Virchows Arch (1996) 428:125–129
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CASE REPORT

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Primary aneurysmal cyst of soft tissue

Report of a case with ultrastructural and MRI studies

Received: 12 September 1995 / Accepted: 18 December 1995

Abstract We report a case of primary aneurysmal cyst of soft tissues in a 57-year-old woman presenting with a painful mass in her left arm. Conventional radiography showed a radiolucent soft tissue mass surrounded by a ring of bone. MRI displayed an unusual, ill-defined soft tissue lesion that was not connected to the nearby humerus and appeared to be an aggressive tumour. Microscopically, the mass consisted of multiple anastomosing cavernous channels surrounded by a peripheral band of mature trabecular bone. These bloody channels were separated by fibrous septa containing fibroblasts, histiocytes and multinucleated giant cells, as well as fibromyxochondroid material. Some of these giant cells lined the septa and partially occupied the lumen of the channels. Ultrastructurally, the features observed in this tumour were similar to those described in aneurysmal bone cyst; the giant cells lining the septa were an additional observation. Whereas most bone tumours have a well-known extraosseous counterpart, this unique lesion is not well recognized by surgical pathologists and the few published cases have been reported under different names. Gross, microscopic, radiological and ultrastructural findings are presented to familiarize pathologists with this underdiagnosed condition.

Key words Soft tissue · Aneurysmal bone cyst · Magnetic resonance imaging

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Introduction

Aneurysmal bone cyst (ABC) is a non-neoplastic expansile bone lesion, first described by Jaffe and Lichtenstein [11] in 1942. Since the original description, many large series of ABCs have been reported [5, 6, 8–10, 12, 13, 21, 22]. This lesion occurs most frequently in children and young adults and has a predilection for the metaphysis of long bones and the vertebral column, although it can occur in any other bone [13, 21, 22]. Until the first description in the lumen of a large artery by Petrik et al. [16] in 1993, this lesion had never been reported outside the bone. We have recently reported the first primary aneurysmal cyst in soft tissues [17]. This paper presents another of these cases and stresses the ultrastructural and MRI features, which have never been described previously.

Case report

A 57-year-old woman was admitted to the hospital for evaluation and management of a 1.5-month history of swelling in her left arm that was spontaneously painful. She was treated with analgesics and anti-inflammatory drugs for 1 week, but the symptoms persisted. The patient practiced yoga and no direct traumatic antecedent to her arm pain was reported. Physical examination revealed a hard, ill-defined, moveable, 8×7.5 cm mass which was deep-seated and fixed to the underlying skeletal muscle. Routine laboratory data were within normal limits. The serum alkaline phosphatase, calcium, phosphorus and uric acid levels were normal.

Roentgenograms showed a left-sided paracortical mass close to the middle third of the humerus. The tumour, sharply circumscribed, displayed a peripheral calcified margin surrounding a radiolucent centre (Fig. 1). The lesion was considered radiologically to be myositis ossificans. MRI showed a soft tissue round mass, with poorly defined smooth edges and heterogeneous signal (hyperintense on STIR- and isointense on T1-weighted MRI sequences), along with abundant surrounding oedema, involving the long portion of the brachial triceps. The mass was clearly separated from the adjacent bony structures with no evidence of bone or neurovascular bundle infiltration. Intense heterogeneous enhancement was observed in the entire lesion (Fig. 2), which was considered to be an aggressive soft tissue mass. At surgery, the tumour was found to be included in soft tissues without contact with the

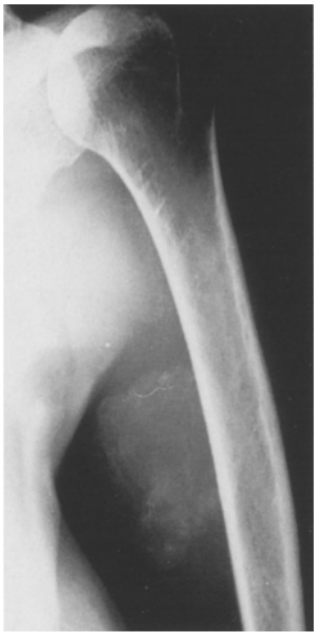


Fig. 1 Plain lateral radiograph showing a round and well-circumscribed soft tissue lesion in the left arm. Note the central radiolucency and the peripheral calcified margin

