Pathology – Research and Practice 213 (2017) 1445–1449

Contents lists available at ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp

Case report

Extraskeletal aneurysmal bone cyst: Report of a case and review of the literature

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ARTICLE INFO

Article history:
Received 14 April 2017
Received in revised form 15 May 2017
Accepted 2 July 2017

Keywords:
Aneurysmal bone cyst
Soft tissue
Giant cell-rich tumor

ABSTRACT

Aneurysmal bone cyst (ABC) is an expansile cystic lesion that may affect any bone of the skeleton. Although exceedingly rare, lesions with histomorphologic characteristics of an ABC have reportedly originated within soft tissue. Extraskeletal ABC may mimic a variety of benign and malignant lesions and can be confused with other more common or rare giant cell-rich tumors of soft tissue, especially myositis ossificans. Clinical, radiological and histologic correlation is crucial in reaching the correct diagnosis. Cytogenetic and/or molecular genetic analysis is a useful adjunct in diagnosing these exquisitely rare lesions. Here we report a case of an ABC arising in an extraskeletal site and provide a comprehensive review of literature on this rare entity.

1. Introduction

Aneurysmal bone cyst (ABC) is an expansile, osteolytic lesion that most commonly occurs in individuals during their second decade of life. It may affect any bone in the body but usually arises in the metaphysis of long bones. It has long been controversial as to whether ABC is neoplastic or reactive in nature. It is now generally accepted that primary ABC is a neoplastic process owing to rearrangements involving the Ubiquitin-specific peptidase 6 (USP6) gene located at chromosome 17p13 as a reproducible genetic event [1]. Although benign, the tumor can be locally aggressive and cause extensive destruction of the affected bone and impinge on the surrounding tissues. Recurrence is not uncommon. ABC may also occur secondarily to other benign or malignant bone tumors. The secondary ABC does not harbor a USP6 gene rearrangement thus is thought to be reactive in nature [1].

Although exceedingly rare, lesions with histomorphologic characteristics of an ABC have reportedly originated within soft tissue, of which some have been further confirmed to harbor a USP6 gene rearrangement [2]. Extraskeletal ABC can be confused with other benign or malignant soft tissue tumors radiologically and histologically given its rarity and resemblance to other mimickers such as myositis ossificans and giant cell-rich tumors of soft tissue, including giant cell tumor of soft tissue, tenosynovial giant cell tumor, brown tumor of hyperparathyroidism, and extraskeletal telangiectatic osteosarcoma. Herein, we report such a case and provide a comprehensive review of literature on this rare entity.

2. Case report

A 26-year-old Caucasian woman presented with pain and a mass in her proximal left thigh that had increased in size over a period of several months. Magnetic resonance images showed an intramuscular lesion in the left vastus lateralis, measuring approximately $6.6 \times 4.3 \times 3.8$ cm. The mass was T1 hypointense with peripheral and septal enhancement, and demonstrated hyperintense foci with associated fluid–fluid levels on T2-weighted images (Fig. 1). Longitudinal sonography revealed a well-circumscribed, heterogeneous, hypoechoic lesion with multiple anechoic and hypoechoic cystic cavities that contained fluid–fluid levels. The patient underwent percutaneous core-needle biopsy of that left thigh mass.

The histologic sections of the core-needle biopsy revealed a blood-containing cystic lesion. The cyst wall consisted of an admixture of spindled cells, inflammatory cells, hemosiderin-laden macrophages and osteoclastic multinucleated giant cells (Fig. 2A&B). No endothelial or epithelial lining was appreciated. No significant nuclear pleomorphism or atypical mitotic figures were noted. A surgical excision was performed following a pathologic diagnosis of ABC. The resected specimen consisted of multiple red-brown, hemorrhagic tissue fragments. Again, the lesion demonstrated blood-containing cysts that were separated by mul-

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http://dx.doi.org/10.1016/j.prp.2017.07.001
0344-0338/

