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Aneurysmal Bone Cyst of the Temporal Bone: A Case Report with Review of Literature

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Abstract

Aneurysmal bone cysts (ABCs) are noncancerous growths that often develop in the metaphyseal region of long tubular bones. As a whole, ABCs consist of cystic cavities containing blood and hemosiderin enclosed in a subperiosteal shell of reactive bone with septae but not endothelium. Though nonmalignant, ABCs can grow aggressively and be locally destructive, leading to pathologic fractures. Early intervention with surgery is preferred to avoid pathological fractures. Radiotherapy must be considered in inoperable and recurrent cases. ABCs are more common in pediatric patients and cause more complications if the growth plate is involved. ABCs in the temporal bone are a rare occurrence. We report a case of ABC of the right petrous part of the temporal bone in a 38-year-old female patient who presented clinically with headache, facial nerve palsy, and same-sided sensorineural hearing loss. Noncontrast computed tomography (NCCT) scan showed a sharply marginated, smooth hyperdense lytic expansile lesion involving the right petrous bone. Magnetic resonance imaging findings corroborated with the NCCT findings. Surgical resection was performed. External beam radiotherapy was delivered to treat the residual tumor at a dose of 50.4 Gy in 28 fractions by intensity modulated radiation therapy technique over 5 weeks, and the patient was symptom-free on follow-up. This underscores the importance of managing ABCs, particularly those involving craniofacial bones, with surgery followed by radiation therapy for better treatment outcomes and patient survival.

Keywords
- aneurysmal bone cyst
- temporal bone
- case report

Introduction

Aneurysmal bone cysts (ABCs) are benign, osteolytic bone tumors characterized by blood-filled cavities and locally destructive growth. They represent ~9.1% of all primary bone tumors.[1] Although non-cancerous, ABCs can grow aggressively, causing local destruction and weakening bones, and increasing the risk of pathologic fractures under normal stress. The term "aneurysmal bone cyst" was first introduced by Jaffe and Lichtenstein in 1942.[2] These cysts are more frequently observed in children and adolescents and can lead to substantial complications, particularly when they affect the growth plates of bones. ABCs commonly occur in the femur, tibia, and vertebrae, although they may involve any bone.[3] Sixty-three skull ABCs have been reported in the literature, of which 21 involved the temporal bone.[4,5] ABCs are known for their expansive growth, which can lead to pain, inflammation, and damage to nearby joints and growth

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plates. ABCs most commonly appear during adolescence, with the average age of diagnosis being around 13 years. In fact, ~90% of cases are identified before the age of 30 years. Studies suggest a slightly higher occurrence in females, with a male-to-female ratio of 1 to 1.3.[6] While ABCs can develop in various bones, those occurring in the skull are particularly concerning due to their potential for severe symptoms. Skull-based ABCs make up ~2 to 6% of all cases.[5] ABCs of the temporal bone are rare, particularly in adults, and are usually treated with surgery alone.

Here, we report a case of an ABC located in the right petrous portion of the temporal bone in a 38-year-old female patient. She was treated with a multimodality treatment approach (i.e., surgery followed by radiation therapy), leading to increased survival and better quality of life. This represents 1 of only 12 reported temporal bone ABCs, and is distinctive for (1) occurrence in a 38-year-old patient (while >90% occur before age 30), (2) use of high-dose adjuvant radiotherapy (50.4 Gy) for residual disease, and (3) 5-year symptom-free follow-up—the longest documented in temporal bone ABCs.

