

Large lumbar aneurysmal bone cyst presenting as an abdominal mass. Management and outcome

Mostafa Elaskary MD, Moamen Mohamed MD.

Department of Neurosurgery, Alexandria University, Egypt.

Abstract

Background Data: Aneurysmal bone cysts (ABCs) of the spine account for 12 to 30% of all ABC cases. They mostly occur in the lumbar vertebrae, followed by the thoracic, cervical, and sacral vertebrae. Despite being benign, they can be locally expansive and destructive, and can result in pathological fractures of the vertebrae and neurological complications. Treatment modalities include surgical resection, radiotherapy, and embolization with reconstruction.

Purpose: To report on a rare case of very large aneurysmal bone cyst originating from the body and the posterior arch of L3 vertebrae and extending from the paraspinal sub-cutaneous tissue posteriorly to the anterior abdominal wall anteriorly and pushing the kidney up and explaining the way of management of such cases.

Study Design: Case report and review of literature.

Patient and Methods: An 18years old female patient presented with back pain, right lower limb numbness and partial foot drop together with a big abdominal mass. A thorough general and neurological examination proceeded by proper history taking including family history and previous trauma were done. Radiological workup was done in the form of plain X-ray, CT scan, and MRI. Total excision and reconstruction was performed under general anesthesia through a posterior only approach.

Results: Post operatively the patient recovered nicely without any neurological deficits within 2 weeks. Post operative CT showed total excision, adequate reconstruction and trans-pedicular screw fixation. This patient was followed up clinically for 8 months till now with very good neurological condition.

Conclusion: Proper diagnosis and adequate surgical techniques are mandatory for proper surgical excision of such a very big ABC especially if highly vascular. (2014ESJ083)

Key Words: Aneurysmal bone cyst (ABC), Lumbar, Large.

Introduction

Aneurysmal bone cysts (ABC) are rare, benign but locally destructive bone tumors. The term aneurysmal bone cyst was first described by Jaffe and Linchtenstien in 1992.¹ Since then, the name has been generally accepted although the lesion is neither aneurysm nor a cyst. The incidence of ABC amounts to 0.14 per 105 per year. Women are slightly more often affected than men, with a ratio of 1.04:1.

The mean age of patients affected lies in the second decade of life. The long bones of the lower extremities are affected most often, with the tibia and femur being respectively affected in 24.7% and 7.3% of all cases, followed by the upper extremities 10%, and the pelvis 9%. About 14% of all ABCs are encountered in the spine, The cervical and thoracic vertebrae are involved more commonly than the lumbar vertebrae or sacrum.^{3,16}

The etiology of ABC is uncertain; it may occur in bone as a solitary lesion or can be found in association with other bone tumors such as giant cell tumor, chondroblastoma, chondromyxoid fibroma, and fibrous dysplasia or in association with a malignant process.²⁰

Most of spinal ABCs affect the cervical region. Multiple vertebral levels is often affected and 50% occur in the pedicles, laminae and the spinous processes.^{2,4} Most patients clinically present with symptoms of diffuse pain, Low back stiffness, and swelling. Symptoms usually persist for about 12 months before definitive diagnosis.¹⁴

Case Report:

An 18 years female patient presented with low back pain and right lower limb numbness along the distribution of L4 and L5 nerve roots as well as weakness together with abdominal swelling. Abdominal examination revealed a large mass filling the renal angle and extending from the anterior to the posterior abdominal wall, firm in consistency, not mobile, not pulsating nor compressible and can't get between it and the kidney.

Neurologically the patient had partial right foot drop. She had no sphincteric disturbances. CT abdomen was performed and revealed a very large

mass, measuring about 14.0x9.0x8.5cm, cystic in nature with fleshy component surrounded by shell of bone and originating from L3 body and pedicle. MRI lumbar spine was done and confirms the CT picture as very large mass, measuring about 14.0x9.0x8.5cm, cystic in nature showing multiple fluid levels indicating multiple chronological ages of hemorrhage with fleshy component surrounded by shell of bone and originating from L3 body and pedicle. (Figure 1) The patient was prepared for surgical excision and reconstruction through a posterior only approach.

