

Case Report

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Teng-Le Huang^{1,3}

Wei-Ming Chen^{1,3}

Winby York-Kwan Chen^{2,3}

Tain-Hsiung Chen^{1,3}

¹ Department of Orthopaedics and Traumatology;

² Department of Pathology, Taipei Veterans General Hospital;

³ Department of Surgery, National Yang-Ming University School of Medicine, Taipei, Taiwan, R.O.C.

Huge Aneurysmal Bone Cyst of Iliac Bone in a Mid-aged Female

Aneurysmal bone cyst is a rare nonneoplastic expansile osteolytic bone lesion of unknown etiology. It usually occurs in the first 2 decades of life. The most common sites are the metaphysis-epiphyseal areas of long bones or vertebrae with eccentric expansion. We present a 42-year-old female with a huge aneurysmal bone cyst of the pelvis with dumbbell-shaped expansion on the both side of the iliac bone, which grew rapidly in 6 months. We also review the literature and discuss its prevalence, clinicopathologic characteristics, differential diagnostic problems, optimal treatment, and the potential of recurrence.

Key Words

aneurysmal;

bone Cyst;

ilium

Aneurysmal bone cyst (ABC) is usually an eccentric osteolytic lesion of bone. The name was first used by Jaffe and Lichtenstein (1950)^{1,2} to separate this tumor-like lesion of the bone from other bone cysts and the giant-cell tumors on the basis of its distinct clinicopathologic appearance. Its typical histological findings are blood-filled spaces surrounded by connective tissue septa, rich giant cells and newly formed bone trabeculae.¹⁻³ It can be a primary bone lesion or a secondary lesion arising from other osseous conditions.⁴ The prevalence represents approximately 1% of primary bone tumors.⁵ In about 80% of cases, the lesion involves the metaphysis of a long bone or the posterior elements of a vertebra.^{1,6} According to the Mayo Clinic's experience, the vast majority (about 80%) of patients with ABC are younger than 20 years of age.⁷ Iliac ABC has rarely been reported to occur in patients with relative old age. We herein describe a case of a huge ABC occurring in the 5th decade of age, which grew rapidly in the iliac bone.

CASE REPORT

A 42-year-old female was seen in March of 2002,

complaining of pain over her left inguinal area off and on for 2 years. The patient started to notice that palpable mass over her left upper hip area 6 months before, and it became gradually larger and firmer for 3 months before she attended to our institute. The physical examination revealed a palpable, non-movable and hard mass over the area of left anterior superior iliac spine (ASIS). There was no local heat or erythema. Motor and sensory examinations of the lower extremities were normal. Rectal examination revealed a surface smooth hard mass in left pelvis. Plain radiography showed an osteolytic "blow-out" lesion outlined by a paper-thin shell of cortex over the left iliac bone (Fig. 1). Magnetic resonance image (MRI) demonstrated the presence of a 10.0 × 9.0 × 8.5 cm large cystic expansile lesion in dumb-bell shape at the both sides of left iliac bone with some destruction of the bone (Fig. 2). Well-demarcated fluid-fluid level and internal hemorrhage were also noted (Fig. 3). No evidence of tumor lesion was seen on a chest computed tomography scan (CT). Tests for tumor markers were all negative.

At surgery, a huge cystic tumor in dumbbell shape was noted in the left pelvis with invasion to the iliac bone. It was rich in blood supply and with well demarcation. After the diagnosis of ABC was established by

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Correspondence to: Tain-Hsiung Chen, MD, Department of Orthopaedics and Traumatology, Taipei Veterans General Hospital, 201, Sec. 2, Shih-Pai Road, Taipei 112, Taiwan.

Fax: +886-2-2875-5482; E-mail: tlhuang@vghtpe.gov.tw

frozen section, marginal resection of the tumor was performed without damage of the neurovascular bundles and other organs. Bone wax was used to stop the oozing



Fig. 1. Preoperative radiography showed an osteolytic “blow-out” lesion outlined by a paper-thin shell of cortex over the left iliac bone.



Fig. 2. Magnetic resonance image (MRI) demonstrated the presence of a 10.0 × 9.0 × 8.5 cm large cystic expansile mass in dumb-bell shape at the both side of left iliac bone with invasion to the iliac bone.

