Case Report

Histopathological Features of Aneurysmal Bone Cyst of the Temporal Bone – A Case Report and Review of Literature

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Aneurysmal bone cyst (ABC) is a rare lesion. We described an ABC of the temporal bone. The patient was a 46-year-old woman presenting with a progressively enlarging mass in the left temporal region with unknown duration. A CT revealed a 3 x 3.5 cm osteolytic lesion involving the left temporal bone. MRI revealed a 3.6 x 2.6 x 2.7 cm well-defined expansile lesion centered in the left temporal bone. The patient subsequently underwent MRI-guided stereotactic left frontotemporoparietal craniectomy and the lesion was completely removed. Following surgery, the patient has been followed for the past 9 months without evidence of residual/recurrent disease. Morphologically, the lesion consisted of cystic and solid areas. The cystic areas demonstrate multi-cystic architecture between bony trabeculae separated by thick septa. These septa contained abundant fibroblasts, spindle-shaped stromal cells, and giant cells with scattered hemosiderin laden macrophages. The solid areas consisted of spindle-shaped stromal cells with oval nuclei, proliferating blood vessels, mononuclear cells and some interspersed multinucleated giant cells.
[N A J Med Sci. 2015;8(1):46-49. DOI: *10.7156/najms.2015.0801046]*

Key Words: *aneurysmal bone cyst (ABC), temporal bone, bony trabeculae*

INTRODUCTION

Aneurysmal bone cyst (ABC) is a rare, benign disease characterized by osteolytic multicystic lesions. The disease is most common in adolescents showing a slight female predominance.[1] ABC can occur virtually in any location, although they are most commonly encountered in the metaphyses of long bones (50%) and vertebrae (20%). ABC of the temporal bone is even rarer with only 27 cases reported in the English and Non-English language medical literature.[2] These reports mainly described clinical manifestations, radiology diagnostic findings and management of these lesions. In this report, we present an ABC of the temporal bone in a 46-year-old female, and focus on its histopathological findings.

CASE REPORT

A 46-year-old woman noted a progressively enlarging mass in the left temporal region which had been present for an unknown duration. The patient denied headache but did report nonspecific visual changes over the past months to years prior to her detection of this mass; she denied other neurologic symptoms. When this mass failed to improve over a number of weeks, she sought care. A CT revealed a 3 x 3.5 cm osteolytic bony lesion involving the left temporal bone. MRI revealed a 3.6 x 2.6 x 2.7 cm well-defined expansile lesion centered in the left temporal bone (**Figure 1**).

The patient subsequently underwent MRI-guided stereotactic left frontotemporoparietal craniectomy and debulking surgery with middle fossa and infratentorial approach. During the procedure, dural involvement was noted, so a large area of dura measuring greater than 3 cm was resected, up to a clear margin on frozen section pathology consultations. The final pathologic diagnosis, as discussed below in further detail, was aneurysmal bone cyst.

Following surgery, a surveillance without adjuvant therapy and close observation was recommended. The patient has been followed with serial MRI studies for the past 9 months without evidence of residual/recurrent disease.

Features of Histopathology

Histopathologically, the lesion appeared to have a biphasic pattern with cystic and solid areas. The cystic areas demonstrated multi-cystic architecture between bony trabeculae separated by thick septa. These cysts varied in size and contained red blood cells. The septa that separated the lesion into multiple cysts contained abundant fibroblasts, spindle-shaped stromal cells, and giant cells with scattered hemosiderin laden macrophages. The solid areas consisted of spindle-shaped stromal cells with oval nuclei, proliferating blood vessels, mononuclear cells and some interspersed multinucleated giant cells (**Figure 2**).

Received: 06/05/2014; Revised: 01/16/2015; Accepted: 01/18/2015
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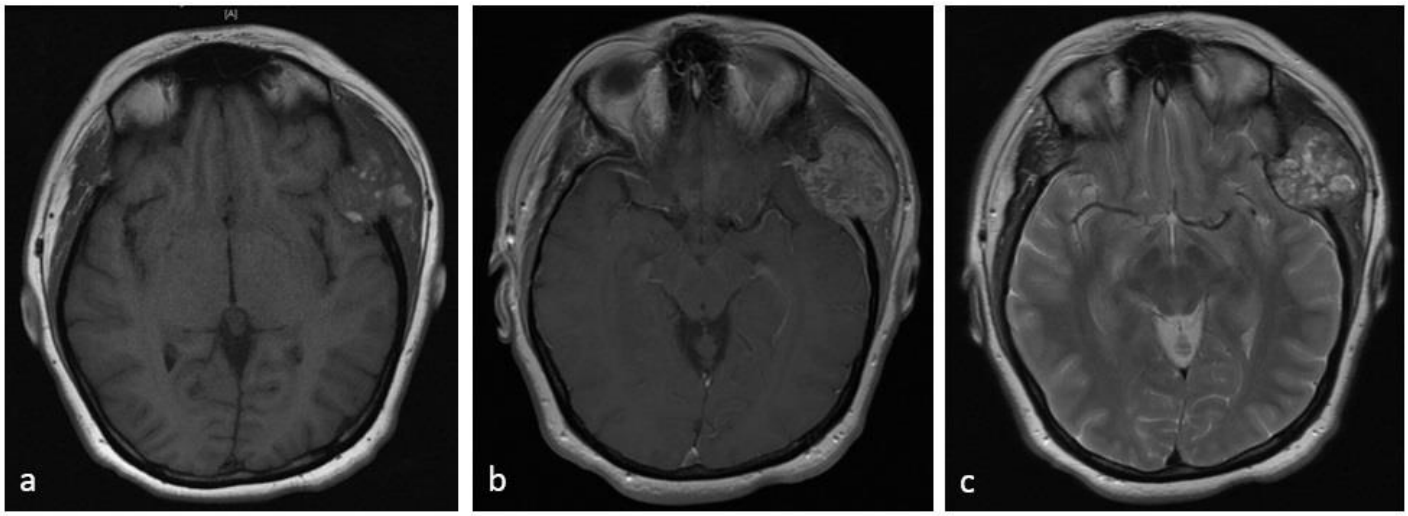

