Case Report

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Primary Rosai-Dorfman Disease in 39-Year-Old Woman With Osseous Tibial Lesion Manifestion: A Case Report and Literature Review

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Abstract

Rosai-Dorfman disease (RDD), otherwise known as sinus histiocytosis with massive lymphadenopathy (SHML), is a very rare and typically benign disorder of unknown etiology with <10% bone involvement. The report is of a case seen at the authors' hospital of a patient presenting with several months' onset unspecified nontraumatic ankle pain. There was no physical mass or lymphadenopathy appreciated on examination. Plain radiographs and magnetic resonance images demonstrated an osteolytic lesion at the medial malleolus. Biopsy revealed the diagnosis of intraosseous manifestation of Rosai-Dorfman disease.

Keywords: Rosai-Dorfman, SHML, bone tumors, tumors, rare tumors, foot and ankle

Introduction

Rosai-Dorfman disease (RDD) is a very rare and typically benign disorder of unknown etiology characterized by a proliferation of histiocytes in lymph nodes. Speculations regarding viral infectious causes are unproven at this time. First described in 1965 by Destombes after observing several patients with similar presentation of cervical lymphadenopathy and identical histologic findings, this disease was officially classified in 1969 by 2 pathologists, Rosai and Dorfman.

RDD predominantly affects children and young adults with symptoms of fever, malaise, painless cervical lymphadenopathy, weight loss, and pharyngitis.[3,10] Laboratory results typically show elevated erythrocyte sedimentation rate or leukocytosis.[3] Almost half of the cases seen have extranodal involvement (skin, soft tissue, central nervous system, or gastrointestinal tract), typically presenting as subcutaneous nodules.

Bony involvement is typically seen in <10% of cases; however, primary bone RDD is even less common. In 2002, Foucar and colleagues reported that 2% of 423 patients in their study had osseous RDD without lymphadenopathy.[3] RDD bone lesions typically are multifocal and lytic within the cancellous areas of long bones; however, they can vary in appearance, with possible soft tissue extension. Differentials include multifocal osteomyelitis, metastatic neuroblastoma, or Langerhans cell histiocytosis.[4,7]

The following is a case seen at our hospital presenting with several months' onset unspecified nontraumatic ankle pain. Biopsy results led to a diagnosis of intraosseous manifestation of Rosai-Dorfman disease.

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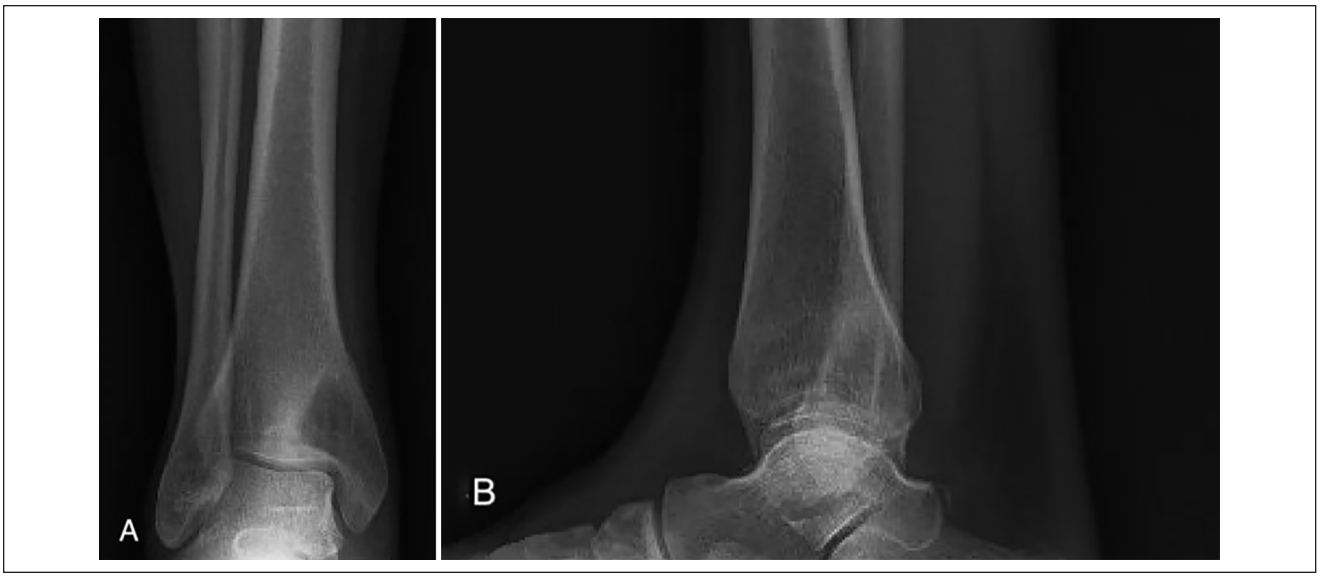

