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Aggressive aneurysmal bone cyst in association with polyostotic fibrous dysplasia: A case report

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ABSTRACT

INTRODUCTION: Aneurysmal bone cyst occurring in the setting of previously diagnosed fibrous dysplasia is rare. While both are benign processes, pain, compression of nearby structures and risk of fracture can require treatment.
PRESENTATION OF CASE: In this report, we describe a 56 year old male who developed an aggressive aneurysmal bone cyst secondary to fibrous dysplasia in the proximal tibia over a period of 8 months. He required an above knee amputation for disease and symptom control due to the aggressive nature of disease and medical comorbidities.
DISCUSSION: The diagnosis of a secondary lesion can prove difficult. It is important to exclude a malignant disease process, particularly when imaging demonstrates an aggressive appearance. In this case, repeat imaging, CT guided biopsies and an open biopsy were performed to exclude malignancy prior to definitive surgical management.
CONCLUSION: In order to exclude secondary lesions, we suggest further investigation for new onset pain in the setting of a benign lesion.

1. Introduction

Fibrous dysplasia is a benign process where normal bone is replaced with fibrous tissue. It represents 2.5% of primary bone tumours and occurs predominantly in the first 3 decades of life [1]. The gene mutation at the α-subunit of the G-protein receptor results in an increase of cAMP [2,3], causing hyperproliferation of abnormal osteoblasts as well as stimulating cytokine pathways that lead to increased bone resorption by osteoclasts. A combination of these two pathways produce the characteristic lesion. It usually affects the long bones, craniofacial bones and ribs, and can occur as a single lesion (monostotic, 70%), multiple lesions (polyostotic, 30%), or as part of McCune–Albright syndrome involving polyostotic fibrous dysplasia, café-au-lait spots and multiple endocrine dysfunction [3,4,5]. Radiologically, the appearance of fibrous dysplasia includes endosteal scalloping of the inner cortex without periosteal reaction, bony expansion and a ground-glass appearance resulting from the radiolucent bone producing no visible trabecular pattern. There is a classic histological appearance with a low to moderate cellular fibrous stroma surrounding irregular trabeculae of woven bone, arranged in a "Chinese characters" pattern [4,6,7]. In mild cases, treatment consists of surveillance and maintenance of bone density through diet, exercise, and bisphosphonates [2,3]. In severe cases, surgical reinforcement or correction may be required with internal or external fixation, sometimes with the use of cortical allografts [2].

Aneurysmal bone cysts appear as a blood-filled cavity separated by connective tissue septa with fibroblasts and osteoclast-like giant cells. They expand the affected bone, usually occurring at the metaphysis of long bones, flat bones or spinal column [7,8]. Aneurysmal bone cysts are benign, but can be locally aggressive and cause weakening of the bony structure, and expansion can cause pain, swelling, deformity, neurological symptoms and pathological fracture [9,10]. Radiologically, they appear as an eccentric expansile lesion; CT and MRI can show internal separations and fluid levels [8]. While pathogenesis is not completely understood, the development of aneurysmal bone cysts has been linked to tumour-induced vascular processes or as a consequence of trauma [6], and can occur as a secondary vascular phenomenon in areas of a previous lesion [11].