Figure 1. Aneurysmal bone cyst on MRI imahing. **(a)** Axial T1 imaging pregadolinium revealing heterogenous hyperintensity. **(b)** Axial T1 imaging postgadolinium revealing extension to dural surface. **(c)** Axial T2 imaging postgadolinium reveals heterogenous T2 hyperintensity.

Table 1. Clinical and histopathologic features of ABC from the previously reported cases and this case.

AGE (YR) AND SEX	# OF CASES	MICROSCOPIC FEATURES	TREATMENT	RECURRENCE AND PERIOD OF FOLLOW-UP (YR)	REFERENCES
13 F	1	Cavernous spaces filled with blood. The spaces did not show any endothelial lining. Septa contained fibroblasts, focal collections of osteoclastic and intermediate giant cells	Near total excision of cyst	No recurrence, 7 months	Purohit et al. 2002 [15]
45 M	1	N/A	Partial resection of the tumor was carried out	The residual tumor mass was somewhat less than before, 2	Buxi et al. 2004 [12]
60 M	1	Biphasic structure with cystic and solid areas. The cystic areas were composed of thick septa of fibromyxoid connective tissue lined by mesenchymal-type cells; The solid areas were composed of spindled-shaped stromal cells interspersed with multinucleated giant cells	Mastoidectomy with piece meal resection of tumor; the dura was extensively coagulated	No recurrence, 6 months	Sabatini et al. 2005 [11]
17 M	1	Numerous variably sized blood filled spaces separated by fibrous septa containing spindle shaped cells and scattered multinucleated giant cells	Cyst was removed completely by a superior approach to the infratemporal fossa	N/A	Goyal et al. 2006 [16]
12 F	1	Blood-containing cystic spaces were separated by fibroblasts and reactive bone formation was also discovered.	Totally removed	No evidence of mass in the orbit, 7 months	Yu et al. 2012 [5]
5 1/2-year-old M	1	Numerous telangiectatic vascular channels, some packed with RBCs. The intervening fibrous septa were cellular with numbers of multinucleate osteoclast giant cells. Within the septa, there were areas of new bone formation.	Partially removed	Being kept under observation	Chidambaram et al. 2001 [17]
46 F	1	Biphasic pattern with cystic and solid areas. Solid areas and septa contained abundant fibroblasts, spindle-shaped stromal cells, and giant cells with scattered hemosiderin laden macrophages. Bony trabeculae were lined by a complete or incomplete layer of endothelial cells or lack lining layer.	Completely removed	No evidence of residual/recurrent disease, 9 months	This case

DISCUSSION

Grossly, ABC is typically described as friable, multilocular, and vascular masses.[3,4] When the mass is squeezed, the unclotted bloody fluid is released.[5] In 1942, Jaffe and Lichtenstein first described ABC as a benign, expansive bone lesion with a "soap bubble" or "blowout" radiographic appearance.[6] In this case, at surgery, the lesion was removed in a piecemeal fashion. We received multiple fragments of red brown tan soft muscular tissue and bony segments so that a detailed gross examination description was not possible. But the MRI image (**Figure 1**) well demonstrated the ABC and the physical relations with its neighbors.