Figure 1. Conventional radiographs of the ankle reveal a lytic lesion without substantial cortical involvement. (A) Anteroposterior and (B) lateral images are shown.

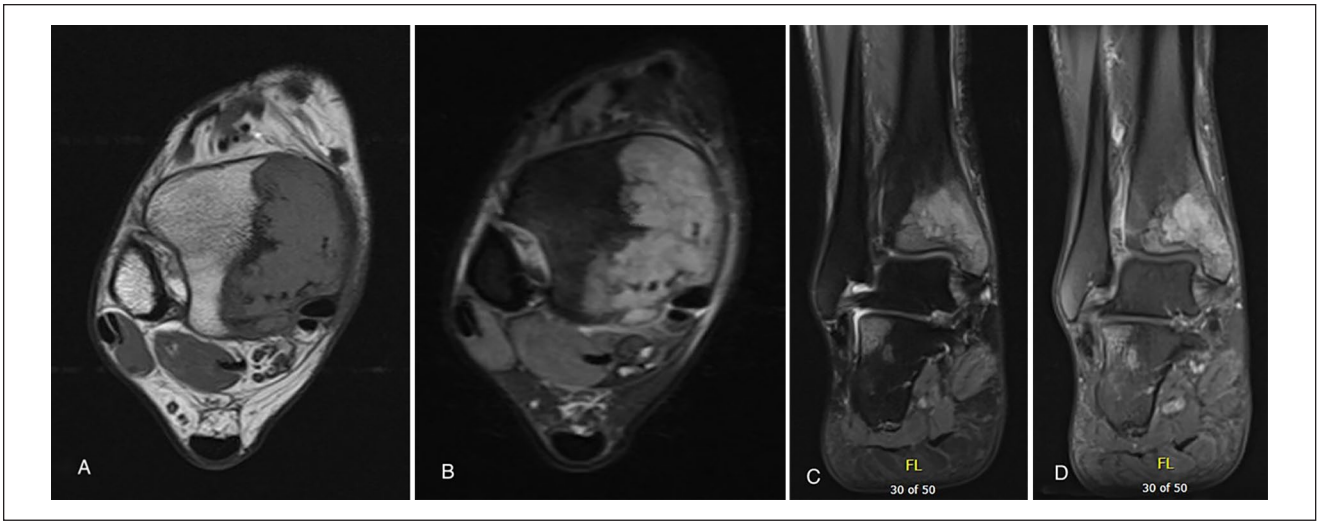

Figure 2. Magnetic resonance imaging reveals enhancing marrow-replacing lesions of the medial malleolus and lateral calcaneus without significant cortical or soft tissue involvement. (A) Axial T1, (B) axial T2 FSE, (C) coronal T2 FSE, and (D) coronal T2 FS. (FS, fat saturated; FSE, fast spin echo.)