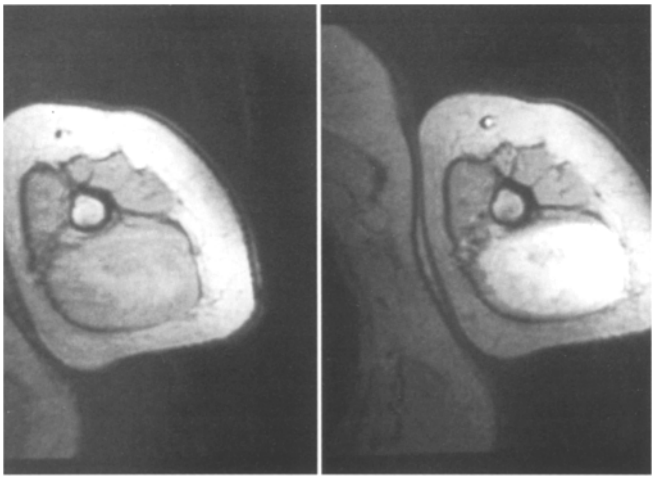


Fig. 2 *Left* Axial gradient echo slices (400/20/70°) demonstrate a soft tissue mass clearly separated from the nearby bone. Note the poorly defined hyperintense area in the centre of the mass. *Right* Same sequence, following contrast injection (Gd-DPTA), displays intense contrast enhancement, in both the centre and at the periphery of the lesion

nearby humerus. A complete resection of the mass along with the surrounding soft tissue was performed. The patient is currently well and free of disease 18 months after excision.

On gross examination, the excisional biopsy revealed a well-circumscribed 7×5×5-cm ovoid mass, well outlined and separated from the surrounding soft tissues by an eggshell-thin layer of bone. On sectioning, the tumour was composed of multiple anastomosing cavernous spaces separated by fibrous septa and filled with unclotted bloody viscous material (Fig. 3).

Microscopically, the bulk of the lesion consisted of anastomosing cavernous blood channels, lacking endothelial lining and separated by fibrous septa composed of fibroblasts, multinucleated giant cells and a lacework of osteoid trabeculae (Fig. 4). Some of these septa contained chondroid-like zones of calcification with heavily mineralized haematoxylin-staining deposits. Osteoclast-like giant cells were included in the cystic spaces and they sometimes lined the septa (Fig. 5). The peripheral ring of the tumour was made up of mature lamellar bone trabeculae with frequent osteoblastic rimming.

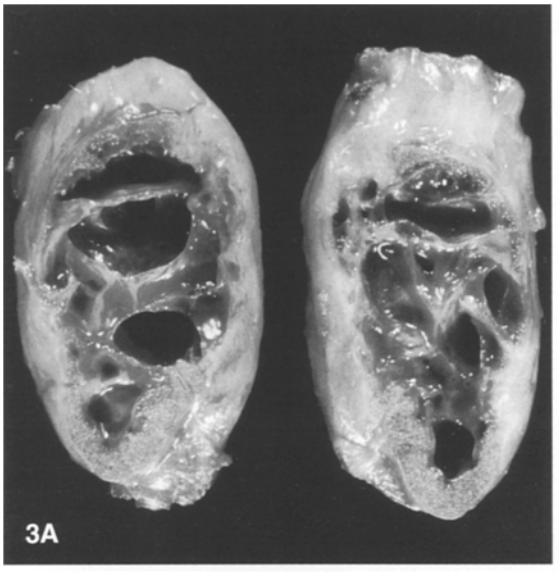
Ultrastructurally, the bulk of the lesion consisted of multiple aneurysmal blood channels, separated by septa mainly composed of fibroblasts with small Golgi apparatus and variably developed rough endoplasmic reticulum, probably representing different stages of differentiation. In addition to the fibroblasts, histiocytes, osteoblasts and giant cells, primitive mesenchymal cells could be also observed. The giant cells showed little evidence of phagocytosis and were ultrastructurally similar to osteoclasts. These giant cells were within fibrous septa or in foci lining the surface of blood-filled cavernous channels with their bases joined to the neighbouring cells by cytoplasmic interdigitations. However, no real junctions were present (Fig. 6). Nevertheless, septa were mostly lined by histiocytes and fibroblasts. Within the septa there were foci of osteoid and bone trabeculae related to metaplastic changes in spindle cell stroma with osteoblastic differentiation.

