Giant Cell Tumor of the Occipital Bone and Secondary Aneurysmal Bone Cyst: Case Report and Review of Literature

R Modkovski, R Elliott, B Rubin, D Zagzag, J Jafar, I Mikolaenko

Citation

Abstract
Giant cell tumors (GCTs) and aneurysmal bone cysts (ABCs) are locally aggressive but benign lesions typically of the long bones of the extremities or spine and rarely involve the cranial vault or skull base. Although they are distinct entities pathologically and cytogenetically, GCTs can occur with secondary ABCs, which are likely reactive in nature. The reported cases have primarily involved bones of the extremities and, to our knowledge, there has been only one case of coexisting GCT and ABC of the calvarium or skull base. We describe a 27-year old woman who underwent complete resection of a GCT with secondary ABC of the left occipital bone and condyle. We discuss the significance of the co-occurrence of these lesions and their optimal management.

INTRODUCTION
Aneurysmal bone cyst (ABC) and giant cell tumor of the bone (GCT) are both benign osseous lesions that were originally described in the literature in 1942 and 1818, respectively. GCTs are a common cause of secondary ABC and are usually found in the bones of the extremities. Their associated presentation in the skull base or calvarium is rare, and, to our knowledge, has been reported only once. We describe the case of a 27-year old woman who presented with pain and swelling in the left posterior auricular region. She was found to have a large tumor of the occipital bone and underwent complete surgical resection. The pathology was consistent with GCT with secondary ABC. The clinical, radiologic and histopathological characteristics of the two conditions are described, and the treatment implication of this association is discussed.

CASE REPORT
History and Presentation: This 27-year old female initially presented to her outside physician complaining of swelling behind the left ear one month following bilateral molar extraction. She was empirically treated with antibiotics for a suspected infection but developed progressive left retroauricular headaches and mild dizziness. The patient had normal neurologic examination, including lower cranial nerve function, but had a large, non-mobile, non-tender mass behind the ear with no signs of erythema or drainage. The CT scan demonstrated an expansile, lytic lesion with a faint rim of residual inner and outer table of the left occipital bone (Figure 1A) and erosion of the condyle (Figure 1B). The MRI showed a multicystic lesion with fluid-fluid levels (Figure 1C) and heterogeneous enhancement (Figure 1D).

Figure 1
Figure 1. Preoperative CT and MR imaging of left occipital intrinsic bony lesion. A. Nonenhanced CT showing large expansile, osteolytic lesion with a thin rim of residual cortical rim in the left occipital bone (A) with erosion of...
much of the occipital condyle (B). T2-weighted MRI (C) showing a mixed-intensity, multicystic lesion with fluid-fluid levels and significant mass effect on the cerebellum and brainstem. T1-weighted MRI with gadolinium (D) demonstrating an enhancing, multicystic lesion invading the jugular foramen and abutting the internal carotid artery. Digital subtracted angiogram showing the hypervascular tumor supplied by the occipital artery (E) that was successfully embolized with coils (F).

Treatment & Postoperative Course: She underwent preoperative endovascular embolization of the hypervascular lesion resulting in a significant decrease in tumor vascularity (Figure 1E & F). The following day she underwent a far lateral suboccipital craniotomy, exposing a large, reddish-brown tumor (Figure 2A-C).

Figure 2

Figure 2. Intraoperative photographs showing a large reddish-brown mass involving the occipital bone. A rim of normal bone was circumferentially removed with a cutting burr (*) exposing normal, uninvolved dura mater. The C1 lamina was left intact (black arrow). Given tumoral adherence to the dura mater, a large section of dura was removed circumferentially around the tumor (B, black arrow). The mass was completely removed leaving the anterior cortical rim of the condyle intact (*). The dural sleeves of the lower cranial nerves (white arrow) and hypoglossal nerves (black arrow) were uninvolved with tumor (C).

A rim of normal bone was removed around the periphery of the lesion to facilitate en bloc removal and to attain negative margins. Given the adhesion of the tumor to the dura, a portion of dura was resected and the defect repaired with synthetic graft material. Following gross-total resection of the lesion, a mesh cranioplasty was inserted to cover the bony defect. Although most of the condyle was destroyed by tumor, instrumentation and arthrodesis was not performed. Postoperative MRI confirmed complete resection of the tumor (Figure 3) and postoperative flexion-extension radiographs demonstrated no evidence of instability at the atlanto-occipital junction.

