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Case Report

Aneurysmal bone cyst of thoracic spine with neurological deficit and its recurrence treated with multimodal intervention – A case report

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ABSTRACT

Background: Aneurysmal bone cysts (ABCs) are rare, representing about 1% of primary bone tumors, and 15% of all primary spine/sacral tumors. Notably, when they are located in poorly accessible regions such as the spine and pelvis, their management may be challenging. Treatment options include selective arterial embolization (SAE), curettage, *en bloc* excision with reconstruction, and radiotherapy.

Case Description: A 16-year-old male presented with 2 months of mid back pain, left-sided thoracic radiculopathy, and left lower limb weakness (MRC – 3/5). MR imaging revealed an expansile, lytic lesion involving the T9 vertebral body, and the left-sided posterior elements resulting in cord compression. He underwent SAE followed by intralesional excision, bone grafting, and a cage – instrumented fusion. ABC was diagnosed from the biopsy sample. Postoperatively, the pain was reduced, and he was neurologically intact. Five months later, he presented with a new lesion that was treated with repeated SAE and three doses of zoledronic acid. At the end of 2 years, the subsequent, MRI and CT studies documented new bone formation in the lytic areas, with healing of lesion; additionally, he clinically demonstrated sustained pain relief.

Conclusion: Here, we emphasized the importance of surgery for patients with ABC who develop focal neurological deficits. Treatment options should include SAE with bisphosphonate therapy for lesions that recur without neurological involvement.

Keywords: Aneurysmal bone cyst, Bisphosphonate therapy, Recurrence, Selective arterial embolization, Surgical excision

INTRODUCTION

Aneurysmal bone cysts (ABCs) are rare, locally aggressive lesions that occur most frequently in the first or second decades of life.[18] They are associated with genetic alterations causing activation of the USP6 gene located at 17p13.[17,22] These are pseudotumoral hyperemic-hemorrhagic lesions that constitute about 1% of all primary bone tumors[5] but 15% of all primary spine/sacral tumors.[19] CT imaging typically shows an expansile, lytic lesion with thin cortices and septae while axial MRI studies document contrast enhancement with edema and "fluid-fluid levels."[15] Clinically, patients present with pain, spinal cord compression, pathological fractures, instability, and neurologic deficits.[16] Treatment options include selective arterial embolization (SAE), direct intralesional injection, intralesional excision (curettage) with

or without fusion, *en bloc* excision/reconstruction, and/or radiotherapy. Here, we report a 16-year-old with left lower limb weakness and a T9 lytic lesion that was first treated surgically; when it recurred, it was managed with SAE and bisphosphonate therapies.

CASE PRESENTATION

A 16-year-old male presented with 2 months of mid back pain and left-sided radiculopathy with the acute onset of the left lower limb monoparesis (MRC – 3/5). Other accompanying neurological findings included a T10 sensory level with loss of pin prick, temperature, vibration/position appreciation with hyperreflexia, and a left-sided positive Babinski response. The computed tomography (CT) scan of the thoracic spine demonstrated an expansile lytic lesion with the classical "egg shell layer" occupying the left side of T9 vertebral body destroying the lamina and pedicle with epidural extension [Figure 1a and b]. The MR showed a bony, cystic mass with internal septation and fluid-fluid levels within the T9 vertebral body; on T1- and T2-weighted images, the lesion was heterogeneous with evident spinal cord compression [Figure 2a-d].

SAE and surgery

The patient had SAE; intercostal feeders were embolized using coils and gel foam [Figure 3a-c]. Within 48 h, he underwent a single stage posterior spinal decompression, with left-sided complete intralesional excision [Figure 4a]. This was accompanied by bone grafting/and cage placement for anterior column reconstruction; posteriorly, an instrumented fusion was performed utilizing titanium screws/rods from T7 to T11 [Figure 4b and c].