Case Report

A 38-year-old female patient presented with intermittent headache, deviation of the right angle of the mouth, decreased hearing in the right ear for 2 years, and diminished vision in the right eye for 3 weeks. There was a history of a road traffic accident 2 years back. There was no history of loss of consciousness, seizures, or weakness of extremities. All the vital parameters were within normal limits. Neurological examination revealed normal higher mental functions and speech with cerebellar gait. Visual acuity in both eyes was 6/6. The right V, VII, VIII, and XII cranial nerves' lower motor neuron palsy was found on examination. Routine laboratory investigations were within normal limits. Pure tone audiometry testing showed evidence of sensorineural hearing loss in the right ear. Noncontrast CT (NCCT) head (►**Fig. 1A**) revealed a 6.3 × 4.2 × 4.3 cm well-defined extra-axial, smoothly marginated, mildly hyperdense lytic expansile lesion epicentered at the right petrous bone. Anteromedially, the lesion is noted to cross the petroclival suture and erode the right lateral aspect of the clivus as well as involve the carotid canal. Anteriorly, it is seen extending into and obliterating the right middle ear and involving the otic capsule. Laterally, there is complete obliteration of the mastoid air cells. Medially, it obliterates the right cerebellopontine angle, involving the jugular fossa and hypoglossal canal, compressing and displacing the right cerebellar hemisphere and fourth ventricle to the left. Posteriorly, the lesion extends into and involves the right lateral aspect of the occipital bone and the sub-occipital region. The lesion has multiple rarefied bony fragments. MRI of the brain (►**Fig. 1B**) showed a 6.9 × 4.2 × 4.4 cm (AP × TR × CC) well-defined extra-axial lytic lesion with avid contrast enhancement. The lesion is obliterating the right internal auditory canal and is surrounding the cochlea and semicircular canal. The right VII/VIII nerve is not seen separately. The petrous segment of the internal carotid artery is displaced anteriorly and is thinned out. The right transverse and sigmoid sinuses are not visualized. Superiorly, the lesion is seen reaching up to Meckel's cave. MRI findings corroborated the NCCT findings. CT cerebral angiography was suggestive of a right petrous bone lesion surrounding and mildly displacing the petrous and lateral segments of the internal carotid artery, which was mildly narrowed. However, no filling defect or hypertrophied vessels supplying the lesion were noted. In digital subtraction angiography, a moderate tumor blush supplied by the ascending pharyngeal artery was embolized. The patient underwent a right retromastoid suboccipital craniotomy by the neurosurgical team, and the excision of the lesion was done. The facial nerve was identified and preserved using intraoperative monitoring. The excised tumor was sent for histopathological examination. Histopathology revealed multiple tissue fragments of blood-filled cystic spaces separated by a fibrous wall, which is composed of moderately dense cellular proliferation of bland fibroblasts, numerous scattered multinucleated osteoclast-type giant cells. At places, new bone formation, reactive woven bone rimmed by osteoblasts, and basophilic blue bone formation are seen. Focally, the cysts are seen to be lined by giant cells. The features are suggestive of ABC (solid variant). Postoperative period was uneventful, and postoperative contrast-enhanced MRI showed a reduced bulk of the lesion measuring 3.1 × 5.6 × 3.5 cm (AP × TR × CC) compared with the preoperative size of 6.9 × 4.2 × 4.4 cm. The multidisciplinary tumor board opined for external beam radiotherapy with a higher dose for the residual tumor. Radiotherapy was administered to the patient at a dose of 50.4 Gy in 28 fractions by intensity modulated radiation therapy technique over 5 weeks (►**Fig. 2**). The patient was followed up monthly for the first 3 months, then 3 monthly for the next 2 years, and then 6 monthly up to 5 years. Following treatment completion, the patient was monitored for 66 months and remained entirely asymptomatic throughout this follow-up period.