Under general anesthesia and in prone position, a midline posterior lumbar skin incision was performed. Dissection in layers on the right side was done under adequate hemostasis to decrease blood loss. Adequate exposure of a hard wall cystic lesion was done. Due to the large size of the tumor, puncture on the cyst wall was done to evacuate some of the content of the cyst to allow complete dissection to the wall, which has been done successfully.

After removal of the cyst, curettage of the body was performed, and decompressive laminectomy was performed to allow removal of the intra canalicular part then, reconstruction of the eroded pedicle and body was done with direct bone cement injection followed by pedicle screw fixation and fusion level above to and one level below the involved vertebra (Figure 1). Closure in layers was done with closed drain 14 French and sub-cuticular skin closure.

Results

Successful total removal of the ABC was achieved through posterior midline approach only with adequate reconstruction and fixation. The surgery took about 2 hours with blood loss of about 700cc. Post operatively the patient recovered smoothly with the same pre operative neurological state. The patient was discharged home on the second post operative day. Follow up visit after two weeks reveals total recovery of the pre operative neurological deficit. Post operative CT was done during this visit confirmed total excision of the ABC and adequate spinal reconstruction.

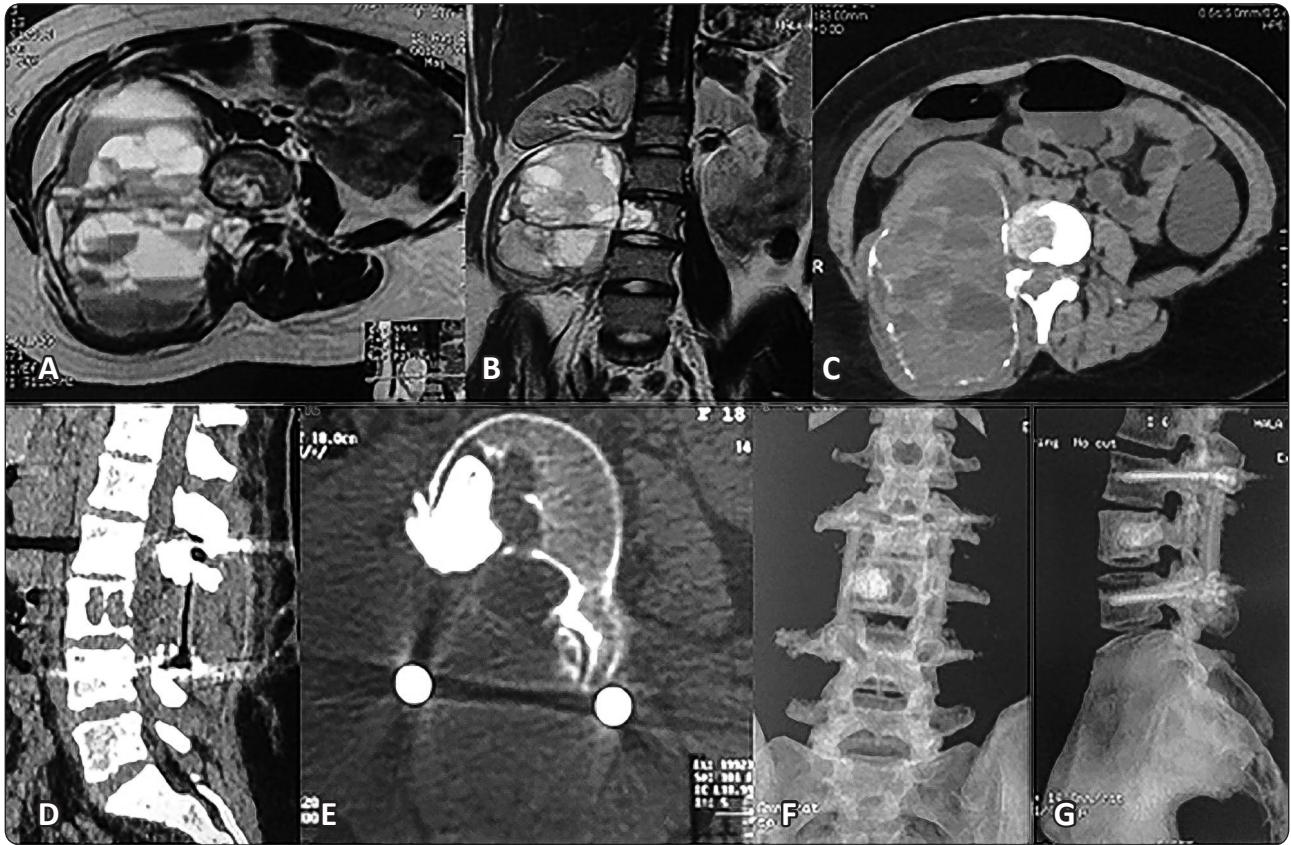


Figure 1. Preoperative T2 MRI axial (A) and coronal (B) Showing large mass measuring 14.0x9.0x8.5cm, cystic in nature with multiple fluid levels indicating multiple chronological ages of hemorrhage with fleshy component surrounded by shell of bone and originating from L3 body and pedicle. CT-scan abdomen axial cut (C) showing the same cyst surrounded by shell of bone. Postoperative CT scan sagittal (D), axial (E), and 3D AP (F) and lateral (G) showing decompressive laminectomy, removal of the intra canalicular part and reconstruction of the eroded pedicle and body with direct bone cement injection followed by pedicle screw fixation and fusion.

Discussion

The case we presented here was of an 18 year old female who manifested with some characteristics of an ABC. She presented with a very large lesion, 14.0x9.0x8.5cm in a less usual site (L3, 4 vertebrae) and a late presentation. A complete literature review shows that more than 90% of the cases presented before the age of 30, which was typical of the case we presented who was in the adolescent age.¹³ This is also proved by the study of Sherman and Soong¹⁹ which stated that ABC is a disease of adolescence with highest incidence between 10 and 19 years of age.