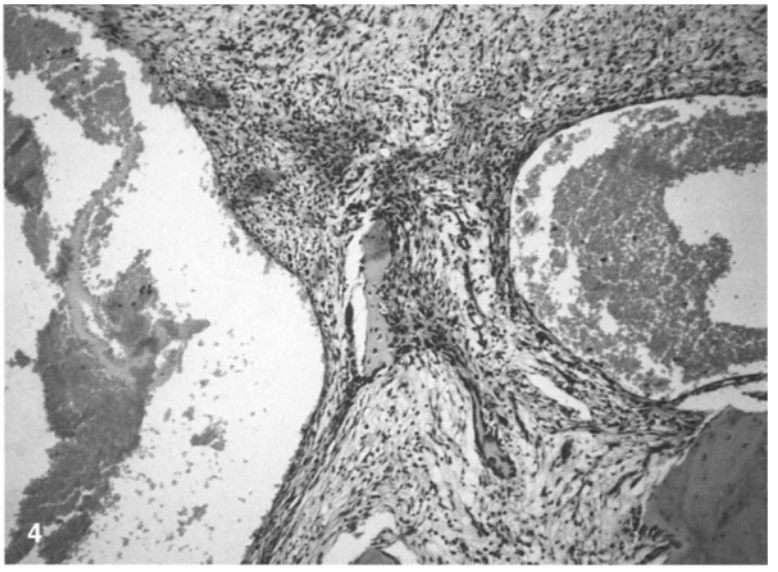
Discussion

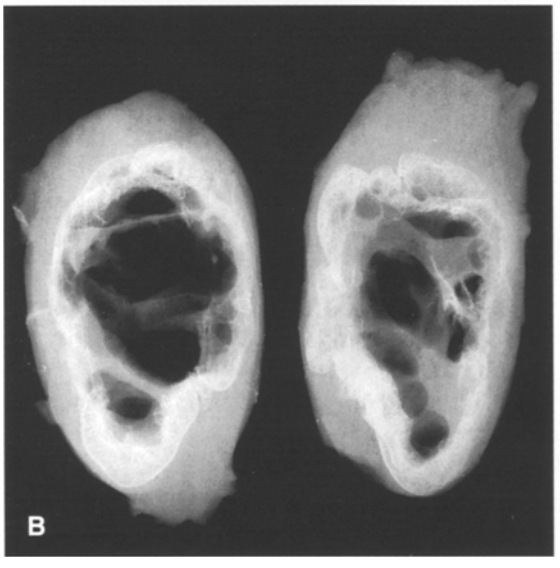
Aneurysmal bone cyst is a non-neoplastic benign lesion, predominantly occurring in children and adolescents in the metaphysis of long bones, the femur being most frequently involved [22]. Classically, this tumour was considered to be a secondary lesion arising from other bone disease. However, nowadays authors consider ABC as a distinct pathological entity [7, 8, 10, 13, 18], existing either as a primary lesion or as a secondary lesion associated with other osseous processes.

It is well known that many typical bone tumours, including osteoma, osteoblastoma, chondroma, chondroblastoma, Ewing's sarcoma, osteosarcoma and chondrosarcoma, can occur outside the skeleton, especially in soft tissues. However, reports of the soft tissue counterparts of ABC are extremely rare. As far as we know, the first reference to a soft tissue tumour similar to the present one was made by Salm and Sissons [19]. In a series of giant cell tumour or soft tissues, these authors described two cases composed of predominantly cystic and vascular spaces and suggested the name "cystic vascular tumour". Recently, Petrik et al. [16] reported an aneurysmal cyst of bone type involving a large vessel of the neck. In 1994, Rodridguez-Peralto et al. [17] coined the term "primary aneurysmal cyst of soft tissues", for the first time, in describing a lesion in the left shoulder of a 20-year-old woman. These authors found one more case in the literature that fitted the concept of true primary aneurysmal cyst, by Amir et al. [3] in the left groin of a 15-year-old and interpreted as a myositis ossificans with aneurysmal bone cyst-like changes.