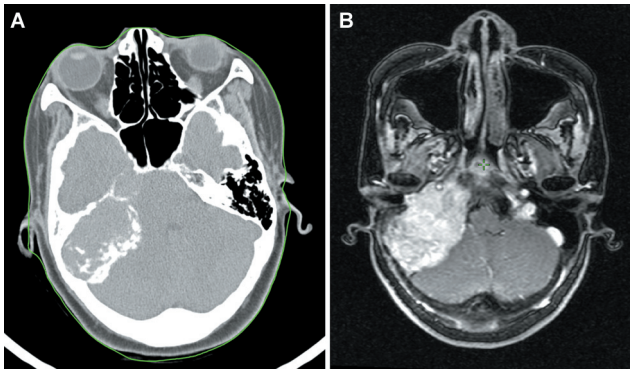

Fig. 1 Axial section of CT (**A**) and T1-weighted MRI (**B**) image showing a large, hyperdense/hyperintense, expansile, multiloculated, osteolytic mass epicentered at the squamous and petrous part of the right temporal bone, compressing the adjacent brain parenchyma.

Discussion

ABCs are rare, benign bone tumors, accounting for approximately 1 to 6% of all primary bone tumors. Their incidence is

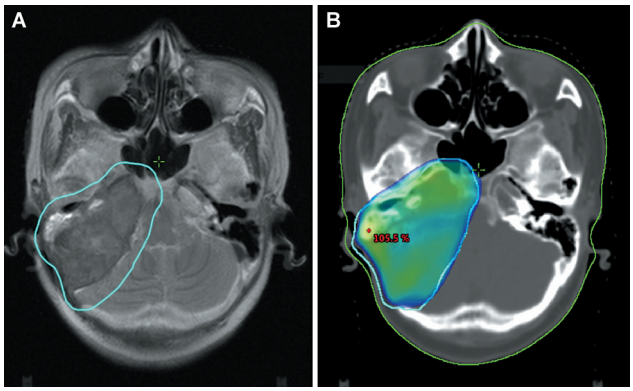

Fig. 2 CT and MRI fusion image shows adequate coverage of residual tumor and postoperative bed (**A**) and dose distribution showing 95% coverage of the planning target volume (cyan) (**B**) at the right temporal bone.

quite low, estimated at 0.14 cases per 1 million people per year. These cysts are most commonly diagnosed in children and adolescents younger than 20 years, with a slight predominance in females (male-to-female ratio of 1:1.3).[5] The majority of ABCs (67%) occur in the metaphysis of long bones, but they can also affect the spine (15%, particularly the posterior elements), pelvis (9%), craniofacial bones, and epiphyses.[7] On X-ray and CT scan, an expansile bony lesion with septations, a hypodense rim of capsule, and numerous lobulations with varying signal intensities giving a "bubbly" appearance should raise the suspicion of ABC. The development of ABCs is primarily linked to the upregulation of the TRE17/USP6 oncogene due to gain-of-function mutations, which disrupt osteoblastic maturation.[8] Current theories suggest that vascular abnormalities lead to increased expansile pressure within the bone, causing erosion and resorption, ultimately forming the cyst.[4] On gross examination, ABCs consist of blood-filled cystic cavities containing blood and hemosiderin, surrounded by a thin shell of reactive bone. The cavities are divided by septa made of osteoid tissue, lacking an endothelial lining. The stroma of ABCs contains fibroblasts, spindle cells, osteoid, and multinucleated giant cells, often clustered at the periphery of the cysts, creating a "pigs-at-the-trough" appearance.[4] While mitotic figures are common, atypical mitoses are absent, helping distinguish ABCs from malignant lesions. Diagnosis primarily relies on radiological imaging, including X-rays and NCCT scans. X-rays typically show expansile, osteolytic lesions with thin, "eggshell" sclerotic borders and internal septations. The aggressive, expansile nature of ABCs can displace surrounding bone. CT scans provide a more detailed visualization of the cystic septa and the "eggshell" rim. Fluid-fluid levels, caused by the separation of cellular debris and serum, are often visible. MRI findings are similar to CT, with T1- and T2-weighted images highlighting the septa and showing areas of hyperintensity due to blood products. Pathologic fractures may also be evident, accompanied by bone and soft tissue edema. Laboratory tests are generally not useful for diagnosis, though alkaline phosphatase levels may be elevated due to increased osteoblast activity.[9]