Figure 3

Figure 3. Postoperative T1-weighted MRI without (A) and with (B) gadolinium showing complete resection of the tumor and fat graft sealing the mastoid air cells.

Sixteen months from surgery, she remains disease-free, without neck pain or instability.

Histological Examination: Cut surfaces of the gross specimen showed blood-filled spaces separated by septa. Microscopic examination revealed a giant cell-rich lesion of bone, predominantly composed of cystic components with features of an ABC, and focal areas of predominantly compact components with features of GCT of bone. There were irregularly shaped cystic spaces surrounded by fibrous septa (Figure 4).

Figure 4

Figure 4. Histopathological examination with hematoxylin and eosin-stained sections. A (original magnification X 10), ABC component of the lesion demonstrating cystic spaces surrounded by septa containing spindle fibroblastic cells without atypia and scattered multinucleated giant cells. B (original magnification X 10), GCT component of the lesion composed of mononuclear cells and giant cells. The giant cells are distributed uniformly. C (original magnification X 40), The mononuclear cells of the GCT show multiple round
or oval nuclei with uniformly distributed chromatin and indistinct nucleoli. The nuclear features are similar to those of mononuclear cells.

The septa contained spindle-shaped cells without cytological atypia, multinucleated giant cells and capillaries. There were foci of chondro-osteoid with characteristic blue quality ("blue bone") throughout the lesion characteristic for ABC. In some areas the lesion demonstrated features of GCT of bone with relatively extensive non-cystic solid component composed of numerous multinucleated giant cells that were scattered relatively uniformly (Figure 7). Focally, giant cells proliferation filled the vascular channels at the periphery of the lesion. In some areas, the lesion was attached to the dura and areas of peripheral reactive bony rim were identified with adjacent giant cell-rich lesion.

**DISCUSSION**

GCT is a benign tumor of the bone, comprising about 4-9.5% of all the bone tumors, and 18-23% of the benign bone tumors. It is slightly more common in females and in Chinese populations with a peak prevalence in the third decade of life. Although it is classified as a benign tumor, it can be locally invasive and aggressive. GCT commonly presents as a large lytic mass compromising the epiphysis of long bones, specially the femur, tibia and radius in patients over the age of 20. Only 2% of these lesions present in the head and neck, with the most common sites being the sphenoid, ethmoid, and temporal bones.

GCTs present radiographically as osteolytic lesions with indistinct margins, without periosteal sclerosis, and typically located in the epiphyseal region of a long bone. On MRI, GCT usually appears hypointense in T1- and T2-weighted imaging and enhance with the administration of contrast. This exam more accurately shows the tumor margin and any soft tissue extension. CT scanning is useful in evaluating the extension of the bony erosion and demonstrates prominent bony trabeculation and loculation areas.

The histopathology of GCT is characterized by frank and marked hemorrhage, numerous multinucleated giant cells and spindle-shaped fibroblast-like stromal cells. A biopsy of the solid components of the lesion is the only definitive way to establish the diagnosis of GCT. Macroscopically these lesions appear gray to yellow brown with small cystic areas and gray-white necrotic regions.

ABC is considered as a non-neoplastic, expansile lesion secondary to an existing pathological process of the bone, consisting of blood-filled spaces separated by connective tissue septa containing bone or osteoid and osteoclast giant cells. The surrounding bone cortex is “blown-out” into a very thin bony shell. ABCs represent 10% of benign osseous tumors and approximately 80% of the patients presenting with these lesions are less than 20 years old. It affects most commonly the metaphyseal region of the long bones, the pelvis and the spine, and only 2-6% involve the skull.