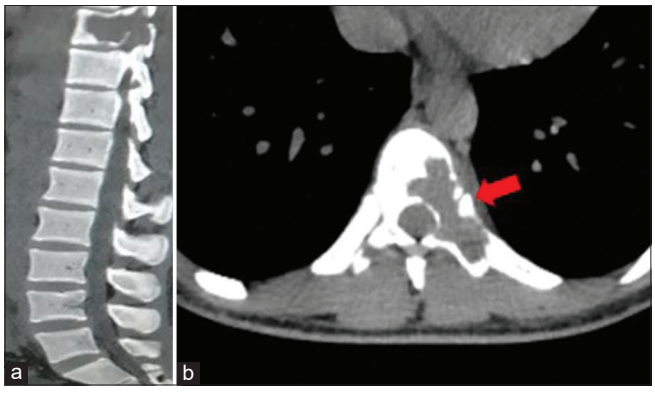

Figure 1: Computed tomography of the spine. (a) Sagittal section showing typical expansile, osteolytic bony destruction of T9 vertebra with posterior epidural extension and cord compression. (b) Axial section showing lytic lesion with egg shell layer (marked with arrow) of the T9 body involving the left pedicle, lamina, and spinous process.

Pathology/histopathology

Grossly, the T9 tumor was a gray-red 3–4 cm fleshy mass containing multiple blood-filled cysts. Histopathological examination showed cavernous spaces filled with blood surrounded by fibrous septa with marked cellular proliferation of band fibroblasts, few spindle cells, and scattered giant cells consistent with the diagnosis of an ABC [Figure 5].

Postoperative course

The pain was reduced, and his deficits improved significantly. Within 3 months, he was walking independently and performing routine activities.

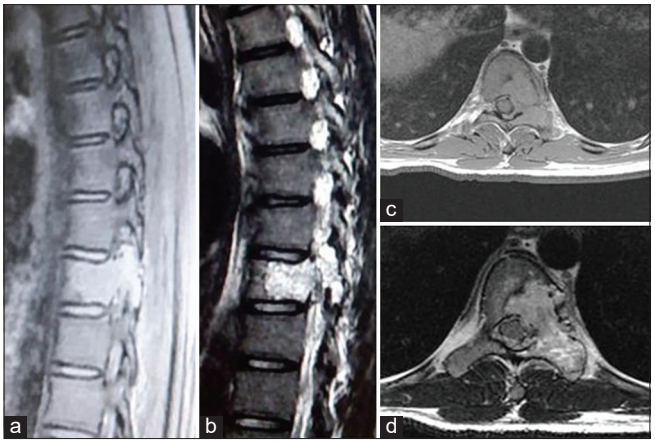

Figure 2: MRI of the spine. (a and b) T1- and T2-weighted sagittal image revealed a heterogeneous bony cystic mass with internal septation and fluid-fluid levels at the T9 vertebra. (c and d) Axial T1- and T2-weighted images showing large, expansile spinal lesion with multiple fluid levels typical for an aneurysmal bone cyst and cord compression.

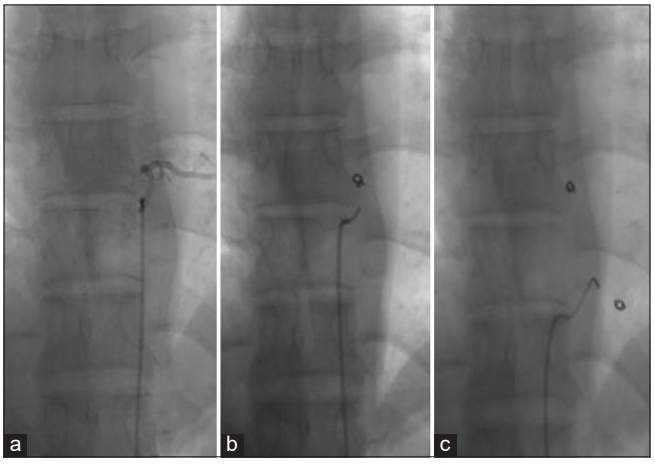

Figure 3: Preoperative embolization. (a) Angiography showing intercostal feeder vessel. (b) Intercostal feeder vessel from T8 vertebra blocked with coil, also showing owl eyed (absent left pedicle) T9 vertebra below. (c) Intercostal feeder vessel to the left T9 vertebra blocked with coil.

