Primary Xanthoma of The Acromion: A Case Report and Review of the Literature.

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Abstract

Primary “xanthoma” of bone is a rare lesion of unsettled histogenesis that may pose a diagnostic challenge owing to its wide range of differential diagnosis. Herein, we present a case of primary xanthoma of the right acromion in a middle aged woman who had no aberrant lipid metabolism or evidence of other pre-existing bone lesions. To our knowledge, this is the second reported case of a primary xanthoma involving the acromion in an adult.

Key words

Xanthoma, acromion, bone tumor.

Introduction

 Sheets of foam cells, giant cells and cholesterol crystals are often seen as secondary or degenerative findings in many neoplastic and non-neoplastic bone lesions. However, primary “xanthoma” of bone consisting essentially of lipid laden “foamy” histiocytes (xanthoma cells) is a rare bony lesion of unsettled histogenesis that may pose a diagnostic challenge to the pathologist, radiologist and clinician. (1) Developmental, degenerative and neoplastic histogeneses have been proposed by different authors and the differential diagnosis includes both non-neoplastic and neoplastic lesions (1,2). Xanthoma of bone is almost always solitary, and the flat bones (pelvis, ribs, and skull) are the most frequent sites of involvement (3). Occasional cases have been reported in the scapula (2). Herein, we present a case of primary xanthoma of the right acromion in a middle aged woman who had no aberrant lipid metabolism or evidence of pre-existing bone lesions. To our knowledge, this is the second reported case of a primary xanthoma involving the acromion in an adult in a patient with a normal lipid profile.

Case Report

A 38-year-old Saudi woman sought medical advice at a primary health care centre because of right shoulder pain for the last two years. The patient was referred to orthopaedic clinic of King Fahd Hospital of King Faisal University, Al-Khobar, Eastern Providence, because of increasing pain which became refractory to oral as well as intra-articular non steroidal anti-inflammatory drugs. On examination a spherical, 5 x 6 cm. hard lump was noted in the right shoulder, with no signs of local inflammation.

Fig. 1: Plain X-ray of the right shoulder: grossly expansible lytic lesion with a sclerotic margin and internal thick septation arising from the acromion.
Lipid profile, as well as all other laboratory investigations were within normal limits.

Right shoulder plain X-ray showed a grossly expansible geographical lytic lesion with a sclerotic margin and internal thick septation in the acromion (Figure 1). CT scan (Figure 2) showed the lesion to be well circumscribed, grossly expansile, lytic lesion arising from the acromion and extending to the scapular spine. There was bony erosion and remodelling with no fluid/fluid level, no soft tissue mass and no identifiable matrix. Surgical excision revealed a well capsulated, spherical, rough edged cystic mass filled with yellow friable material. The resulting space was packed with a bone graft. The patient was discharged with no complications and her symptoms were markedly relieved.

Pathological examination showed multiple, irregular, yellow and greyish-white tissue fragments measuring 8 x 8 x 4 cm in aggregate and weighing 30 grams. Representative sections were initially processed but subsequently all the remainder of the specimen was submitted in the pathology lab. Microscopic examination revealed a lesion consisting predominantly of foamy histiocytes (xanthoma cells) admixed with few multinucleate giant cells (osteoclastic and Touton’s types) and a small amount of fibrous tissue (Figure 3A). Cholesterol crystals, sometimes present within necrotic debris, and small numbers of mononuclear inflammatory cells were also focally observed (Figure 3B). There was no significant nuclear pleomorphism and mitoses could not be demonstrated. Immunohistochemically, the foam cells and the giant cells showed immunoreactivity for CD68 (Figure 4) and were negative for pancytokeratin,
indicating a histiocytic origin and excluding a metastatic clear cell carcinoma. Other co-existing bone lesions could not be found after total processing of the received material. The final diagnosis was “primary bone xanthoma”.

