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Aneurysmal Bone Cyst of Clivus and C1 C2: Case Report and Review of Literature

Klivus, Servikal 1-2'de Yerleşmiş Anevrizmal Kemik Kisti: Olgu Sunumu ve Literatür Taraması

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ABSTRACT

Aneurysmal bone cyst are benign rapidly expanding bone destructive lesions of any bone. They are common in the metaphysis of long bones but 10-30% involve the spine. Cervical region involvement is uncommon. We report a case of aneurysmal bone cyst of clivus C1 and C2 with minimum symptoms. Involvement of C1 and clivus separately had been reported in past, but simultaneous involvement of both is presented in this report for the very first time.

KEYWORDS: Aneurysmal bone cyst, Spine, Clivus

ÖZ

Anevrizmal kemik kistleri herhangi bir kemikte görülebilen iyi huylu ancak tutmuş olduğu kemikte hızla genişleme yaparak harabiyete neden olur. Uzun kemiklerin metafizlerini yaygın olarak tutmakla beraber yüzde 10-30 oranında omurgada tutulum yapabilir. Servikal bölge tutulumu nadirdir. Klivus, servikal 1 ve 2. omurlarında anevrizmal kemik kistine bağlı tutulumu olan ve minimal semptomu olan hasta olgu olarak sunulmaktadır. Geçmişte, clivus veya servikal 1. omurun anevrizmal kemik kisti ile tutulumu ayrı vakalar halinde sunulmuştur, bizim olgumuz ise eşzamanlı olarak klivus ve servikal 1-2 omurgalarının aynı olguda tutulması ilk kez sunulmaktadır.

ANAHTAR SÖZCÜKLER: Anevrizmal kemik kisti, Omurga, Klivus

INTRODUCTION

Aneurysmal bone cysts are benign bone tumors that most commonly occur in people younger than 30 years. The cysts are most often found in the metaphyses of long bones and rarely affect the skull and cervical spine. The present article reports a 45-year-old woman with aneurysmal bone cyst of the atlas C2 vertebral body and clivus and reviews the literature in regard to the presentation, radiology and treatment options for an aneurysmal bone cyst.

CASE REPORT

A 45-year-old female presented at the Neurosurgery outpatient department with complaints of mild to moderate localized pain in the nape of the neck, for which she took analgesics occasionally. She had developed slurred speech, difficulty in swallowing and difficulty in breathing through nose so she used to breathe through mouth. She had no history of fever, trauma or limb weakness. There was no comorbid condition. Her general physical examination was normal. Neurological examination revealed hypernasality of speech. Cranial nerve examination was normal. However, there was a bulge over the posterior pharyngeal wall. X ray cervical spine showed destruction of the C1 and clivus. Computed tomogram of the spine showed an expansile lesion with a thin rim of cortical margin involving the clivus, C1 anterior arch and C2 vertebral body (Figure 1). Craniospinal MRI showed a heterointense mass on T1W1 and T2W1 with heterogeneous enhancement on contrast in the retropharyngeal region also involving the clivus and anterior and posterior arches of atlas and C2 vertebral body. The lesion had classical fluid-fluid levels. The lesion extended to the posterior aspect into the muscle plane and was more on right side (Figure 2).

Decompression at the craniovertebral junction was done anteriorly as well as posteriorly in two stages on the same day. Posterior decompression was done through a midline incision from inion to C5 spinous process in the prone position. The tumor was moderately vascular with reddish-brown color encasing the third part of the vertebral artery. The posterior arch of C1 was destroyed. The vertebral artery was mobilized from the foramen transvarsarium of C2 and decompression was done till facet joints bilaterally. Reconstruction was done with pedicle screws, rod and occipital plate between occiput, C2, C3 and C4. The position was changed to supine and decompression was done through the transoral route. Anteriorly, the C1 arch was not seen separately. In the postoperative period she was kept on elective ventilation for two days and then gradually weaned off. There was subjective improvement in speech, breathing and deglutition.

Histopathology revealed multiple cystic lesions separated by fibrous septae having spindle-shaped cells with multiple multinucleated giant cells (Figure 5).