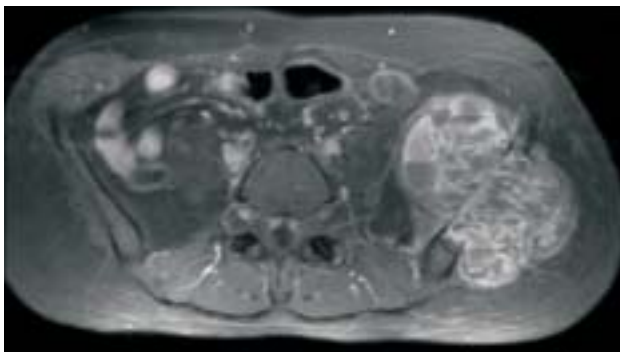


Fig. 3. Well-demarcated fluid-fluid level and internal hemorrhage are also noted on post-contrast MRI.

from bone marrow of the remaining iliac bone. The intra-operative blood loss was about 1450 mL totally.

The mass was 10.0 × 8.0 × 5.0 cm in size. The extraosseous part of the lesion was significantly larger than the intraosseous part (Fig. 4). Grossly, the specimen was cystic, with several hollow spaces of varying sizes, which were filled up with blood clots (Fig. 5). Microscopic examination revealed many septae forming blood filled channels (Fig. 6). Multinucleated giant cells, osteoid and woven bone were apparent in the septae (Fig. 7). No malignant cell was found.

She resumed full daily activity 3 months after the



Fig. 4. The extraosseous part of the lesion was significantly larger than the intraosseous, measuring up to 10.0 × 8.0 × 5.0 cm.



Fig. 5. The tumor was cystic, with several hollow spaces of varying sizes, which were filled up with blood clots.

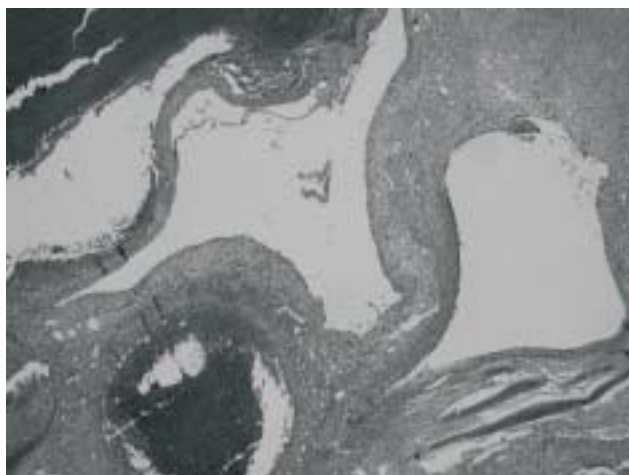


Fig. 6. Aneurysmal bone cyst, note the septae and the cystic space with blood. (Hematoxylin and Eosine. 60×).

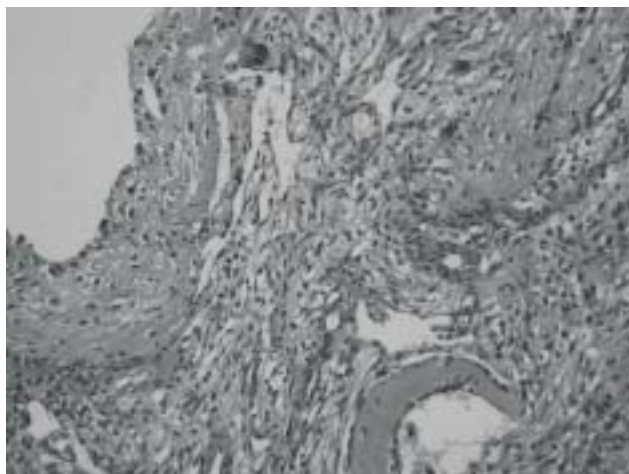


Fig. 7. Both osteoclast-typed giant cells and osteoid are identified in the septa. (Hematoxylin and Eosine. 300×).

surgery. The follow-up examinations done in 15 months, including radiography and MRI, found no recurrence.

