Aneurysmal bone cyst

D Basson *ND: Diagnostic Radiography; Postgraduate Certificate: Computerised Tomography (CPUT)*

Abstract: Aneurysmal bone cysts are unusual benign masses that have the potential for rapid growth, bone destruction and extension into adjacent soft tissue. Computerised tomography (CT) is useful in delineating the size and location of the intraosseous and extraosseous components of the tumor [1]. This case study is a most unusual presentation of an aneurysmal bone cyst, as only 1% of this type of tumour is found in the calcaneum. The most usual presentation in the calcaneum being that of a simple (unicameral) bone cyst or a lipoma. This article discusses the aetiology, radiological features, symptoms and preferred examinations of aneurysmal bone cysts.

Keywords: bone destruction, rapid mass growth, radiotherapy.

Case report

A 20 year old male patient presented with pain and swelling of his right foot and ankle after injuring the latter three months earlier. On clinical examination there was an abnormal bone mass with spongy swelling medially. Lateral radiograph of the left calcaneus demonstrated a visible lesion (Figure 1). The lesion was well-defined, except for the anterior aspect. The patient was referred for computed tomography (CT) of his right foot. Three millimeter axial cuts were employed and these demonstrated a large subcutaneous cystic mass of the medial aspect of the right foot, involving the underlying bony elements. The mass lesion (Figures 2 and 3) measured 4 cm x 4 cm in diameter. There was an impression of a blood fluid level in this mass (Figure 3), suspicious of an aneurysmal bone cyst with an extra-osseus component. Approximately 1% of aneurysmal bone cysts occur in the calcaneus. The patient is currently awaiting further evaluation and treatment.

Figure 1: *Radiograph of calcaneus demonstrating the bone cyst.*

Discussion

Aneurysmal bone cysts are unusual benign masses that have the potential for rapid growth, bone destruction and extension into adjacent soft tissue. The masses contain a network of multiple blood-filled cysts lined by fibroblasts and multinucleated giant cells of the osteoclast type. Although a sclerotic margin around the lesion may indicate that it is benign, soft tissue extension can make it difficult to differentiate aneurysmal bone cyst from sarcoma. CT is useful in delineating the size and location of the intra-osseous and extra-osseous components of the tumour. Magnetic resonance (MR) images typically show a lobulated or septated mass with a thin, well-defined rim of low signal intensity. The detection of a fluid-fluid level within the tumour shows the hemorrhagic nature of the cyst contents [1].

The term aneurysmal bone cyst was originated by Jaffe and Lichtenstein in 1942 to describe the radiographic appearance of a subset of unicameral bone cysts with marked expansile remodeling due to large blood-filled spaces [2].

The exact aetiology of this tumour is unknown. The descriptive name is derived from the macroscopic appearances of blood-filled, expansile, sponge-like tumour containing numerous giant cells [3]. These lesions can develop de novo or as a result of cystic changes in a pre-existing lesion such as a chondroblastoma, osteoblastoma, giant cell tumour or fibrous dysplasia [4]. A giant cell tumour is the most common cause, occurring in 19%-39% of cases and, whatever the aetiology, aneurysmal bone cyst has a close relationship to all the giant cell variants [3]. Some reports suggest that aneurysmal bone cyst is not a true neoplasm since it has been shown to arise in association with other abnormalities of the skeleton [3].

Trauma is also considered an initiating factor in the pathogenesis of some cysts in well-documented cases involving acute fracture. Local haemodynamic alterations to venous obstruction or arteriovenous fistulae that occur after an injury are important in the pathogenesis of an aneurysmal bone cyst [5].

Aneurysmal bone cysts are seen predominantly in children: 90% of these lesions occur in patients under 20 years [4]. An aneurysmal bone cyst may occur in patients between the ages of 10-30 years with a peak incidence in those aged 16 years [5]. Three quarters of aneurysmal bone cysts occur before epiphyseal fusion has commenced [3].