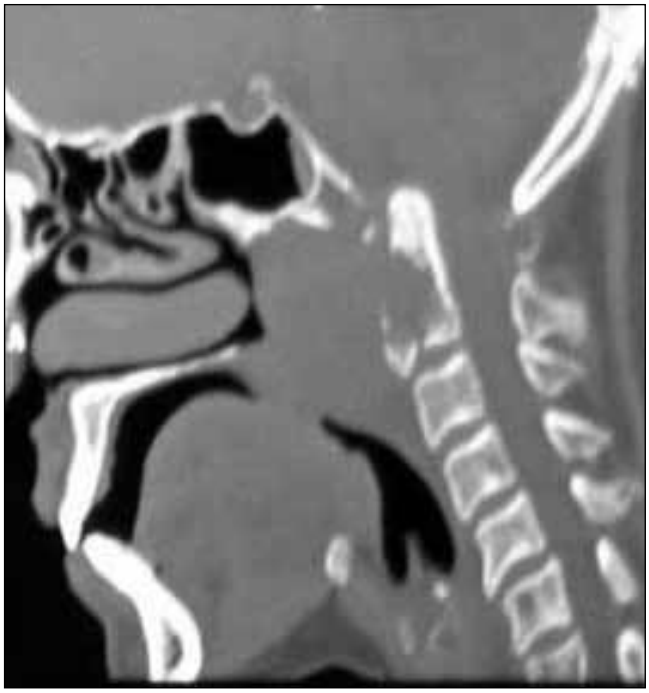

Figure 1: CT scan of the patient showing destruction of clivus, C1 and C2 vertebral body.

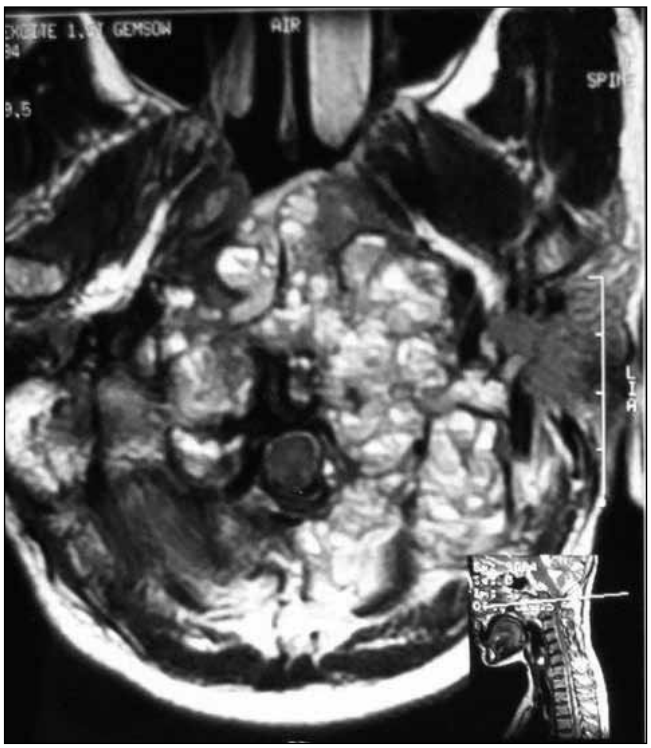

Figure 2: MRI of the patient showing typical fluid-fluid appearance of aneurysmal bone cyst involving clivus, C1 and C2.

Though post operative MRI (Figure 3) showed sufficient decompression, the tumor size had increased on the right side. Adequate stabilization was apparent on postoperative CT scan and X rays of the patient (Figure 4). She was considered for adjuvant radiotherapy and referred to the radiotherapy department. During follow-up for 1 year, she had no new complaints or neurological deficit.

DISCUSSION

Aneurysmal bone cysts are relatively uncommon, well-differentiated expansile benign tumors that present commonly in the first and second decade and are more common in females (19,9). Most of them are located in the metaphyses of long bones, and the knee joint is commonly affected where it affects tibia upper end. Other sites of involvement are flat bones and the vertebral column. 25% involve the vertebral body, hyoid, mandible and odontoid (5, 10). Aneurysmal bone cysts are common in the lumbar and thoracic spine. Involvement of the cervical region is a rare entity seen at a rate of 2%. Cases of cysts involving the C1 alone have been reported (7, 12, 15, 14). Cases with involvement of clivus and skull base without involving C1 were also reported (1, 17). Our case is unusual as the patient is a 45-year-old female and there is involvement of the posterior and anterior arch of the C1 C2 vertebral body and clivus, which has not been reported previously. It starts to involve

