

Education-Family Physician Corner

Challenges in the Treatment of Spinal Aneurysmal Bone Cysts

Badriya Toorani, BS, MBBS, CABS, FCPS* Fatema A. Abdulla, MD FFR-RCSI**
Osama Al Marzooq, MB BCH***

A nine-year-old male presented with persistent lower back pain for several weeks. Clinical examination revealed limitation of back flexion. Radiological examination revealed a lytic lesion in L4 vertebra, which was confirmed as an Aneurysmal Bone Cyst (ABC) by Fine Needle Aspiration Cytology.

The patient underwent two surgeries five months apart, which resulted in a recurrence of larger tumor discovered four months following the second surgery. A conservative approach was adopted to treat the final recurrence.

Bahrain Med Bull 2018; 40(3): 175 - 177

Aneurysmal Bone Cysts (ABC) are rare benign reactive bone lesions filled with multiple blood cavities which can be locally destructive to normal bone and neighboring soft tissue. Most ABCs occur in patients below 20 years. It occurs in the metaphysis of long bones 20% of the time (femur, tibia and proximal humerus), 25% in the spine, and 30% are associated with other tumors. Such tumors include Giant Cell Tumor, Chondroblastoma, Fibrous Dysplasia and Chondromyxoid Fibroma¹.

Clinical symptoms of spinal ABCs include local pain, swelling, muscle stiffness, and movement restriction; they are generally discovered a few months after symptom presentation. Advanced ABC's can lead to neurological deficits as the pressure exerted on the spinal cord increases¹.

The diagnosis of ABC's requires a multidisciplinary team approach which includes surgeons, radiologists, and histopathologists. The diagnosis is usually confirmed by the histopathologist and the treatment approach is determined by the anticipated prognosis². The treatment of ABCs surgically or medically had been controversial thus far³.

The aim of this report is to present a case of a spinal ABC in a 9-year-old boy, who was treated both surgically and medically.

THE CASE

A nine-year-old male patient presented with lower back pain lasting for several weeks. Upon examination, the patient showed limitation of flexion of the back; however, he was able to bend his back with a knee and hip flexion. Further radiological

investigations revealed that plain X-ray was unremarkable, CT scan, and MRI of the lumbosacral spine revealed a lesion at the L4 vertebral body compressing the neural foramen, see figure 1.

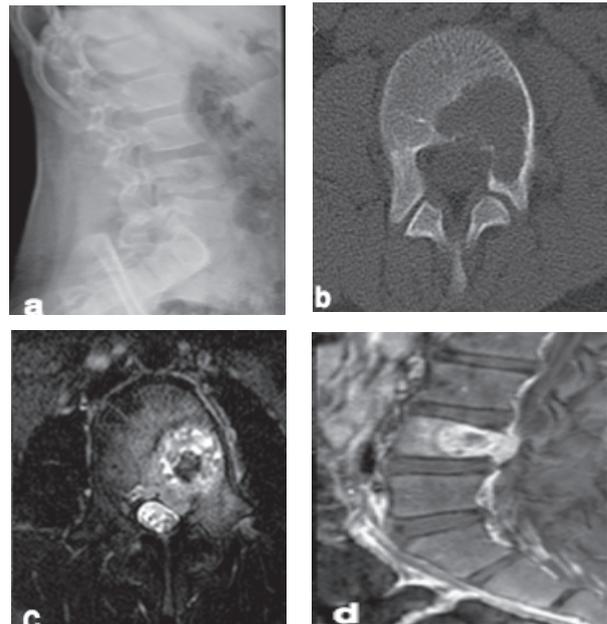


Figure 1: (a) CT Showing a Well-Defined Lytic Lesion at L4 Extending to the LT Pedicle and Focally Violating the Posterior Vertebral Wall (b) MRI Showing Multiple Small Fluid-Fluid Level with Soft Tissue Component and Mild Extension into the Epidural Space (c) Vertebral Compression Posteriorly (d) Post Contrast Enhancement

* Consultant Orthopedic, Trauma and Spine Surgery
Chairman, Department of Orthopedics
** Consultant Radiologist
Department of Radiology
*** Resident
Department of Orthopedics
Salmaniya Medical Complex
Kingdom of Bahrain
E-mail: badriyatoorani@hotmail.com

Embolization and fine needle aspiration were performed; the histopathology report was inconclusive: either Aneurysmal Bone Cyst or Osteoblastoma.

The patient presented 3 months later with pain in both calf muscles and an inability to walk long distances. Upon examination, the straight leg raise test scored 30/30, with normal lower limb power. The patient had a severe limitation of back flexion with a tilt to the right.

MRI and CT scan showed recurrence of the lesion with complete collapse of L4 vertebra and involvement of the posterior elements and cord compression, see figure 2.