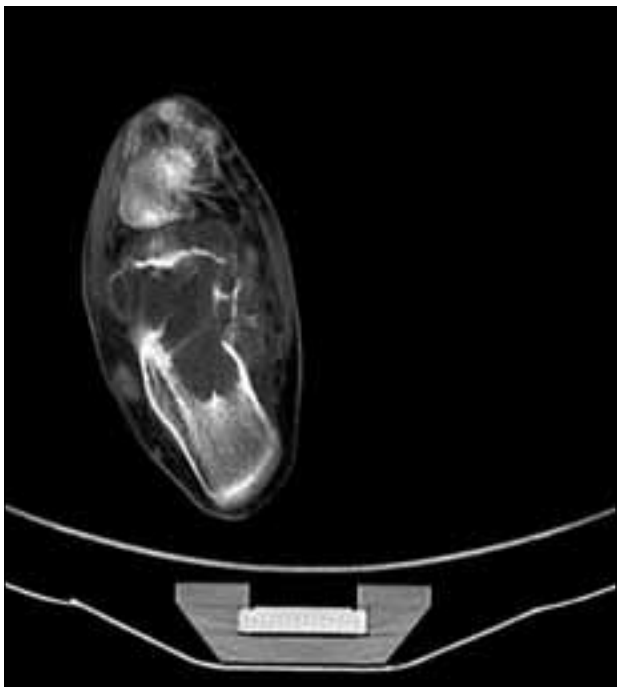

Figure 2: Axial CT image demonstrating aneurysmal bone cyst with bony involvement.

***Figure 3:** Axial CT image demonstrating aneurysmal bone cyst with blood fluid level*

The male-to-female ratio is 1.2:1 and the location is usually metaphyseal when the long bones are involved. Purely diaphyseal lesions are seen in 8% of cases. Extension into the epiphysis is even more rare [6]. Aneurysmal bone cysts have a predilection for the long bones and lumbar spine [3]. Those tumours arising in the spine occur slightly later, between 10 and 20 years. The neural arch is more commonly involved than the body, half of these cases involving more than one vertebra. The prognosis is entirely benign apart from secondary neurological lesions due to spinal canal compression [3].

Regarding the location of the lesions any bone may be affected. Approximate frequencies by site are listed below:

- Skull and mandible (4%)
- Spine (16%)
- Clavicle and ribs (5%)
- Upper extremities (21%)
- Pelvis and sacrum (12%)
- Femur (13%); (24%)
- Foot (3%)
- Lower leg (23%) [5]

The most common site is the metaphyseal region of the knee.

Short tubular bones are less frequently affected and are involved in about 10% of cases. Spinal involvement demonstrates a predeliction of the posterior elements. In decreasing order of frequency, the following parts of the spine are involved: cervical, thoracic and lumbar. Contiguous vertebrae may be involved in 25% of cases [5].

There are two types of aneurysmal bone cysts: intra and extra-osseous.

- Intra-osseous: the tumour originates in the bone marrow cavity. The tumour is primarily cystic and slowly expands into the cortex. This type of tumour is seldom related to a clinical history of trauma [7].
- Extra-osseous: post-traumatic hemorrhagic cyst. The tumor arises from the surface of bones, eroding adjacent cortex and extending into the marrow space [7].

Four phases of pathogenesis are recognized:

1. Initial phase of osteolysis.
2. Active growth phase, characterized by rapid destruction of bone and a sub-periosteal blow-out pattern.
3. Mature stage, also known as stage of stabilization, which is manifested by formation of a distinct peripheral bony shell and internal bony septae and trabeculae that produce the classic soap-bubble appearance.
4. Healing phase with progressive calcification and ossification of the cyst and its eventual transformation into a dense bony mass with an irregular structure [5].

Histologically, the lesion consists of multiple blood-filled sinusoid spaces alternating with more solid areas. The solid tissue is composed of fibrous elements containing numerous multinucleated giant cells and is richly vascular. The sinusoids have fibrous walls often containing osteoid tissue or even mature bone [4]. The clinical manifestation depends on the specific site of involvement. A common presentation includes pain of relatively acute onset that rapidly increases in severity over 6-12 weeks: the local skin temperature may increase, a palpable bony swelling may be present and movement in an adjacent joint may be restricted; spinal lesions may cause neurologic radiculopathy or quadriplegia and patients may have moderate to severe headaches [5, 7].

Diagnostic imaging plays a role in patient management. At

radiography, aneurysmal bone cysts are well-defined, cortically based, rapidly expansile lytic lesions. They can grow large enough to involve the medullary cavity, although their tendency is for eccentric expansion, a so-called blister lesion [6]. A typical area of bone resorption occurs with slight or marked expansion. The size of the lesion can vary between 2 cm and in gross examples as much as 20 cms in diameter. The overlying cortex is thinned and may be expanded to such a degree that in places it can be identified only by tomography or CT [3].