ABCs have some features that are sometimes misinterpreted as malignancy, as most patients present with swelling and a rapidly growing,

destructive lesion of the bone. Also, according to the report of Cser et al,⁵ 84% of the cases present with more than half of their bone destroyed at the time of recognition. ABCs typically involve long bones of the extremities, membranous bones of the thorax or vertebrae¹¹ with the cervical and thoracic vertebrae being more commonly involved than the lumbar or sacral vertebrae. Spinal lesions account for approximately only 12- 30% of cases.^{15,6}

There is a strong co-relation between the formation of an ABC and history trauma at the site of its formation and in this aspect it matches with our case presentation with history of trauma at the site of the tumor. But the exact pathogenesis of an ABC is not fully understood. The natural history of aneurysmal bone cyst has been described as evolving through 4 radiologic stages: initial, active,

stabilization and healing.¹⁰ In the initial phase, the lesion is characterized by a well-defined area of osteolysis with discrete elevation of the periosteum.

This is followed by a growth phase, in which the lesion grows rapidly with "progressive" destruction of bone and development of the characteristic "blown-out" radiographic appearance. The growth phase is succeeded by a period of stabilization, in which the characteristic "soap bubble appearance" develops as a result of maturation of the bony shell. Final healing results in progressive calcification and ossification, and the lesion transforms into a dense bony mass.¹²

Depending on its location, stage of development, and growth activity, ABCs may manifest a wide variety of patterns radiologically.^{3,7,15} Rapid disappearance of bone structure in less than 3 weeks, the apparent "blown-out" expansion of bone and the appearance of a fluid-fluid level on the CT scan and MRI are all characteristic of an ABC formation.⁸ However, "ballooned-out" eccentric lytic lesion may be radiological differential diagnosis of a giant cell tumor or atypical myeloma, giant cell reparative granulomas, hemorrhagic cysts and fibrous dysplasia as well. Thus, the location in the skeleton and the age of the patient can both play important roles in the roentgen appearance.

Taking age as an example, giant cell tumors are usually seen in an older age group, more than 30 years of age, and only 1.7-10.6% of giant cell tumors occur in children and adolescents.¹⁶ Besides, giant cell tumors have their epicenters at the end of the bone, whereas the ABC is in the metaphysis. Although ABC was formerly considered to be a variant of giant cell tumor of bone, the giant cell itself is not a constant finding. To differentiate the 2 lesions, attention should be given to the stroma. The stroma is more fibrogenic in an ABC than in a giant cell tumor.

