Vanishing Bone Disease of foot

M Singh, S Sharma, A Tikoo, D Singh

Citation

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Abstract

Vanishing bone disease, also known as idiopathic osteolysis and Gorham Stout disease is a rare clinicopathological entity characterized by proliferation of intra osseous capillaries and lymphatics thereby leading to bony destruction. All age groups can be affected. The most common sites for predilection are the shoulder and pelvis. The authors report a rare and interesting case of idiopathic osteolysis of right foot in a 50 year old female who presented with progressive regression of 2nd and 3rd toes.

INTRODUCTION

Idiopathic osteolysis, also known as vanishing bone disease, massive osteolysis, or Gorham-Stout disease; is a very rare bone condition of unknown origin that was described as a distinct pathological entity by Gorham and Stout in 1954 [1]. The first case of massive osteolysis was reported in1838 by Jakson [2].The disease has no racial or sexual predilection and has been described in patients from infancy to 75 years of age. Fujiku, et a.l in 2002 observed that the syndrome may occur in any bone, but common sites include the shoulder, spine, hand, ribs, jaw foot, spine and the pelvis.

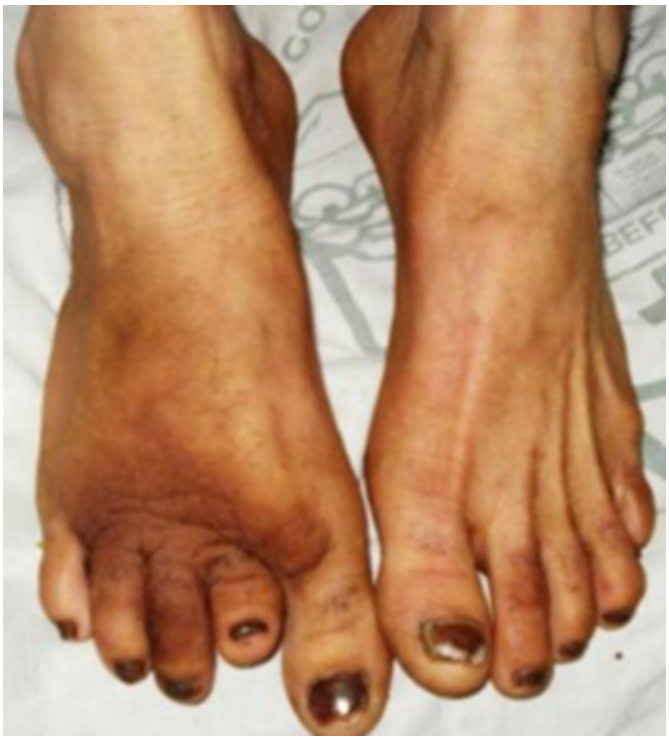
CASE REPORT

A 50-year old woman presented to us with complaints of pain and progressive decrease in size of 2 nd and 3 rd toes of the right foot since the last six months. This was accompanied by a slight difficulty in walking. Past history, family history and occupational history were insignificant.

On examination, the skin over 2 nd and 3 rd toes was hyper pigmented and there was obvious regression of 2nd and 3rd toes (Fig.1). The 2 nd and 3 rd metatarsals were not palpable. There was no movement of these toes and sensations were absent. Examination of rest of the phalanges, metacarpals and ankle joint was within normal limits. Examination of other systems was also within normal limits.

Figure 1

Figure 1: Showing regression of 2 and 3 toes of right foot

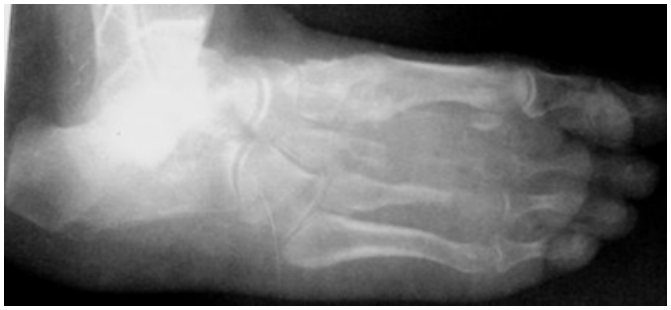


Serum biochemistry tests were all within the normal range.