Lesion recurrence

At the 5th postoperative months, he presented with a new right-sided thoracic radiculopathy without any focal neurological deficits. The repeat CT scan showed recurrence of the lesion; it now involved the right side of the T9 vertebral body [Figure 6a and b]. He underwent SAE alone, following which his radicular pain markedly improved. Subsequently, he was also treated with bisphosphonate therapy (intravenous zoledronic acid) at a dose of 0.04 mg/kg every 4 months intervals, for 1 year (3 doses). At 2 years follow-up with MRI and CT studies, there was complete bone formation within the lytic areas [Figure 7a and b] and good pain relief.

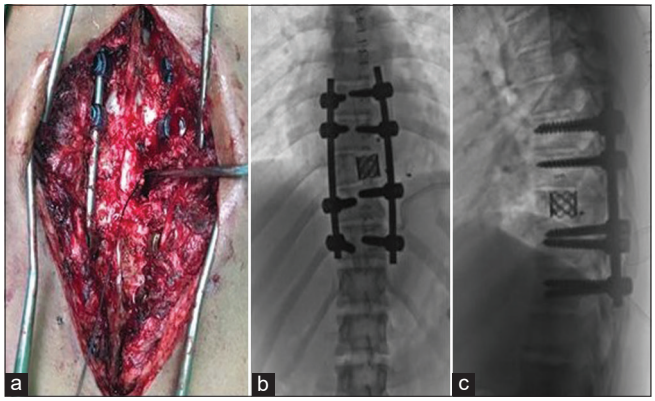

Figure 4: (a) Intraoperative picture showing cord decompression left side excision of lesion. (b and c) Postoperative thoracic spine X-ray AP view and lateral view showing posterior spinal decompression and instrumented fusion T7-T11 with anterior cage reconstruction of T9 vertebra.

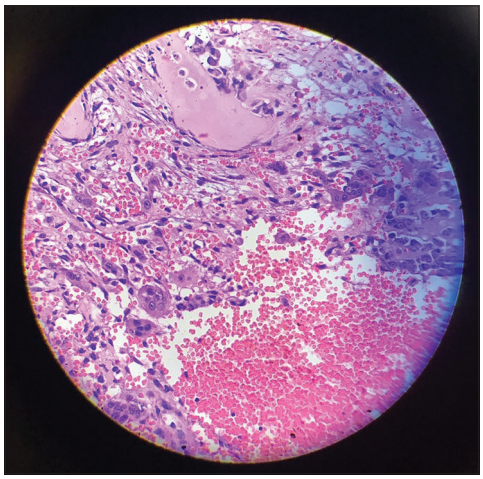

Figure 5: Cavernous spaces filled with blood surrounded by fibrous septa composed of a cellular proliferation of band fibroblasts, few spindle cells, and scattered giant cells.

DISCUSSION

ABCs predominantly occur at the second decade and have a slight preponderance for women.[7] In the spine, lumbar involvement (34%) is followed by thoracic spine (32%) and cervical spine.[12] ABC arises from posterior elements of a vertebra and later involves the pedicles and vertebral body; later on, there is intraspinal extension with resultant neurological deficits. Although most cases involve small lesions and only one spinal level, they may spread to another vertebra/rib.[2,6] In this case, the patient originally presented with – back pain and left thoracic radiculopathy of 2 months duration followed by the acute onset of the left lower limb paresis. On CT and MRI studies, the ABC aggressively breached the medial wall of pedicle, the lamina on the left side, and the posterior cortex of the T9 vertebral body resulting in spinal cord compression. Further, the MRI