Histologically, the most unusual feature of an ABC is the mineralized matrix with a chondroid aura. It has been reported to be peculiar to this condition.⁷ This mineralized matrix, considered by some to be osteoid, chondroid⁴ or fibromyxoid, is often heavily calcified. If present, it can be regarded as a reliable diagnostic feature of an ABC.¹⁷

ABCs, especially when they present in an aggressive manner, must be distinguished from a telangiectatic osteosarcoma.¹⁶ Although

telangiectatic osteosarcoma mimics this gross morphologic feature,¹⁷ it is not pathognomonic. Morphologically, no cellular atypism is detected, and the osteoid formation appears reactive in ABCs.

The surgical interventions were classified into 3 types by Enneking:⁹ 1) intralesional (curettage and bone grafting), 2) marginal (en bloc) resection, and 3) wide resection (segmental resection). In preoperative planning, the location and growth pattern of the ABC are decisive. However, in sites such as the pelvis or spine or when the size of the cyst is particularly large, surgical treatment of extraperiosteal excision and bone grafting become difficult and risky.⁹ Thus, careful curettage and bone grafting still remain the surgical method of choice in such cases as we did in our case except instead of bone grafting we use bone cement in reconstructing as the patient doesn't prefer taking iliac or rib bone for grafting. Although the location and size of the present case made surgery difficult, we still resected the tumor with marginal margin to reduce the possibility of recurrence.

The factors influencing recurrence are still unknown. Age, lesion location, lesion size, and number of mitotic figures have been suggested. According to Enneking⁹ more than 90% of recurrent lesions occurred in patients younger than 20 years of age.

Conclusion

Very large ABC in the lumbar vertebrae is not that common. It comes after the long bone and cervical and thoracic vertebrae. Location, size, and tumor pattern, as well as symptoms are extremely important clues to make a correct diagnosis. A variety of entities should be considered and the CT and MRI might be the most helpful tools in the differential diagnosis of ABC. Therefore, it is essential to keep such possibilities in mind, especially when the clinical feature is indistinct.

References

1. Barnes R: Aneurysmal bone cyst. *The Journal of Bone and Joint Surgery* 38B: 301-311, 1956
2. Bertoni F, Bacchini P, Capanna R: Solid variant of aneurysmal bone cyst. *Cancer* 71: 729- 734, 1993
3. Campanacci M, Capanna R, Picci P: Unicameral

- and aneurysmal bone cysts. *Clin Orthop* 204:25-36, 1986
4. Capanna R, Albisinni U, Picci P, Caderoni P, Campanacci M, Springfield DS: Aneurysmal bone cyst of the spine. *The Journal of Bone and Joint Surgery* 67-A N4: 527-531, 1985
 5. Cser MSI, Kónya A, Rényi-Vámos A: Aneurysmal Bone Cyst: A review of 52 primary and 16 secondary cases. *Arch Orthop Trauma Surg* 111:318-22, 1992
 6. Dahlin DC, McLeod RA: Aneurysmal bone cyst and other non-neoplastic conditions. *Skeletal Radiol* 243-50, 1982
 7. Dahlin DC, Unni KK: Bone tumors: general aspects and data on 8542 Cases, ed.4. Springfield, IL: Charles C. Thomas 420-30, 1986
 8. Diercks RL, Sauter AJM, Mallens WMC: Aneurysmal bone cyst in association with fibrous dysplasia: A case report. *J Bone Joint Surg (Br)* 68:144-6, 1986
 9. Enneking WF: *Musculoskeletal Tumor Surgery*. New York: Churchill Livingstone 1530, 1983
 10. Gold RH, Mirra JM: Case report 234: Aneurysmal bone cyst of left scapula with intramural calcified chondroid. *Skeletal Radiol* 10:57-60, 1983
 11. Hrishikesh KA, Narlawar RS, Deasi SB, Aniruddha K, Maheshwari S: Aneurysmal bone cyst of the ethmoid bone. *Br J Radiol* 75:916-8, 2002
 12. Kransdorf MJ, Sweet DE: Aneurysmal bone cyst: Concept, controversy, clinical presentation and imaging. *Am J Roentgenol* 164:573-80, 1995
 13. Leithner A, Windhager R, Lang S, Haas OA, Kainberger F, Kotz R: Aneurysmal bone cyst: a Population based epidemiologic study and literature review. *Clin Orthop* 363:176-9, 1999
 14. Meyers SP: MRI of bone and soft tissue tumors and tumor like lesions, differential diagnosis and atlas. Thieme Publishing Group ISBN 31:313-322, 2008
 15. Mirra JM, Picci P, Gold RH: *Bone tumors: Clinical, Radiologic, and Pathologic Correlations*, Philadelphia: Lea & Febiger 1267-309, 1989
 16. Randall RL, Nork SE, James, PJ: Aggressive Aneurysmal Bone Cyst of the Proximal Humerus. *Clin Orthop* 370: 212-8, 2000
 17. Schütte HE, Taconis WK: Giant cell tumor in children and adolescents. *Skeletal Radiol* 22:173-6, 1993
 18. Scully SP, Temple HT, O'keefe RJ, Gebhardt MC: Case report 830. Aneurysmal bone cyst. *Skeletal Radiol* 23:157-60, 1994
 19. Sherman RS, Soong KY: Aneurysmal bone cyst: its roentgen diagnosis. *Radiology* 68:54-64, 1957
 20. Topouchian V, Mazda K, Hamze B: Aneurysmal bone cysts in children: complications of fibrosing agent injection. *Radiology* 232 (2): 522-6, 2004