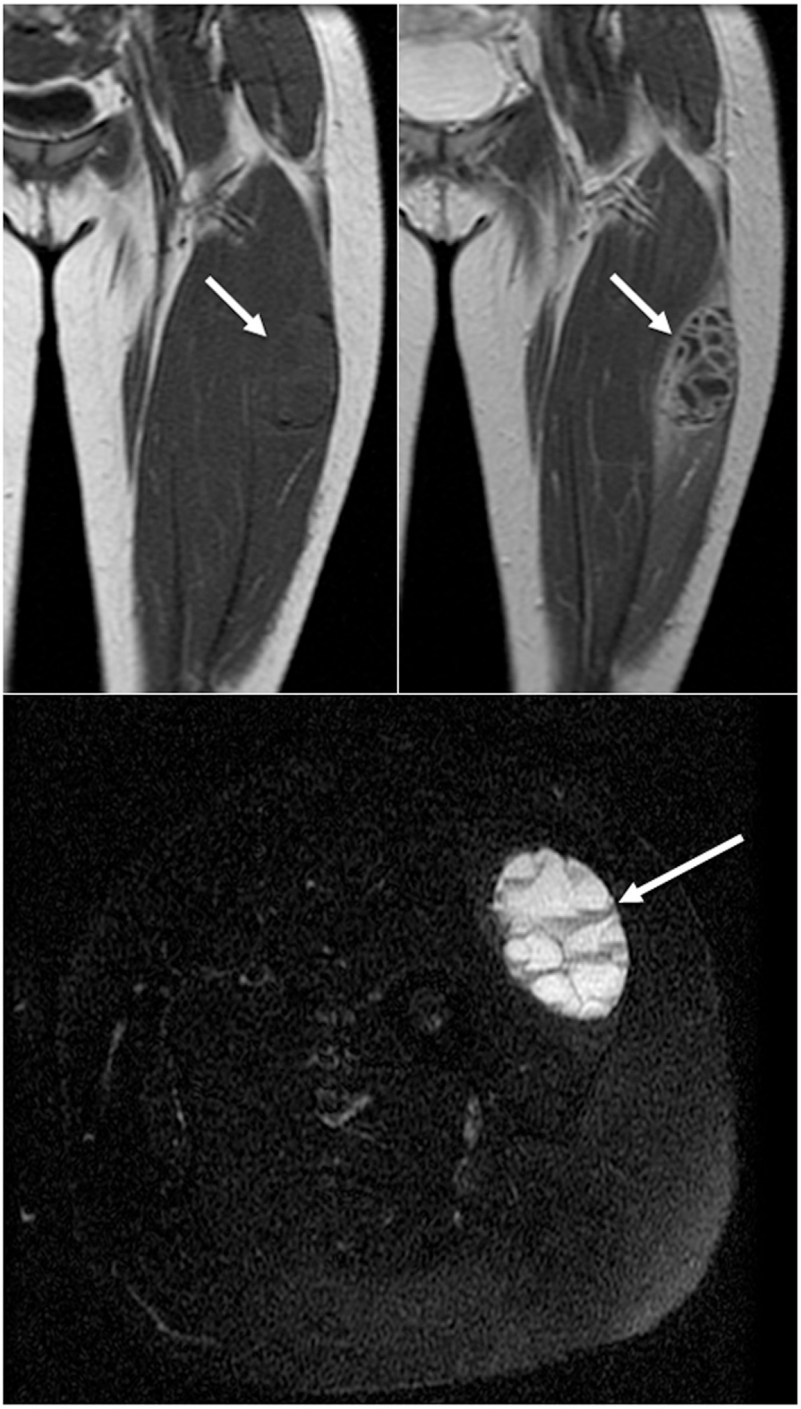

Fig. 1. Magnetic resonance images of a left thigh mass. Coronal T1-weighted, non-fat-suppressed pre-contrast (top left) and post-contrast (top right) images show an intramuscular lesion in the left vastus lateralis that was T1 isointense to skeletal muscle and demonstrated peripheral and septal enhancement (indicated by arrows). The lesion was T2 hyperintense and had associated fluid–fluid levels (arrow) on this axial T2-weighted, fat-suppressed image (bottom).

tiple internal fibrous septa composed of spindle cells, inflammatory cells and multinucleated giant cells along with hemosiderin containing macrophages, features characteristic of an ABC (Fig. 2C&D). There was subtle osteoid seen along the cyst wall. Fluorescence in-situ hybridization (FISH) was performed using break-apart probes which clearly demonstrated a USP6 rearrangement, thus confirming the diagnosis of ABC (Fig. 3).

The patient had an initial uneventful recovery but developed recurrent pain with associated swelling in the left thigh region 5 months later. Imaging studies demonstrated a fluid collection with features concerning for recurrence. A second wide local excision was performed. Microscopic examination of the recurrent lesion revealed near identical histologic features with a partial solid growth pattern in areas. Interestingly, a shell of lamellar bone at the edge of the lesion was evident. The newly formed bone was more mature at the periphery thus closely mimicking that seen in myositis ossificans (Fig. 2E&F). The patient has had no evidence of disease 2 years after re-excision.