Given the risk of pathologic fractures due to their expansile nature, surgical intervention is the primary treatment for ABCs. The surgical approach–intralesional curettage, excision, or en bloc resection—depends on the lesion's size and location. Intralesional excision is often preferred. For recurrent ABCs, particularly in the temporal bone, radiotherapy may be considered.[10] Megavoltage radiotherapy is an option for inoperable or recurrent cases. Studies, including a retrospective review by Zhu et al, have shown favorable outcomes with radiotherapy, with a 4-year local control rate of 90.9% compared with 55% for surgery alone.[5,11] Unique features of this case include adult onset, extensive petrous involvement, nerve preservation via intraoperative monitoring, and durable control using 50.4 Gy radiotherapy—a dose exceeding conventional recommendations.

Despite ongoing debates about their pathogenesis, ABCs generally have an excellent prognosis. The standard treatment is intralesional curettage, with or without adjuvant therapy.[12] However, surgery has limitations, including a high recurrence rate and difficulty accessing lesions in certain anatomical locations.[13] Radiotherapy serves as an effective adjuvant or alternative, particularly for residual disease or hard-to-reach areas. Studies, including one by Zhu et al,[11] support the use of radiotherapy, reporting a 100% local control rate when used alone or postsurgery. However, due to the risk of radiation-induced malignancies, especially in young patients, radiotherapy should be reserved for cases where surgery alone is insufficient or impractical.

The recommended radiation dose is 26 to 30 Gy, delivered in 1.8 to 2.0 Gy daily fractions or 1.2 Gy twice daily, to balance efficacy and minimize side effects.[11] In some cases, a slightly higher dose may be used to manage residual disease and improve local control. While standard RT doses for ABCs range from 26 to 30 Gy,[11] we administered 50.4 Gy due to (1) large residual volume ($3.1 \times 5.6 \times 3.5$ cm) compressing critical structures, (2) solid-variant histology suggesting aggressive potential, and (3) proximity to the brainstem requiring precise coverage. This aligns with Feigenberg et al's finding that doses >35 Gy improve control in complex skull-base ABCs.[13] A watchful-waiting strategy was deemed unsafe given the risk of irreversible cranial neuropathy from regrowth. The 5-year remission without radiation necrosis (►**Fig. 2B**) supports this approach, though lifelong monitoring for secondary malignancies is warranted. Radiotherapy is contraindicated in ABCs associated with fibrous dysplasia due to the risk of malignant transformation. Chemotherapy has no role in ABC management, but preoperative endovascular embolization can reduce bleeding during surgery, especially for tumors in difficult locations.[14] There is still no available consensus guideline for the use of radiation in ABCs, adjunct to surgery for better treatment outcomes; mostly due to the rarity of the cases. Further research to treat ABCs with a multimodality approach (i.e., surgery followed by radiation therapy) should be considered to develop a consensus guideline for better patient care.

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Conclusion

ABCs of the temporal bone are exceptionally rare, particularly in adults, and pose unique therapeutic challenges. Surgery remains the cornerstone of management; however, adjuvant radiotherapy has an important role when complete resection is not feasible or when residual disease persists. This case highlights the effectiveness of a multimodality approach, with surgery followed by radiotherapy, in achieving durable 5-year local control and long-term symptom-free survival. While radiotherapy must be used judiciously due to potential risks, carefully selected patients can benefit significantly, especially in complex skull-base lesions. Adjuvant radiotherapy (50.4 Gy) is safe and effective for residual skull-base ABCs when surgery alone is insufficient. Reporting such unusual presentations not only expands clinical understanding but also emphasizes the need for developing consensus guidelines to optimize outcomes in rare craniofacial ABCs.

Patients' Consent

The authors certify that they have obtained all appropriate patient consent forms before preparing the manuscript. In the forms, patients have given their consent for their images and other clinical information to be reported in the journal.

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Conflict of Interest

None declared.

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