**Discussion**

Intraosseous xanthomas are rare, particularly in normolipidemic patients, in whom the presenting symptom is pain without skin lesions. Hyperlipidemia is present in most patients with xanthomas $(1,4)$. A lytic lesion with a rim of sclerosis is seen on radiographs. Histology shows foam cells, giant cells, and fibrosis. Intraosseous xanthoma is a benign tumor, and other diagnoses must be ruled out (histiocytosis X, Erdheim Chester disease, clear cell carcinoma metastasis). Surgical excision of the lesion is the elective treatment. The histogenesis and classification of fibrohistiocytic lesions involving bone and containing an admixture of fibrous tissue, foam cells, and giant cells are confusing and include several overlapping entities, such as metaphyseal fibrous defect, nonossifying fibroma, fibrous cortical defect, fibroxanthoma, xanthoma and benign fibrous histiocytoma (BFH) of bone $(2-4)$.

Some of these entities are considered to be developmental abnormalities, while others are considered to be neoplasms $(2)$. They may alternatively represent degenerative lesions. It has been suggested that bone xanthoma may represent a «burnt-out» benign condition such as fibrous dysplasia or histiocytosis X $(4)$.

Histological appearances vary from lesions showing a predominantly storiform, fibrous growth pattern with multinucleated giant cells, to lesions consisting predominantly of lipid laden “foamy” histiocytes (xanthoma cells). Some authors consider these latter xanthomatous lesions as neoplastic under the broad category of “benign fibrous histiocytoma”, while others favour a “primary” xanthoma of bone as a separate, possibly reactive entity $(3,4)$. In some cases there is a primary underlying disorder of cholesterol metabolism or other lipid abnormalities $(5)$. In the present case, the lipid profile as well as all other laboratory investigations were within normal limits. Moreover, other co-existing bone lesions could not be found after complete processing of the material received.

Xanthoma of bone usually presents in patients over the age of 20 years and has a male: female ratio of 2:1. The main presenting symptom is pain. It is almost always solitary, and the flat bones (pelvis, ribs, and skull) are the most frequently involved sites $(5)$. Our case was 38 years old female who presented with right shoulder pain. The lesion was present in the acromion. To our knowledge, this is the second reported case of a primary xanthoma involving the acromion in an adult. However, the previous case $(2)$ was reported under the designation of “Benign fibrohistiocytoma, xanthomatous variant”, a term that reflects the view of the author regarding the histogenesis and classification of the lesion as neoplastic and as a variant of BFH.

Radiographically, a well-defined, sometimes expansible lytic lesion, with either a small area of surrounding reactive bone or a distinct sclerotic margin is seen. Computed tomographic scans show multiple osteolytic areas, with an irregular trabecular pattern in the surrounding sclerotic bone $(5)$. T1-weighted magnetic resonance images show a lesion with central low signal intensity, surrounded by a peripheral ring with high signal intensity. The entire lesion shows high signal intensity on T2-weighted images, partially surrounded by areas with low signal intensity, concordant with reactive bone sclerosis $(6)$. In the present case, right shoulder plain X-ray showed a grossly expansible geographical lytic lesion with a sclerotic margin and internal thick septation in the acromion. CT scan showed the lesion to be well defined, arising from the acromion and extending to the scapular spine with bony erosion and remodelling. These features, although indicative of a benign lesion, are non-specific as to its precise nature and type.

The differential diagnosis of bone xanthoma includes both neoplastic and non-neoplastic lesions, such as Erdheim-Chester disease (a multisystemic granulomatosis), bone involvement insinushistiocytosis withmassivelymphadenopathy (Rosai-Dorfman disease), giant cell tumor of bone, eosinophilic granuloma, malignant fibrous histiocytoma, and in children, non-ossifying fibroma and fibrous cortical defect. More important
is the differential diagnosis with metastatic clear cell carcinoma\(^{(1,4)}\). In our case, systematic clinical examination, immunoreactivity for CD68 (a histiocytic marker) and negative immunostaining for pancytokeratin (an epithelial marker) helped exclude these conditions and establish a diagnosis of primary bone xanthoma. Surgical excision of the lesion is the elective treatment. The prognosis is satisfactory, even after partial excision \(^{(4)}\).

**Conclusion**

This is a rare case of bone lesion with unsettled histogenesis.

**References**