DISCUSSION

The case we presented here manifested several unique characteristics such as relatively older age, less usual site (the iliac bone) and huge size of lesion, $10.0 \times 9.0 \times 8.5$ cm. According to Sherman and Soong,⁸ ABC is not existent in infancy and is rare among young children and older adult. It is a disease of adolescence with highest incidence between 10 and 19 years of age. A complete literature review has found more than 90% of the cases be-

fore the age of 30.³

ABCs typically involve the long bones of the extremities, membranous bones of the thorax, or vertebrae.⁹ Long bone lesions are most common in the femur and tibia, and usually eccentric and metaphyseal. Epiphyseal lesions are usually intramedullary and associated with chondroblastoma or giant cell tumor.⁵ Spinal lesions account for approximately 12-30% of cases.^{5,7} The cervical and thoracic vertebrae are involved more commonly than the lumbar vertebrae or sacrum. The pelvis is not the site of predilection for ABC and iliac wing involvement is less.⁴

Most patients present with swelling and a rapid growing, destructive lesion of bone, features that are often misinterpreted as a sign of malignancy.⁶ According to the report of Cser *et al.*¹⁰ 84% of the cases have already destroyed more than half of the bone in width at recognition. However, when a flat bone, such as the iliac wing, is involved, the usual bone expansion may not be appreciable on the routine films, and MRI may then be helpful.

The exact pathogenesis of an ABC is not fully understood. Trauma, however, is felt to be important with ABC formation. The natural history of aneurysmal bone cyst has been described as evolving through 4 radiologic stages: initial, active, stabilization and healing.¹¹ 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

Radiologically, ABC may manifest a diversity of patterns depending upon its location, stage of development, and growth activity.^{5,13,14} The radiological evidence of extremely 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 all suggest a diagnosis of ABC formation.¹⁵ However, "ballooned-out" eccentric lytic lesion may be produced by a giant cell tumor or atypical

myeloma as well.¹⁰ Furthermore, radiological differential diagnoses of such a lesion also include giant cell reparative granulomas, giant cell tumor, hemorrhagic cysts and fibrous dysplasia. 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.¹⁷ 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.¹³ 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.⁴ Occasionally, it is impossible to tell the two apart microscopically; the lesion's location, the patient's age, and its radiographic features should be helpful in these instances.⁴

ABCs, especially when they present in an aggressive manner, must be distinguished from a telangiectatic osteosarcoma.¹⁷ Although telangiectatic osteosarcoma mimics this gross morphologic feature,⁴ 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:¹⁸ (1) intralesional (curettage and bone grafting), (2) marginal (en bloc) resection, and (3) wide resection (segmental resection).¹⁷ In preoperative planning, the location and growth pattern of the ABC are decisive. It is most advantageous if the ABC grows superficially and involves no more than one-third of the bone width. These cases are suitable for intra-lesional excision to remove the cyst in normal bony tissue. This is a good intervention for ABC and results in normal joint function and few local recurrence.¹⁰ 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.¹⁸ Thus, careful curettage and bone grafting still remain the surgical method of choice in such cases.

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. Besides, according to the classification of internal hemipelvectomy,¹⁹ resection of iliac bone is classified as type 1 resection (P1 resection). Because stability of the pelvic girdle is not impaired after partial P1 resection, there is no need for reconstruction.

Recurrence occurs most commonly during the first 2 postoperative years. The factors influencing recurrence are still unknown. Age, lesion location, lesion size, and number of mitotic figures have been suggested. According to Vergel De Dios *et al.*⁴ more than 90% of recurrent lesions occurred in patients younger than 20 years of age. Patients with recurrence were in general younger than patients with ABC.⁴ The recurrence rate after wide excision approaches zero, whereas the recurrence rate with curettage is significantly higher.⁴ Campanacci *et al.*¹⁴ reported that 26% of patients with aggressive ABCs treated with curettage had a recurrence, whereas no patients treated with partial or complete resection had a recurrence.

In summary, ABC is rare in middle-aged adults. 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 MRI might be the most helpful tool in the differential diagnosis of ABC. Therefore, it is essential to keep such possibilities in mind, especially when the clinical feature is indistinct. Close observation with serial radiographic examinations for at least 2 years after surgery might be sufficient in most patients.

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