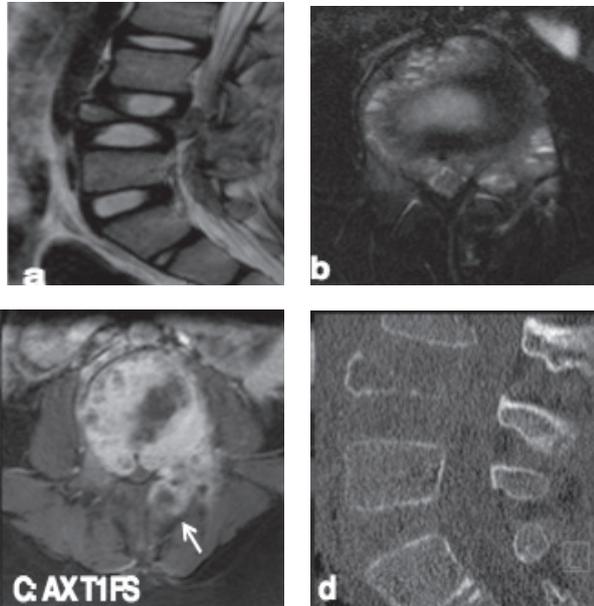


Figure 2: Complete Involvement and Collapse of L4 Vertebral Body (Vertebra Plana) (A & D) with Significant Encroachment on and Compression of the Thecal Sac (B & C). Extension of the Lesion into the Posterior Element (Arrow in C)

The patient underwent a decompression of anterior corpectomy and a posterior decompression with fixation. Two months later, the tumor recurred, and histopathology confirmed the diagnosis of Aneurysmal Bone Cyst of the spine, see figure 3. Therefore, the patient underwent revision anterior decompression with total excision of the tumor, and instrumentation with syncage, see figure 4. The patient recovered neurologically; however, he still experienced mild intermittent right lower limb pain, which was treated with analgesia.

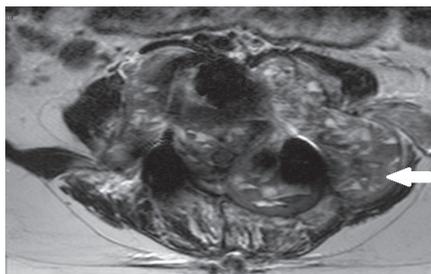


Figure 3: Recurrence of the Lesion One Month Post Operatively (Arrow)

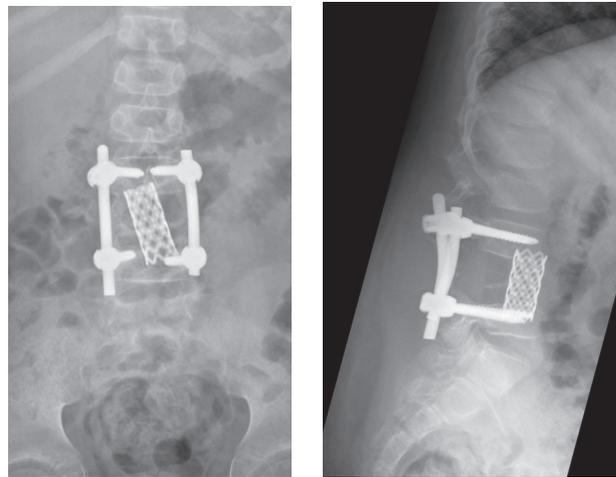


Figure 4: Second Postoperative X-rays

MRI and CT four months later revealed a second recurrence with huge paravertebral and retroperitoneal expansile lesion with multiple fluid-fluid levels significantly compromising the central canal, see figure 5.

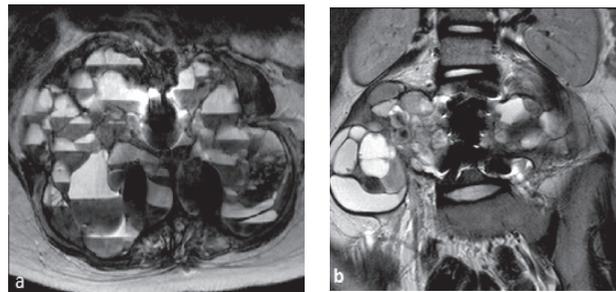


Figure 5: Axial (A) and Coronal (B) T2WI Showing Large Expansile Lesion with Multiple Fluid-Fluid Levels Displacing the Adjacent Structures and Significantly Compromising and Obliteration the Thecal Sac

The patient was informed of the treatment options, which included mesenchymal stem cell injections and Prolia (Denosumab). The guardians opted for Prolia treatment and are currently following this treatment plan overseas.