As many as 50% of the reported cases of ABC are secondary to a preexisting condition, but other large series found no instances of underlying pathology. It may arise in the setting of GCT, chondroblastoma, chondromyxoid fibroma, osteoblastoma, or fibrous dysplasia or in areas with a previous history of a local trauma. Less often it may arise from an osteosarcoma, chondrosarcoma, eosinophilic granuloma, solitary bone cyst and hemangioendothelioma. Among the reported cases in which a concomitant lesion was diagnosed (including cranial and extracranial sites), GCT was the most common underlying pathology, accounting for 14-39% of cases of secondary ABC. Our review identified 28 prior cases of secondary ABC involving the skull base or calvarium and they are summarized in Table 1.
The most common etiology was fibrous dysplasia (68%) and only 1 prior case of GCT and secondary ABC has been reported.11

The pathogenesis of ABC is unknown. It has been postulated that ABCs form secondary to vascular weakness caused by direct endothelial damage or indirectly caused by a defective collagen matrix of vessels, the surrounding parenchyma or both.9 Thus, the thin-walled vessels are markedly dilate over time and may give rise to balloon-like swellings within the bone forming an ABC over time.37

Painless swelling is the most common presenting symptom in patients with cranial ABCs but lesions are occasionally tender or pulsating in those instances where the inner table of the cranium is compromised.24 Neurological deficits may be found in tumors directly compressing the brain or cranial nerves. Consistent with their vascular pathophysiology, there are some isolated reports of ABCs presenting in association with an epidural hematoma and intracerebral hemorrhage.2,4

Radiographs of ABC typically show an eccentric lytic lesion with a “ballooned” contour of the bone peripheral, sclerosis and loculations, sometimes described as “soap bubble” or “blowout” appearance.6,15,27,31 In those cases with skull presentation, it usually involves both the inner and outer tables.31 CT scans will demonstrate a well-demarcated, multiloculated, osteolytic lesion with different densities and fluid-fluid levels in some cases.19,24,27 MR imaging will show a well-defined expansible lesion surrounded by a hypointense fibrous capsule and may have hypointense internal septations surrounding multiple cystic cavities.5,15,27,31 These internal compartments usually are enhanced with the infusion of gadolinium.15,27,31 Bone scintigraphy will show increased uptake of radionucleotides, markedly in the periphery of the lesion, reflecting the extent of bone involvement.15,17 Angiography will demonstrate increased vascularity predominantly in the periphery of the lesion, but no pathological vessels.15

The treatment of choice for ABCs is complete excision.24,33,37 However, appropriate treatment of ABC requires a thorough evaluation to detect any underlying pathologies, as these causative lesions often dictate the ultimate treatment and prognosis.15 Those cases of GCT associated with ABC should be treated with complete surgical resection of the lesion whenever possible, given that the cases described as partial resection or curettage showed a high index of recurrence.10 Recurrence rates for GCTs range from 7% in cases of total excision to 60% in cases of partial resection.10 Some cases of temporal bone GCTs treated with curettage (removal of intracavity tumor and drilling of the surrounding bone) showed no recurrence of the disease in a 2-year follow-up.10 Preoperative embolization reduces bleeding during surgery, and can be very helpful.8,15,30,33 There are case-reports of ABC treated with successful embolization of the feeding vessels resulting in progressive ossification of the lesion.15 The downside to this treatment approach is the inability to analyze the histopathological features of the lesion and possibly fail to diagnose an underlying malignancy. There are some reports of GCT treated with interferon alpha-2a demonstrating tumor stabilization and regression, suggesting that it can be used to control unresectable lesions.30 There are some reported cases of treatment with adjuvant radiation therapy without significant
decrease in the rate of recurrence or progression, and with documented cases of an occasional malignant degeneration.

10,17,23,40

CONCLUSION

We report a case of an ABC of the occipital bone in association with a GCT. Although GCT are a common cause of ABC, the co-occurrence of these lesions in the skull is rare. According to our review, this is the second case in the skull base or calvarium reported in the literature. In all cases of skull lesions presenting with an ABC-like image it is important to consider the possible existence of an underlying pathology. The associated primary condition often determines the clinical presentation, disease course of the ABC and the need for adjuvant therapy. The ideal treatment is complete resection of the lesion whenever possible, with or without preoperative embolization to limit blood loss, and to potentially make the surgical resection safer.

References

Author Information

Rafael Modkovski, M.D.
Department of Neurosurgery, Hospital de Clinicas de Porto Alegre

Robert E. Elliott, M.D.
Department of Neurosurgery, New York University Langone Medical Center

Benjamin Rubin, M.D.
Department of Neurosurgery, New York University Langone Medical Center

David Zagzag, M.D.
Department of Neurosurgery, New York University Langone Medical Center

Jafar J. Jafar, M.D.
Department of Neurosurgery, New York University Langone Medical Center

Irina Mikolaenko, M.D.
Neuropathology, New York University Langone Medical Center