Clavicle pain

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Discussion

In the case presented, a 16-year-old male with a history of repeated trauma to his left clavicle complained of intermittent and worsening left clavicular pain for several months. He was found to have a nondisplaced clavicle fracture with marked osteolysis on X-ray (Fig. 1) and worked up for a possible neoplasm. An open intralesional biopsy of the area of bone loss was taken and showed bone trabeculae replaced by fibrous connective tissue (Fig. 2a) and prominent vessels lined by a single layer of endothelial cells (Fig. 2b). No neoplastic, acute inflammatory, or infectious process was identified. One month later, he subsequently underwent open reduction and internal fixation with a major autologous bone graft to stabilize the middle segment of his left clavicle (Fig. 3). Lab values were all within normal limits. The patient was started on bisphosphonate. Seven months later, X-ray showed that several of the medially based screws had loosened with progression of osteolysis (Fig. 4). Four screws were subsequently removed from the left clavicle plate. At 11 months after the initial surgery, the patient had complete removal of the hardware (Fig. 5), since his bone resorption had progressed to a point where his clavicle was difficult to visualize on X-ray. After excluding other causes of osteolysis, such as infection, cancer, inflammatory and endocrine disorders, a final diagnosis of Gorham's disease (GD) was made based on the radiographic and histologic findings.

GD is a rare idiopathic bone disease characterized by spontaneous and progressive osteolysis and hypervascular and/or angiomatous fibrous connective tissue proliferation without osteoblastic reaction [1]. It does not have race or gender preference, but occurs more frequently in children and young adults. The disease can affect any single bone or multiple bones. The cranium, shoulder, and pelvis are the most common sites [2]. Clinical manifestations vary depending on the site and the extent of involvement. Some patients may present with abrupt pain due to pathologic fracture [3].

The diagnosis of GD is based on radiology and clinicopathology. The X-ray features are similar to those of CT, which can be patchy osteopenia progressing to osseous resorption without sclerosis or osteoblastic reaction. The osteolysis may extend to the contiguous bone and may not spare the intervening joint [4]. Intramedullary or subcortical lucent foci may be the earliest manifestation, with later slowly progressive atrophy, dissolution, fracture, fragmentation, and disappearance of a part of a bone. MRI demonstrates heterogeneously lobulated hyperintensity on T2WI with heterogeneous enhancement, related to the mix of hemorrhage and angiomatous and fibrous tissue [4]. Both CT and MRI are useful in the delineation of the soft tissue extension, and enable biopsy guidance and a surgical plan. Tc-99 m bone scintigraphy may demonstrate increased uptake of the radiopharmaceutical agents on the initial images, and subsequently decreased uptake corresponding to the osteolysis [5]. Besides neoplastic, infectious, metabolic osteolysis, differential diagnoses of GD include hereditary osteolysis, generalized lymphatic anomaly (GLA), and nonhereditary osteolysis with neuropathy. Hereditary osteolysis occurs during infancy and commonly involves the hands and feet without vascular changes. Lala et al. found that osteolysis seen in GLA is more insidious. Findings favoring GLA include more extensive involvement, the presence of discrete macrocystic lymphatic malformations, and visceral organ lesions [6].

GD is often progressive but sometimes can be self-limiting [7]. Serious complications are rare, but chylous pericardial and pleural effusions, spinal, and/or visceral involvement

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can be fatal [8, 9]. Its etiology and pathophysiology remain unclear, and trauma may be the trigger [9]. Current treatment includes anti-osteoclastic medication, interferon, radiotherapy, and surgery [10].

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

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