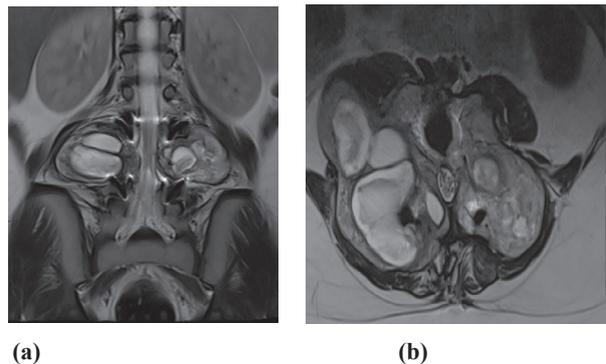


Figure 6: Regression of the Lesion Size by Approximately 40% Post-Prolia Treatment and Decompression of the Central Canal. The Lesion Though Still Having Cystic Component; However, it is Partly Solidified and Shows No Fluid-Fluid Levels

CT scan and MRI revealed shrinkage of the tumor by 40% following the treatment plan overseas fourteen months later, see figure 6. MRI was repeated five months later and showed no significant interval change in the size of the lesion.

DISCUSSION

Aneurysmal Bone Cysts are non-neoplastic expansile primary bone lesions that are uncommon in the spine. They commonly occur in long bones and flat bones. They are locally aggressive and destructive and continue to expand until treated¹. They have a high recurrence rate after excision, which presents a great challenge in achieving a cure⁴.

Treatment options include arterial embolization (either as an adjunct to surgery or as sole therapy), curettage, complete wide resection (with or without bone grafting) with stabilization of the spine, and radiation therapy⁵.

In this case, the recurrence of the tumor initiated a debate whether to have used a conservative approach prior to resorting to a surgical approach. For this particular patient, it seemed to be the consensus that conservation and observation would have been more favorable before opting for surgery. One conservative treatment plan option would include denosumab monoclonal antibody that inhibits osteoclast function by blocking the cytokine receptor activator of the nuclear factor-kappa B ligand, which results in tumor regression⁶. Another option is mesenchymal stem cell injection on bone matrix to repair bone cysts. However, this modality is still undergoing clinical trials⁷.

In 2013, a study was published regarding the use of denosumab in treating spinal ABCs; in the study, two boys (aged 8 and 11) had recurrent ABCs at C5 after surgery with intralesional tumor resection. Arterial embolization was attempted and was unsuccessful. Thus, denosumab therapy was initiated as an individualized treatment, despite the absence of scientific evidence. After the start of denosumab therapy, both patients recovered from pain and neurologic symptoms significantly and are now in a healthy condition with no severe side effects. Magnetic resonance imaging after 2 and 4 months of denosumab treatment, respectively, showed tumor regression in both patients⁸.

It still remains a controversial topic for debate whether to approach Spinal Aneurysmal Bone Cyst surgically or conservatively. So far, the most favorable treatment approach can only be determined through an individualized case-by-case investigation, with a unanimous decision by the physicians' treating team.

CONCLUSION

It still remains a controversial topic whether to approach a Spinal Aneurysmal Bone Cyst surgically or conservatively. So far, the most favorable treatment approach can only be determined through an individualized case-by-case investigation, with a unanimous decision by the treating team physicians.

Author Contribution: All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Acceptance Date: 4 July 2018.

Ethical Approval: Approved by the Department of Orthopedics, Salmaniya Medical Complex, Bahrain.

REFERENCES

1. McKean, J. (2014, August 22). Aneurysmal Bone Cyst. <http://www.orthobullets.com/pathology/8036/aneurysmal-bone-cyst> Accessed on 9 April 2016.
2. Hauschild O, Lüdemann M, Engelhardt M, et al. Aneurysmal Bone Cyst (ABC): Treatment Options and Proposal of a Follow-Up Regime. *Acta Orthop Belg* 2016; 82(3):474-483.
3. Batische F, Schmitt A, Vendevre T, et al. Aneurysmal Bone Cyst: A 19-Case Series Managed by Percutaneous Sclerotherapy. *Orthop Traumatol Surg Res* 2016; 102(2):213-6.
4. Lin PP, Brown C, Raymond AK, et al. Aneurysmal Bone Cysts Recur at Juxtaphyseal Locations in Skeletally Immature Patients. *Clin Orthop Relat Res* 2008; 466(3):722-8.
5. Tsagozis P, Brosjö O. Current Strategies for the Treatment of Aneurysmal Bone Cysts. *Orthop Rev (Pavia)* 2015; 7(4):6182.
6. Lange T, Stehling C, Fröhlich B, et al. Denosumab: A Potential New and Innovative Treatment Option for Aneurysmal Bone Cysts. *Eur Spine J* 2013; 22(6):1417-22.
7. Gourabi H, Nejad M., Emadeddin M, et al. Treatment of Bone Cyst with Bone Marrow Mesenchymal Cell Transplantation. 2011. <https://clinicaltrials.gov/ct2/show/NCT01207193> Accessed on 9 April 2016.
8. Lange T, Stehling C, Fröhlich B, et al. Denosumab: A Potential New and Innovative Treatment Option for Aneurysmal Bone Cysts. *Eur Spine J* 2013; 22(6): 1417-1422.