The endosteal margin is relatively well-defined against cortex, and an ill-defined zone of transition is usual between the lesion and medullary bone, occasionally with slight sclerosis [3].

Most lesions evolve slowly but extremely rapid growth is also characteristic of aneurysmal bone cysts as expansion has more to do with vascular engorgement than with cellular proliferation and aids in its differentiation from other eccentric, expansile lesions of bone [6]. A few lesions showing a highly aggressive radiologic pattern may increase alarming in size, even doubling in a few weeks [3]. The possibility of a very malignant vascular tumour, such as an angiosarcoma, may then be entertained. Aneurysmal bone cyst rarely extends to the articular surface and is often central [3].

Radiographs usually are adequate for diagnosis. The classic description of an aneurysmal bone cyst in tubular bones includes an eccentric radiolucency and a purely lytic or, occasionally, trabecular process, with its epicenter in the metaphysis of an unfused long bone. The trabeculae in the cyst may create a soap-bubble appearance in the lesion [5]. The margins of the lesion are well defined, with a smooth inner margin and a rim of bone sclerosis. The tumor does not usually extend into the epiphyseal plate until after complete fusion, when it may occasionally do so. The expansion or ballooning of the cortex occasionally may result in the loss of the sharp definition of its margin and should then be interpreted as an aggressive lesion.

New bone may horizontally traverse the angle between the original cortex and the expanded part of the bone: this occurs because the periosteum is lifted. No periosteal reaction occurs except when the periosteum is fractured [5].

Typically, the spinal lesion is osteolytic, with a predilection for the posterior elements. The lesion may involve the lamina, arches, pedicles or spinous processes, with or without extension into the vertebral body. The lesion may extend into the adjacent vertebral body, violating the intervertebral disk and causing vertebral collapse and/or extension into the spinal canal, adjacent ribs and paravertebral soft tissue [5].

CT is the most useful examination because it can demonstrate the intra and extra-osseous extent of the lesion. CT can be used to determine the nature of the matrix of the tumour, especially when tumours are in complex locations. Fluid levels may be seen in the cysts but are depicted only when the patient is lying motionless for about 10 minutes and when the scans are obtained in the plane perpendicular to that of the fluid levels. Fluid levels are not specific to aneurysmal bone cysts [5]. CT of the spine can demonstrate stenosis of the spinal canal due to involvement of the posterior elements [5].

Magnetic resonance T1-weighted images (MRI) show predominantly low to intermediate signal intensity with or without fluid levels. Acute hemorrhage into the cyst may have high signal intensity. T2-weighted images show areas of low to intermediate signal intensity or some areas of heterogeneous high signal intensity, depending on the contents of the cyst. A rim of low signal intensity with internal septa may produce a mutli-cystic appearance [5]. MRI images of aggressive lesions show tumour enhancement with gadolinium enhancement especially when they are associated with other tumours. Spinal cord compression and signal-intensity alteration in the cord can be evaluated when neurologic symptoms are present [5].

Aneurysmal bone cysts are treated by curettage or radiotherapy. The latter method being particularly valuable in spinal lesions where surgery may be considered hazardous. An increasing role, however, has developed for trans-catheter embolisation in the successful management of these tumours. Aneurysmal bone cyst is a particularly good example of the importance of radiological investigation being complete before biopsy is undertaken, since these lesions, not surprisingly, bleed considerably and the pre-operative demonstration of the vascular nature of this tumour is very important [3].

Conclusion

Although the accuracy of radiography is high for diagnosing aneurysmal bone cysts, cross-sectional CT is the most useful examination due to the fact that it can demonstrate the intra and extra-osseous extent of the lesion. CT is useful in delineating the size and location of the tumour. This facilitates recording the tumour growth rate. The extent of the disease can be estimated better with CT than with conventional radiography. CT can be used to determine the nature of the matrix of the tumour; the specificity is increased when fluid levels are depicted. Although fluid levels can also be demonstrated with MRI studies, CT still remains a fast and cost effective method of indicating the presence of fluid levels in aneurysmal bone cysts.

CT of the spine can also demonstrate stenosis of the spinal canal due to involvement of the posterior elements.

Plain radiographs can indicate the presence of aneurysmal bone cysts but CT is a most effective modality yielding enough quality information to establish the correct method of treatment.

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