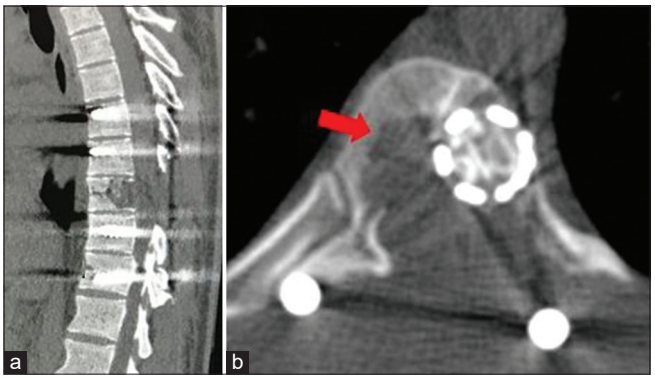

Figure 6: Computed tomography of the spine at the 5th month. (a) Sagittal section showing recurrence of lytic lesion at T9 vertebra with spinal stabilization. (b) Axial section showing lytic lesion at the right side of T9 vertebral body (marked with arrow) and cage with bone growth at previous left side lesion.

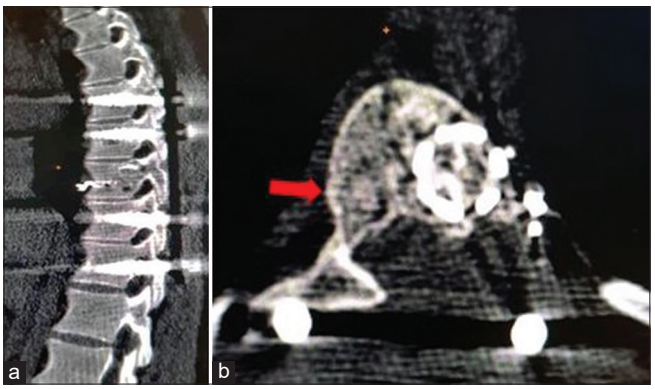

Figure 7: Computed tomography of the spine at end of 2 years. (a) Sagittal section showing complete bone T9 vertebra with intact stabilization and no signs of bony lysis. (b) Axial section showing peripheral sclerotic bone rim formation right side (marked with arrow) and bone formation inside the aneurysmal bone cysts mass.

better demonstrates epidural extension and neurological compression versus the CT study.[8] Treatment options include intralesional excision, with or without fusion,[3] *en bloc* resection, radiation therapy, intracystic injections (with osteoconductive cement,[11] doxiciclina,[10] and demineralized bone matrix with stem cells[4]), and SAE.[1]

Treatment options for primary and recurrent lesions

Tumor recurrences managed with "*en bloc*" resection, although optimal for lesion control, may not be technically feasible due to – high intraoperative and postoperative morbidity for such extensive resections. Radiation therapy, although very effective, does introduce the risk of radiation-induced sarcoma/myelopathy. SAE may be used preoperatively and/or to treat local recurrences.[1] In this case, the patient underwent SAE with coils and gel foam to decrease the vascularity of the lesion before surgery.[9] Within 48 h of the SAE, surgery was done. Most ABC recurs within 1 year and needs close follow-up. In this patient, the lesion recurred within 5 months after the index surgery; as he had no deficit, he was managed with SAE.

Adjunctive use of denosumab versus bisphosphonate therapy

Denosumab is a human monoclonal antibody that binds the cytokine receptor activator of nuclear factor-kappa B ligand,[14] this is an excellent treatment option for symptomatic ABC not amenable to surgical intervention.[21] Denosumab is, however, very costly; here, we alternatively planned for bisphosphonate therapy. The anti-inflammatory effect of zoledronic acid and the antiresorptive effect of osteoclast inhibition;[20] this allows for resolution of the bony edema and ossification of the lesion.[13] For our patient, bisphosphonate therapy was effective as documented on MR/CT studies 2 years later.

CONCLUSION

We emphasized the importance of surgery for patients with ABC who have focal neurological deficits. However, for those with recurrent lesions without specific neurological findings, SAE with bisphosphonate therapy is effective alternatives to repeated surgical intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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