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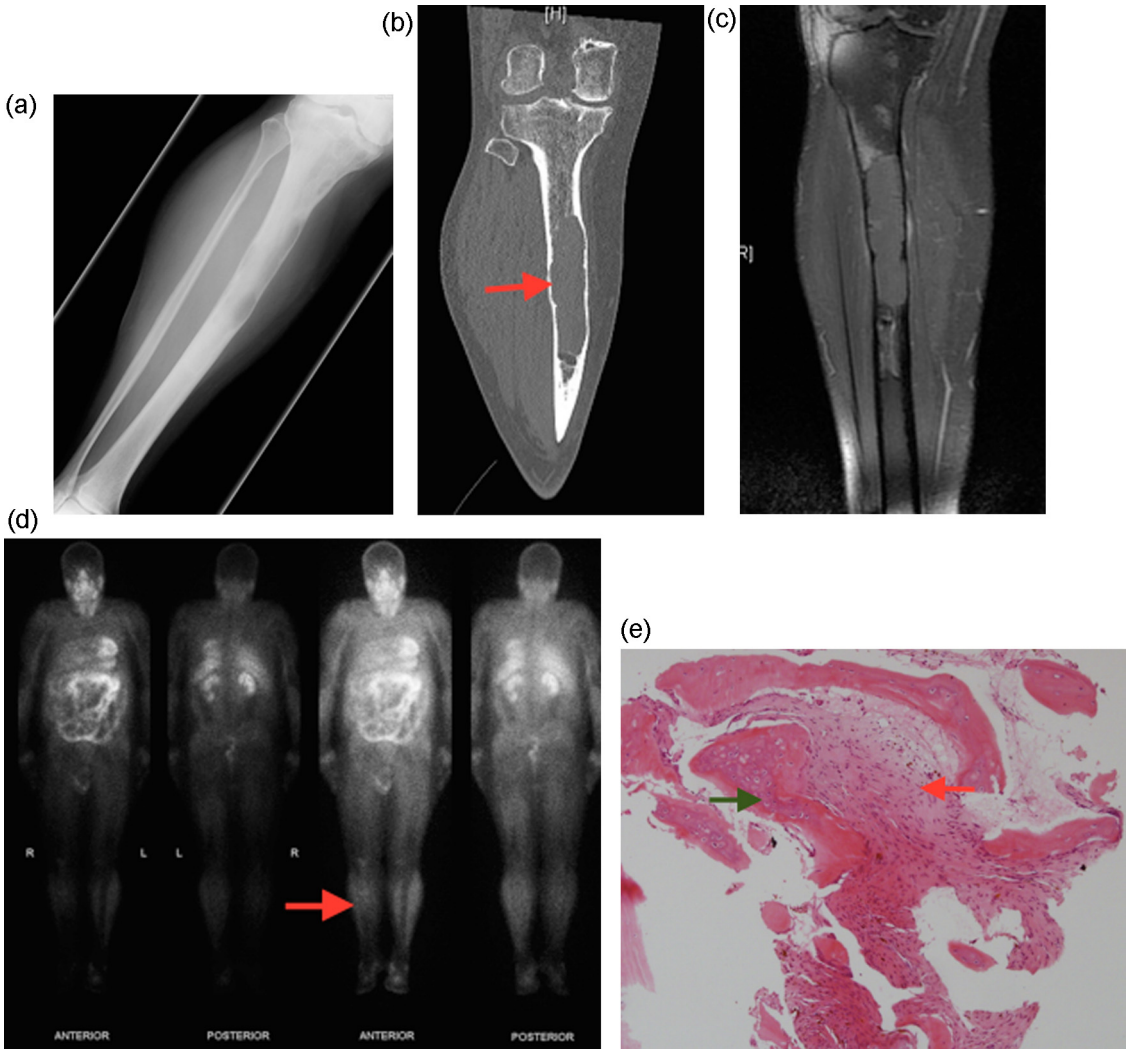

Fig. 1. Initial presentation of right tibial diaphyseal lesion:
Radiolucent lesion with medullary expansion and endosteal scalloping shown in: (A) AP radiograph; (B) Coronal CT scan showing a well circumscribed lesion with endosteal scalloping; (C) Coronal T1 with fat saturation MRI; (D) Thallium scan showing moderate heterogeneous uptake in the right proximal tibia on delayed 4 h planar imaging; (E) Core biopsy showing spindle cell proliferation (red arrow) and fibrous stroma with immature bone formation (green arrow), consistent with fibrous dysplasia.

2. Presentation of case

A 56 year old male presented with right proximal tibial pain. He had swelling and localized tenderness in his proximal leg, and required crutches due to pain; he had no skin pigmentations or neurovascular compromise. Two months prior to his presentation, a drug eluting stent had been inserted for treatment of coronary artery disease. He had been commenced on clopidogrel and aspirin.

Imaging was consistent with fibrous dysplasia in the proximal tibia; the lesion measured 15 cm in length and demonstrated endosteal scalloping, ground-glass opacity and medullary expansion without periosteal reaction (Fig. 1). A skeletal survey performed prior to referral to our hospital revealed multiple radiolucent lesions throughout both femoral and right tibial diaphysis, consistent with polyostotic fibrous dysplasia. Thallium scan, which highlights metabolic activity [6,12], showed heterogeneous mild uptake within the tibial lesion and no uptake in other lesions. Core biopsy was performed under CT guidance targeting the area of thallium avidity, and pathology showed spindle cell proliferation and fibrous stroma with immature bone formation (Fig. 1). There were no features of malignancy and the lesion was placed under surveillance after the diagnosis of fibrous dysplasia was made.

Five months later, he re-presented with increased pain in the right leg. Plain radiographs showed cortical thinning associated with a pathological fracture of the anterior cortex of the tibia and an increase in lesion size (Fig. 2). CT showed cortical breach and periosteal reaction. There was concern of malignant change with MRI features of heterogeneous T2 hyperintensity as well as medullary soft tissue expansion. Fluid-fluid levels were also noted (Fig. 2). A CT guided core biopsy was performed targeting the area of uptake on a repeat thallium scan. This showed a cellular lesion with numerous mulitnucleated giant cells together with smaller mononuclear cells, haemorrhage, fibrin and granulation tissue (Fig. 2). These features were suggestive of solid aneurysmal bone cyst, however, an open biopsy was performed due to clinical concern of malignancy. Intra-operatively it was noted that there was extensive thinning of the bony cortex and replacement of the bony architecture with haematoma. Curettage was performed and analysed, and showed a similar lesion to the previous biopsy. This was consistent with solid aneurysmal bone cyst.

Due to the anticoagulants preventing definitive surgical intervention, angiographic embolisation was attempted but was unsuccessful [13]. The patient was therefore temporarily placed into a bi-valved thermoplastic splint and commenced on zoledronic acid in an attempt to control the growth of the lesion [3,14,15]. His