Differential diagnoses include telangiectatic osteosarcoma, giant cell tumor, low grade osteosarcoma especially with the reactive osteoid beams surrounding ABC lesion, and so on. In telangiectatic osteosarcoma, the tumor consists of large spaces filled with blood and separated by thin bony septa. Microscopically the tumor comprises vascular sinusoids surrounded by thin septa, osteoid matrix and significant pleomorphic cells with high mitotic rate. These giant cells have key histomorphologic features - up to a hundred nuclei with prominent nucleoli. Surrounding mononuclear and small multinucleated cells have nuclei that are similar to those in giant cells. In low grade osteosarcoma especially with the reactive osteoid beams surrounding ABC lesion, the tumor cells demonstrate low-moderate nuclear pleomorphism, clumped chromatin and minimal mitotic activity with no giant cell proliferation. Some areas of mineralization and chondroblastic differentiation of the osteoid can be identified. There are mature woven bone and cortical bone components present. In addition, sometimes a small piece of ABC tissue on biopsy may mimic a spindle cell malignancy or meningioma.[7] Martinez et al[9] reported that one third of ABC (36 cases out of 123) did co-exist with other bone tumors, such as giant cell tumor, chondroblastoma, chondromyxoid fibroma, nonossifying fibroma, osteoblastoma, fibrosarcoma, fibrous histiocytoma, osteosarcoma, and fibrous dysplasia.[7]

The etiology of ABC has not been determined. Based on the previous reports, we summarized some clinicopathological features, treatments, as well as prognosis of ABC in **Table 1**. Panoutsakopoulos et al revealed a clonal nature of ABC and its association with t(16;17)(q22;p13).[13] Later, Herens et al found that the translocation (16;17)(q22;p13) was a recurrent anomaly of ABC.[14] Recently, Oliveira et al reported that the gene rearrangements of the USP6 gene on chromosome 17, and/or the CDH11 gene on chromosome 16 was involved in ABC.[18] However, the recent identification of recurrent chromosome abnormalities indicated that the lesion might result from arrest of maturation of the osteoblasts caused by USP6 overexpression and dysregulation of autocrine BMP.[19]

In short, aneurysmal bone cyst is a rare, benign lesion with pathologic features commonly leading to underdiagnosis or misdiagnosis. This case mainly described the pathologic findings of a clinically aggressive ABC with dural invasion and demonstrated the primary role of surgery in the management of this disorder.

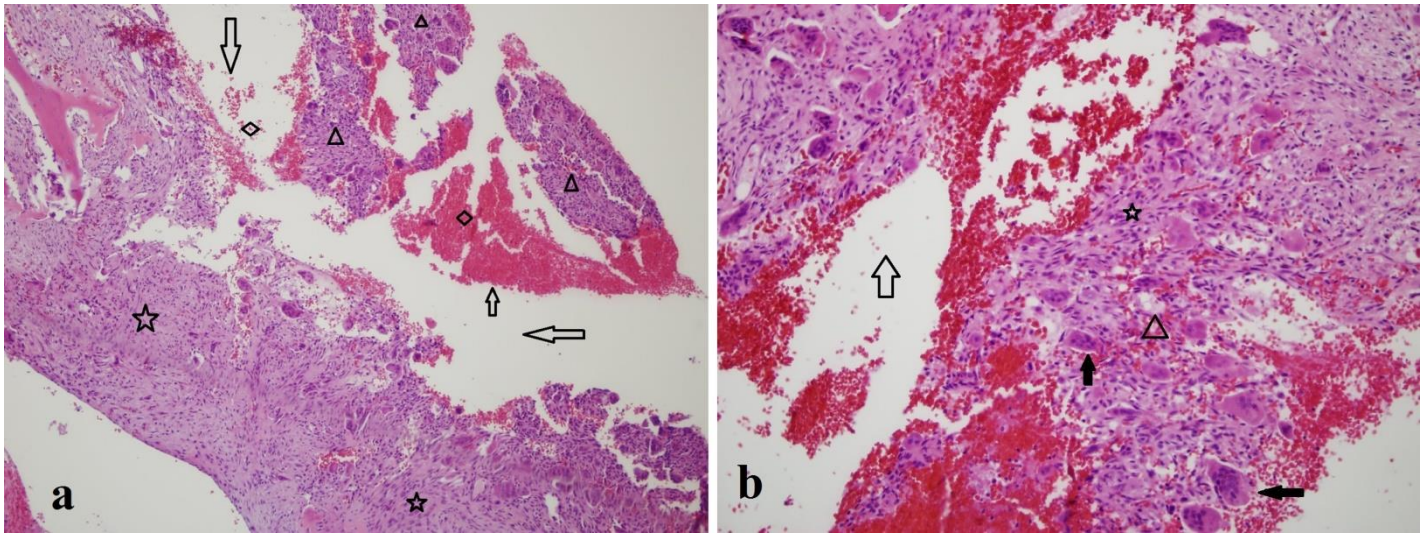

Figure 2. (a) The microphotograph showed cystic areas (open arrow) and septal structures (triangle) of aneurysmal bone cyst as well as solid areas (star). Some cysts contain red blood cells (diamond). (Haematoxylin-eosin, x 100); **(b)** The Higher magnification microphotograph demonstrated the cystic areas (open arrow) and septa (triangle) of aneurysmal bone cyst. The spindle-shaped stromal cells (star) and giant cells (solid black arrow) are seen. (Haematoxylin-eosin, x 400)

CONFLICT OF INTEREST

The authors have no conflict of interest to disclose.

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