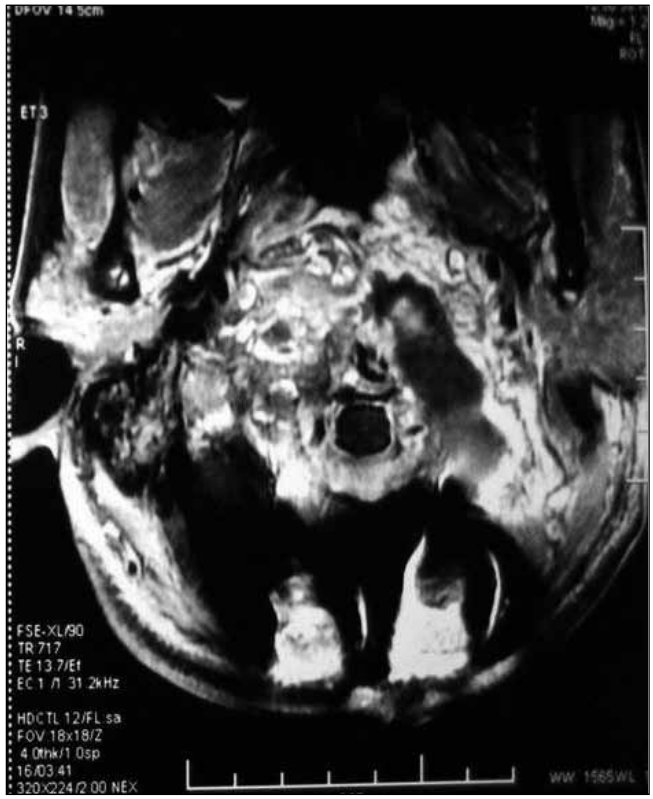

Figure 3: Postoperative MRI of the patient revealing decompression of the cyst on the left nasopharyngeal region with increased growth in right petrous region.

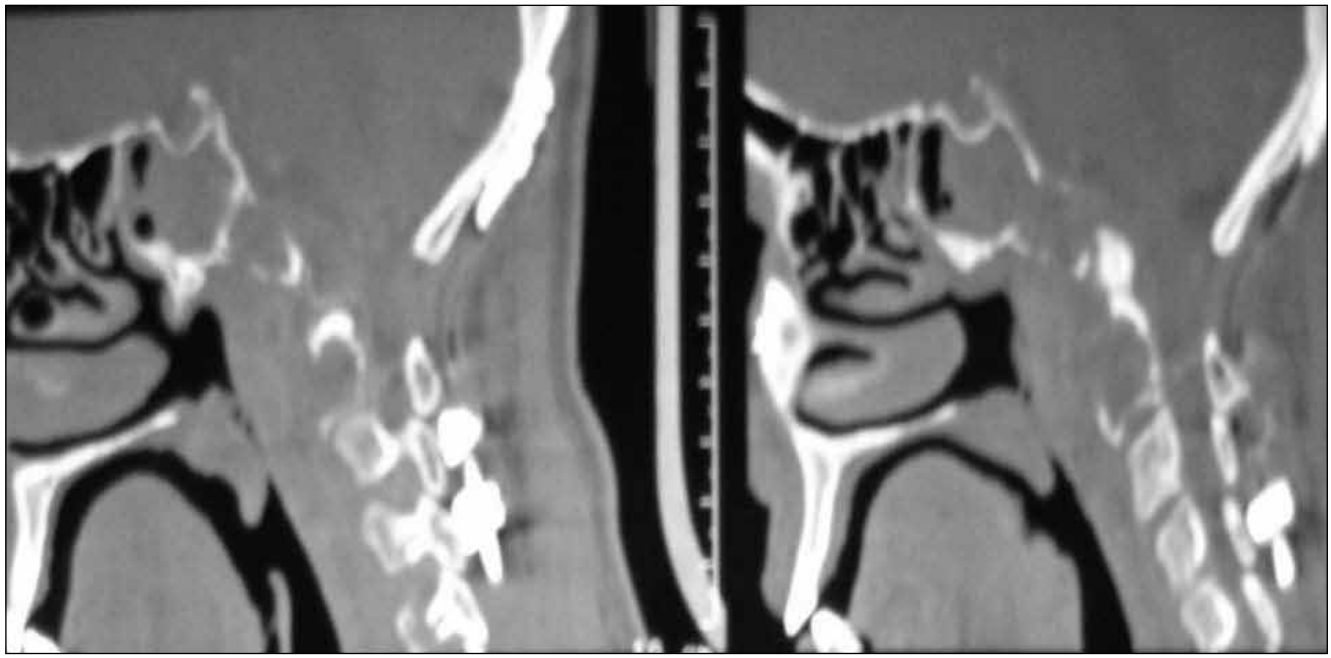

Figure 4: Postoperative X rays of the cervicooccipital junction, Lateral and AP views.

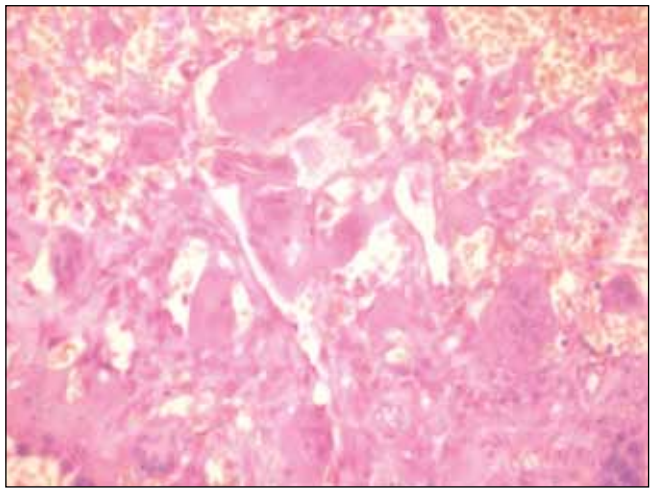

Figure 5: Histopathology (Hematoxylin and eosin staining, x25).

posterior elements first, followed by involvement of anterior part of vertebra.