Address reprint
request to:

Mostafa Elaskary, MD

Neurosurgical Department, Faculty of Medicine, Alexandria University, Alexandria, Egypt
Email: d.elaskary@gmail.com

الملخص العربي

كيس أم الدم الضخم بالمنطقة القطنية ككتلة بالبطن: الإدارة والنتائج تقدمية

المقدمة: يمثل كيس ام الدم بعظام العمود الفقري حوالي من ١٢ الي ٣٠% من كيس ام الدم العظمي. يتواجد كيس ام الدم بعظام العمود الفقري بصورة اكبر في الفقرات القطنية يليها الفقرات الظهرية ثم تاتي الفقرات العنقيه والعجزية. بالرغم من ان كيس ام الدم بعظام العمود الفقري يعد من الاورام الحميدة الا انه قد يؤدي الي تاكل جسم الفقره واضعافها مما قد يؤدي الي انضغاط جسم الفقره وضغط علي الحبل الشوكي والاعصاب الطرفيه مما قد يسبب شلل في الاعصاب المضغوطة ومن هنا تاتي اهمية التشخيص المبكر له وكيفيه التعامل مع مثل هذه الحالات. الغرض من دراسته: هو عرض حاله نادره من حيث الحجم الغير مسبوق وكيفيه التعامل مع مثل هذه الحالات ومتابعتها لتحقيق افضل النتائج الجراحية.

خطة الدراسة: عرض حاله وتقريرها.

طريقة البحث: حاله لسيدة تبلغ من العمر ١٨ سنه تعاني من توروم ممتد من جدار البطن الامامي الي العمود الفقري بالخلف مع ضعف بسيط بالطرف السفلي الايمن. وتم توقيع الكشف الطبي عليها واجراء الفحوصات الطبيه والتي اظهرت كيس ام الدم خارج من جسم الفقره الثالثه القطنيه وممتد بحجم كبير للامام وللخلف ورافع الكلي لاعلي. تم اجراء الجراحه من الخلف فقط وتم استئصال الكيس بالكامل وتم حقن جسم الفقره باسمنت العظم لتقويه جسم الفقره .

النتائج: تم اجراء الجراحه واستئصال الورم الكيسي بالكامل وتم كحت عظام جسم الفقره الثالثه وحقن جسم الفقره باسمنت عظم وتثبيت الفقرتين الثانيه والرابعه القطنيه بمسامير وشرائح لتثبيت الفقرات حتي يتم التئام العظام. وتحسنت حاله الاعصاب بصورة ملحوظه بعد الجراحه ورفع الضغط عن الاعصاب.

الاستنتاج: يعد كيس ام الدم بعظام العمود الفقري من الاورام الحميده ولكن اذا تاخر تشخيصها وبقيت حتي وصولها لهذا الحجم الكبير وحدوث تاكل في العظام يؤدي هذ التاخر الي ضعف في الاعصاب قد يصل الي الشلل وهذ يعطي هذه الحالات صعوبه في علاجها واستشفاء الاعصاب بعد رفع الضغط عنها لذا نعرض في تقرير هذه الحاله طرق التشخيص وكيفيه العلاج والمتابعه لهذه الحالات.