The two previously reported cases [3, 17] and the present one share radiological, macroscopic and microscopic characteristics. However, the previous two cases were located near large joints, hip [3] and shoulder [17] in young patients, while this case arose in the middle third of an older woman's upper arm. All three cases were diagnosed clinically as myositis ossificans. Grossly, the three cases are similar. The masses are included in soft tissues without connection to the nearby bone struc-







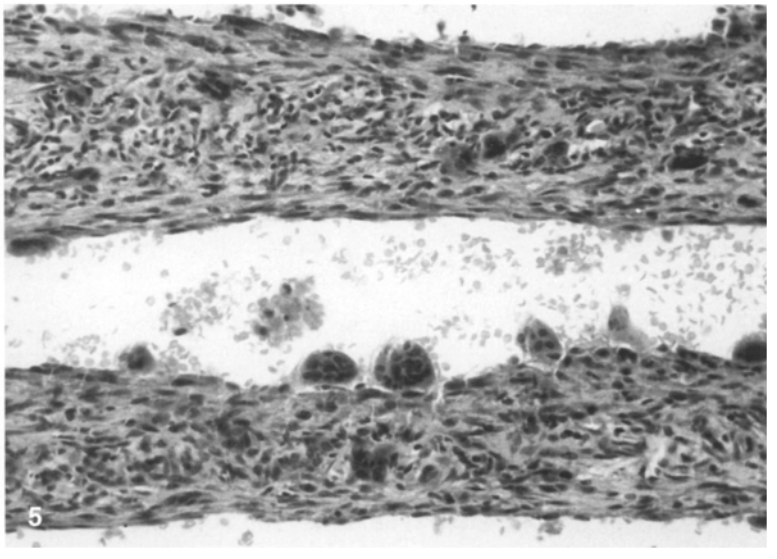


Fig. 3 **A** Cut macroscopic section of the surgical specimen. The lesion is ovoid, circumscribed and composed of multicystic anastomosing cavernous channels. **B** Roentgenographic appearance of the resected mass. Note the peripheral bone ring surrounding the lesion

Fig. 4 Panoramic view showing cavernous channels separated by fibrous septa along with peripheral bone trabeculae. Haematoxylin and eosin (H&E), ×100

Fig. 5 Fibrous septa composed of spindle and multinucleated giant cells. Some of these giant cells can be seen lining the septa and protruding into the cavernous spaces. H&E, ×250

tures, and are ovoid, well outlined and separated from the surrounding soft tissues by an eggshell-thin layer of bone. Microscopically, all three consist of anastomosing cavernous spaces full of viscous bloody material and separated by fibrous septa composed of fibroblasts, osteoclast-like giant cells, and osteoid. The whole lesion is surrounded by a ring of mature lamellar bone trabeculae. Moreover, Rodriguez-Peralto's case [17] and the present one display masses of fibromyxoid calcified material with chondroid aura. This fibromyxoid matrix is a specific diagnostic feature of ABC that unfortunately is only present in 35–52% of cases [22, 23].

There is no documented description of the ultrastructure of aneurysmal cyst of soft tissues in the two previously reported cases but the electron microscopic features of this case are similar to the findings already described in ABC [1, 2, 20]. The heterogeneity of cells observed in the septa between aneurysmal cavities support the concept of a reactive rather than a neoplastic origin for this peculiar soft tissue lesion [1, 20]. An interesting observation, not reported in ABC, is the presence of multinucleated giant cells lining the surface of blood-filled cavernous channels. However, the ultrastructural absence of endothelial, pericytic or smooth muscle differentiation rules out a vascular neoplastic or malformative source.

Earlier MRI studies of ABC have usually shown specific diagnostic features [4, 15, 22]. The constellation of an expansile lesion bordered by a thin low-signal rim,