The etiology of aneurysmal bone cyst is not known. These may present as primary in the bone and sometimes may be associated with other bone pathology like giant cell tumors, osteosarcoma, osteoblastoma, chondroid myxoma, chondroblastoma, fibrous dysplasia or with trauma (6). They are supposed to develop because of local hemodynamic changes. This has been described as a "reparative process gone out of control" by Cooley and as hemorrhagic "blow out of a pre- existing lesion" by Jaffe (11,18).

These lesions are characterized by the presence of many cavernous blood-filled cavities separated by thin osseous tissue or bony trabeculae. These are not lined by endothelium. These septations are highly vascular. This section is in turn surrounded by a spindle cell stroma with osteoclast-like giant cell and osteoid and bone production. Three stages have been described; an early lytic phase, a mature phase with a peripheral bony shell and internal septa and a late calcified stage. Another variant called the solid variant has also have been reported and is an intraosseous fibroblastic lesion with scattered osteoclastic , osteoblastic and fibromyxoid elements without a prominent component of blood-filled cavities (6).

Clinical presentation is varied. Patients commonly complain of pain and restriction of movements in affected parts. Other presentations are local deformity, torticolis, muscular stiffness and sometimes a murmur over affected region. The present case had mild to moderate localized pain at the nape of neck, slurred speech, difficulty swallowing and difficulty breathing. The natural history is also varied (11). Spontaneous regression and regression after biopsy have been reported (13) In contrast, they are also reported to recur in a well-healed fusion mass (18). Though they are benign, they have a rapid course and the cyst may present with compression of adjacent structures or with pathological fractures (19). Sarcomatous degeneration has been reported after radiotherapy (3). However we found only two well-documented reports of malignant transformation of an aneurysmal bone cyst without a history of irradiation. One transformation was to a malignant fibrous histiocytoma, and the other was to an osteosarcoma.

X ray spine may detect advanced disease in the form of expansile bone or pathological fractures. CT scan is more reliable and detects early stages of the disease, showing well-demarcated multiloculated expansile lesions with thin cortical margins and also fluid/fluid levels in about 30%

of cases. MRI shows heterointensities in T2W1 and T1W1 images with heterogeneous enhancement on contrast. There are multiple fluid-fluid cavities which show the presence of degraded blood. The MRI of our patient had a typical appearance of fluid-fluid levels. Fluid/fluid levels are also seen in chondroblastomas, telangiectatic osteosarcomas and giant cell tumors (16).

Various treatment options are available. Best modality of treatment is complete resection of lesions with reconstruction. Local recurrence rates of up to 20 % have been described for lesions treated with surgery alone. Other options are curette of lesion and grafting, radiotherapy (2), selective arterial embolisation (12) and percutaneous intralesional injection of calcitonin and methyl prednisone (7). Curettage of the lesion is associated with incomplete removal of tumor and high rate of recurrence. Radiotherapy causes asymmetrical growth of exposed bones and development of deformities in growing children (10). It also known to cause sarcomatous degeneration of he lesion. As aneurysmal bone cysts are highly vascular tumors, presurgical embolisation of feeders is done as adjunct treatment. Cases have been reported to cured by selective arterial embolisation at several sittings (4,8,12). However, this technique requires highly advanced skill to negotiate the feeders and there is also a chance of cord ischemia, especially in the thoracic region which already has less vascularity (8,12,4). Percutaneous intralesional injections of calcitonin and methyl prednisone may cure patients if the lesion size is small and there is no spinal instability (7).

If an aneurysmal bone cyst is quite large and affects the whole of the vertebral body as commonly occurs in the developing world, treatment is resection of tumor and reconstruction. The tumor was very extensive in our case and involved C1, C2 and clivus with extension into the retropharyngeal space. Complete excision was therefore not possible and could have lead to severe morbidity in a patient with minimal deficits. We therefore decompressed anteriorly as well as posteriorly and reconstruction was done to maintain stability of the spine. The patient was then referred to radiotherapy for adjuvant radiation treatment.

CONCLUSION

Aneurysmal bone cyst is a benign pathology but has a rapid course. These lesions may need surgical resection or decompression depending upon accessibility of affected site and the size of the lesion followed by reconstruction especially in less developed countries where presentation occurs at the late stage. Any residual tumor may be treated by radiotherapy.

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