Radiographic evaluation revealed extensive destruction of the 2nd, 3 rd and 4 th metatarsals and proximal phalanx of 2 nd toe of right foot (Figure 2). Radiographs of opposite foot, shoulder, hands, pelvis and ribs were normal.

Figure 2

Figure 2: AP Radiograph of right foot showing destruction of the 2nd, 3 and 4 metatarsals and proximal phalanx of 2 toe



Histopathological examination of the curettage material revealed presence of thin walled capillaries, endothelial tissue and fibrous tissue.

On the basis of clinical presentation, radiological evaluation and histopathology, a diagnosis of Gorham-Stout disease involving the right foot was made. The disease, its treatment modalities and prognosis were discussed at length with the patient. She was put on bisphosphonates and at follow ups after two years; the disease has not progressed further.

DISCUSSION

Gorham and Stout first recognized this disease as rapidly progressive osteolysis of one or more adjacent bones resulting from excessive intra-osseous proliferation of small blood vessels or lymphatic ducts.

The disease begins in infancy, but the usual age of presentation is above 35 years [3]. Males and females are equally affected. Shoulder and pelvic girdle are common sites of predilection [3]. Clinical manifestations vary and depend on the affected bone and spread of pathology to adjacent tissues. Most patients describe an insidious onset of painless, bony, or overlying soft tissue deformity and instability, which may have been present for several months to years. Another feature of the disease is the tendency to involve one bone only initially and then to spread frequently to adjacent bones, not respecting anatomic boundaries and readily crossing articular barriers. Spontaneous resolutions are known to occur.

The proposed mechanisms include hyperaemic granulation tissue in the bone and underlying endothelial dysplasia of lymphatics, blood vessels or both [4]. Moller et al have proposed that the disease is the result of deranged osteoclastic activity [5]. The stimulus which initiates osteolysis is not known,but mutations in the Matrix Metalloproteinase gene 2 (MMP2) have been linked to multicentric osteolysis and arthritis[6].

The characteristic radiological feature is increasing translucency of the involved bone without any sclerosis and progressive resorption of the bony structure [7].

Involvement of the contiguous bones can occur irrespective of intervening joints.

On histopathology, the osseous tissue is replaced by thin walled, sinusoidal blood vessels without reactive bone formation.

Numerous methods of treating the disease have been tried, including surgical excision with bone grafting and prosthetic replacement, radiation, and the administration of vitamin D, androgens, amino acids, adrenal extracts, calcium, fluoride, calcitonin and bisphosphonates. In the early stages, radiation therapy, bisphosphonates or calcitonin can arrest the disease. At present, there is no effective therapy. Surgical interventions are frequently not possible due to pronounced osteolysis and inadequate bone for fixation [8]. Radiotherapy with moderate doses (40–45 Gy in 2 Gy fractions) has been reported to lead to pronounced clinical improvement [9]. Because the disease is often self-limiting, conservative treatment appears to represent an appropriate treatment option.

The course of the disease is variable and often unpredictable. The prognosis is worse when the course of disease is complicated by neurological deficit or pleural effusion. Though rare, spontaneous resolution is known to occur.

CORRESPONDENCE TO

Siddhartha Sharma, House N0. 2, B Block, Housing Colony, Sector 2, Pocket 3, Channi Himmat, Jammu.
sids_82@yahoo.com +919469210778

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Author Information

Manjeet Singh, MBBS
Post Graduate Department of Orthopaedics, Government Medical College, Jammu

Siddhartha Sharma, MBBS
Post Graduate Department of Orthopaedics, Government Medical College, Jammu

Agnivesh Tikoo, MBBS
Post Graduate Department of Orthopaedics, Government Medical College, Jammu

Dara Singh, MS (Ortho)
Post Graduate Department of Orthopaedics, Government Medical College, Jammu