivus.</td>
<td>Proptosis, headache</td>
<td>None</td>
</tr>
</tbody>
</table>

**Figure 2.** Axial T2WI MRI a. reveals multiple small septated cysts with fluid-fluid level, characteristic of aneurysmal bone cyst and the sediment corresponds to hemorrhage. Coronal T2WI MRI b. shows the mass located in the middle cranial fossa abutting on the cavernous sinus and extending inferiorly to infratemporal fossa.
on an existing pathological process of the bone as demonstrated by the variety of tumors and processes from the reviewed literatures [2, 7]. 30% of aneurysmal bone cysts are accompanied by other bony pathology and may be regarded as reactive phenomenon. The associated bony pathology included fibrous dysplasia, chondroblastoma, osteoblastoma, nonossifying fibroma, and giant cell tumors. The reviewed series showed that three of eleven cases, or 27%, of aneurysmal bone cyst arose from the skull base region associated with other bony pathologies.

Diagnosis of aneurysmal bone cyst usually poses no challenges preoperatively. CT often exhibits soap bubbly appearance for the multi-cystic mass and demarcates thinning of bony cortex with infrequent new bone formation around the lesions. On MRI, these lesions manifest with fluid-fluid levels and peripheral rim enhancement which are rather characteristic of aneurysmal bone cyst but not pathognomonic. Other lesions such as telangiectatic osteosarcoma, giant cell granuloma, and benign fibro-osseous lesions also have been reported to have similar fluid-fluid level appearance on the MR images [10-12]. Angiography study may reveal feeding artery with tumor blush and preoperative embolization could be attempted to reduce intraoperative bleeding. The imaging findings of the present case harbor most ingredients for the diagnosis of aneurysmal bone cyst, although it is rare to find ABC at this age and at this atypical location. When ABC on skull base is secondary to other bony pathologies, the imaging studies usually reveal the characteristic findings of the related pathologies such as ground glass lesions in fibrous dysplasia in addition to fluid-fluid levels to suggest underlying ABC. These lesions may exhibit larger proportion of solid component as in the cases of ABC associated with osteoblastoma or giant cell tumor in these reviewed cases.

The variegated symptoms caused by skull base ABC are related to the anatomical involvement of the skull base structures, as would be expected from the other more common skull base tumors. Of the eleven reviewed cases and the demonstrated case, six tumors originating in the anterior cranial fossa result in symptoms varying from slowly growing facial mass to headache, stuffed nose, blurred vision and loss of vision acuity or smelling. In four tumors including the present one, the masses located primarily in the middle cranial fossa presented with symptoms of slowly enlarging non-tender mass or hearing loss. In the last two tumors, the masses were situated in the posterior cranial fossa, presenting with headache, nausea or vomiting. The late onset of symptoms coincides with the insidious growth nature of the aneurysmal bone cyst.

Treatment of the skull base aneurysmal bone cyst is by surgery, ranging from enucleation to complete excision when possible, or partial resection when total removal cannot be accomplished. Preoperative embolization may reduce the tumor size and decrease intraoperative bleeding. Since aneurysmal bone cyst is a benign lesion, there is no role for radiotherapy or chemotherapy as these modalities may induce sarcoma in the head and neck regions.

Skull base aneurysmal bone cyst, although rare, can be found at the skull base occasionally with or without other associated bony pathologies. The signs and symptoms are insidious because of the slow growth nature. The imaging studies reveal characteristic diagnostic findings such as fluid-fluid levels regardless there is other associated bony pathology.

REFERENCE

顱底動脈瘤性骨囊腫：病例報告和文獻回顧

李懿蒔¹  何治軍²  林偉哲³  吕鎮中³

財團法人義大醫院/義守大學 放射診斷科¹
長庚大學 醫學院 高雄長庚紀念醫院 神經外科²  放射診斷科³

顱底動脈瘤性骨囊腫並不常見，而長在顱底的動脈瘤性骨囊腫更罕見，回顧以往的英文文獻只有一些零星記載，通常發現於小孩或青少族群。我們在此報告一位 40 歲男性罕見病例，因為長在顱底的動脈瘤性骨囊腫，造成視野受損。我們也同時回顧文獻上的病例和做簡短的討論。