Case Report

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

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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 18 years 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.

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. 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.

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.

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. Most patients clinically present with symptoms of diffuse pain, Low back stiffness, and swelling. Symptoms usually persist for about 12 months before definitive diagnosis.

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 canicular 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.
**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. This is also proved by the study of Sherman and Soong 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, 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 vertebrae 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.

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 ay 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. 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.

Depending on its location, stage of development, and growth activity, ABCs may manifest a wide variety of patterns radiologically. 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. 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. 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. This mineralized matrix, considered by some to be osteoid, chondroid or fibromyxoid, is often heavily calcified. If present, it can be regarded as a reliable diagnostic feature of an ABC.

ABCs, especially when they present in an aggressive manner, must be distinguished from a telangiectatic osteosarcoma. Telangiectatic osteosarcoma mimics this gross morphologic feature, 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: 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. 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 Enneking more than 90% of recurrent lesions occurred in patients younger than 20 years of age.

Conclusion

Very large ABC in the lumber 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


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

المقدمة: يمثل كيس أم الدم بعض الأفكار الفقيرة حوالي من 1-3% من كيس أم الدم العظمي. يتواجد كيس أم الدم ببعض العظام الفقرية بطرق أكبر في الفقاطة خليلا للفقرات الظهرية ثم تأتي الفقرات العنقية والعجزية. بالرغم من أن كيس أم الدم بعض العظام الفقرية يعد من الزمر الحميده إلا أنه قد يؤدي إلى تأكد جسم الفقرة اضعافها مما قد يؤدي إلى انضغاط جسم الفقرة وضغط على الحبل الشوكي والاعصاب الطرفية مما قد يسبب شلل في الأعصاب المضغوطه ومن هنا تأتي أهمية التشخيص المبكر وكيفية التعامل مع مثل هذه الحالات.

الفقر من الدراسة: هو عرض حالة نادرة من حيث الحجم الغير مسبوق وكيفية التعامل مع مثل هذه الحالات ومتآبعتها لتحقيق أفضل النتائج الجراحية.

خطة الدراسة: عرض حالة وتقديرها.

طريقة البحث: حالة نادرة تبلغ من العمر 18 سنة تعاني من تورم ممتد من جدار البطن الامامي إلى العظام العظمي بالخلف مع ضعف في الحركة العضلية السفلية. وتتم توقيع الخشايط الطبي عليها وإجراء الفحوصات الطبيه والتي أظهرت كيس أم الدم خارج من جسم الفقرة الثالثة القطنية وممتدة بحجم كبير للجسم وآلت وراء الكلي لعلي. تم إجراء الدراسة من الخلف فقط وتم استئصال الكيس بالكامل وتم حفظ جسم الفقرة باستثناء العظام لتقوية جسم الفقره.

النتائج: تم إجراء الجراحة واستئصال الورم الكسي بالكامل وتم حفظ عظام جسم الفقرة الثالثة وحفر جسم الفقره باستثناء عظام وثبيت الفقرتين الثانية والرابعة القطنية بمسامير وشوات السرة لثبيت الفقرات حيث يتم التمثيل العظام.

وتحتنت حالة الأعصاب بتصوير ملمعية بعد الجراحة ورفع الضغط عن الأعصاب.

الاستنتاج: يعد كيس أم الدم بعض العظام الفقرية من الزمر الحميده ولكن إذا تأخر تشخيصها وثبت تحيز ولا يدوي هذا الحجم الكبير ووجود كي في العظام يدوي هذا التأخر يضر في الأعصاب قد يصل إلى الشلل وهذا يعطي هذه الحالات صعوبة في علاجها واستشيف العناصر بعد رفع الضغط عنها لذا تعرض في تقرير هذه الحالة طرق التشخيص وكيفية العلاج والمتآبعته لهذه الحالات.