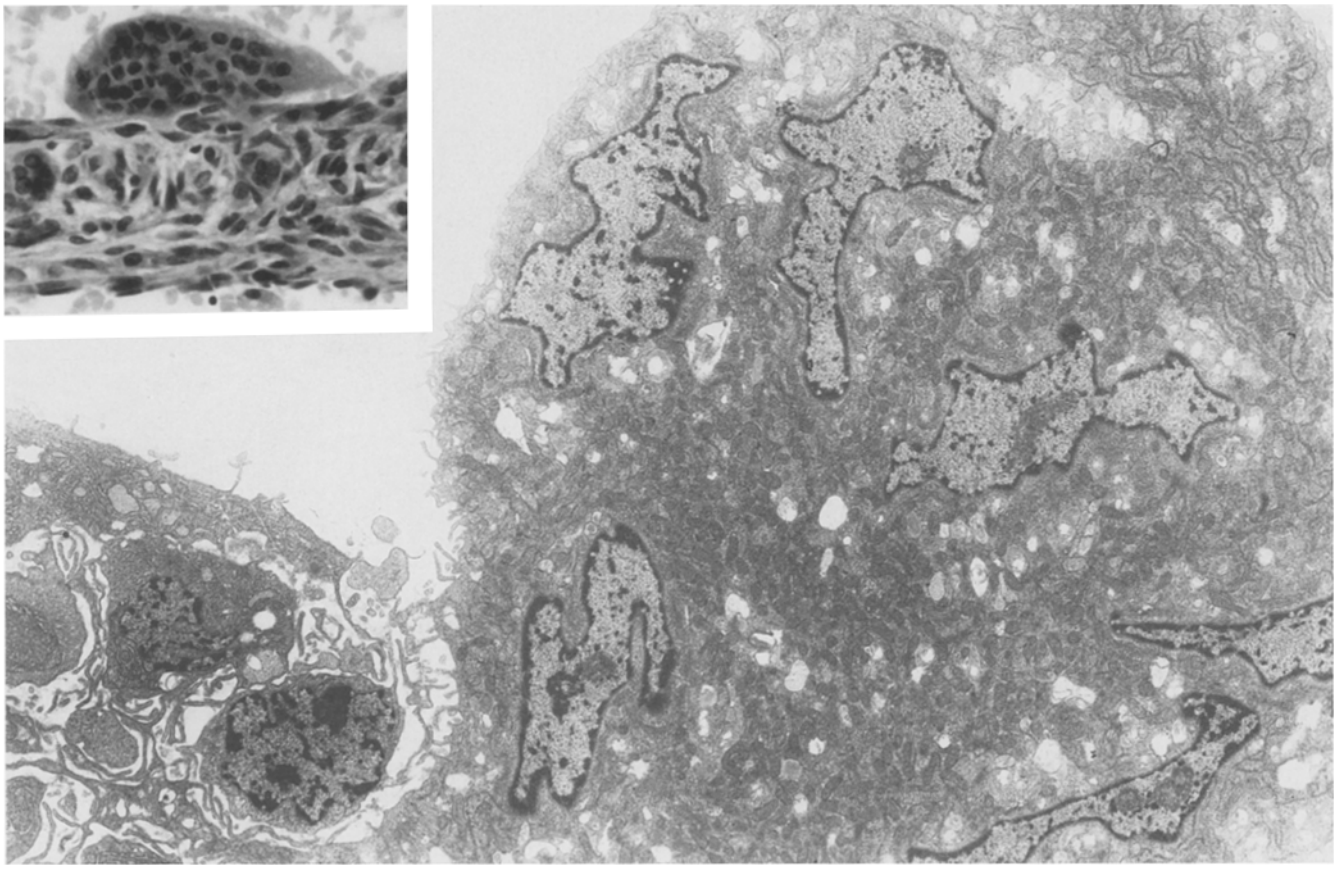


Fig. 6 Osteoclast-like giant cells lining the septa and protruding into the cavernous channel. Note the presence of interdigitations toward the neighbouring cells. Electron microscopy, ×1650. *Inset* Same features on light microscopy. H&E, ×400

the increased signal with augmented T2 weighting and a lobulated contour with fluid-fluid levels strongly suggests the diagnosis of aneurysmal bone cyst [15]. However, the present case does not display these characteristic features. Moreover, the presence in the coronal slices of a poorly defined mass with heterogeneous signal and abundant surrounding oedema may produce mislading MRI features and lead to the erroneous diagnosis of an aggressive process. Our experience in the previously reported case [17] indicates that CT is superior to MRI for the characterization and evaluation of aneurysmal cyst of soft tissues.

The nature of primary aneurysmal cyst of soft tissues is unclear. Nevertheless, Rodriguez-Peralto et al. [17] proposed a pathogenetic scheme based on the one described by Mirra [14] for ABC. This soft tissue lesion could be considered secondary to an unperceived trauma which provoked a haemorrhagic soft tissue vascular abnormality followed by a reactive florid mesenchymal proliferation. Thus, myositis ossificans and extraosseous aneurysmal cyst may be two different morphological responses to the same injury, the difference depending on the type and amount of tissue damage.

Acknowledgements This paper was supported in part by the Fondo de Investigaciones de la Seguridad Social (FISS) and Caja de Ahorros de Madrid.

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