Case

A 39-year-old Hispanic woman presented with atraumatic nonspecific right ankle pain that had been progressively getting worse over the past 8 months. The pain was predominantly at night and relieved with activity modification. She related no other symptoms. Physical examination demonstrated limited and painful range of motion of the right ankle. There were no skin lesions, masses, edema, or lymphadenopathy noted. Patient had intact neurovascular status. Clinical diagnosis at the time was posterior tibial tendonitis. Pain was noted to be recalcitrant to treatment, consisting of ankle brace, oral steroids, and physical therapy.

Conventional radiographs demonstrated a lytic bony lesion in the right medial malleolus (Figure 1). Magnetic resonance imaging scan demonstrated a contrast-enhancing marrow replacement lesion in the medial malleolus as well as the lateral calcaneus (Figure 2). Whole body bone scan demonstrated multiple lesions throughout the bilateral lower extremites (Figure 3).

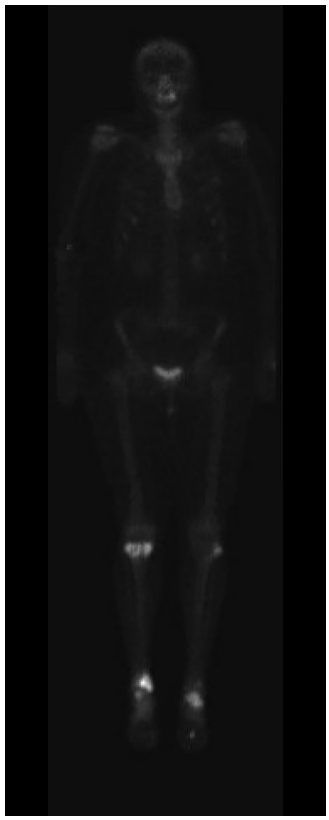

Figure 3. Whole body bone scan demonstrating multiple lesions in the bilateral lower extremities.

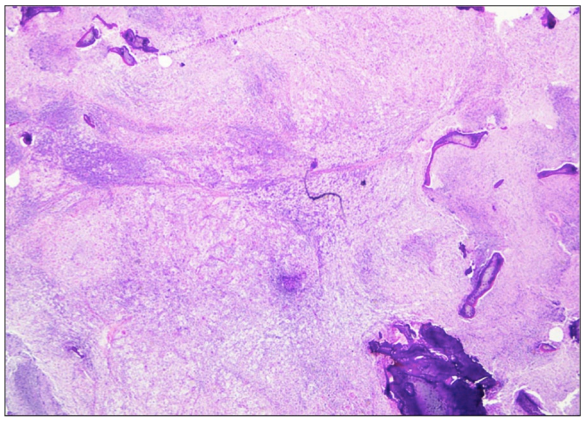

Figure 4. Low-power image of specimen demonstrating inflammatory process.

Erythrocyte sedimentation rate was elevated, at 54, as was the serum C reactive protein level, at 32.65. The white blood cell count was normal.

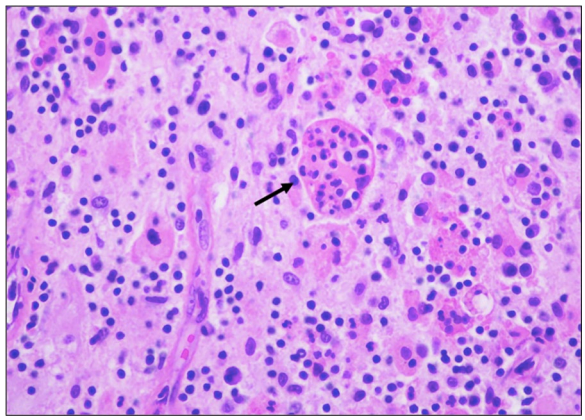

Figure 5. Image of emperipolesis (arrow) with multiple inflammatory cells within the cytoplasm of a histiocyte (hematoxylin-eosin, original magnification × 40).

At that time, the patient was referred to an orthopaedic oncology surgeon on suspicion of bone tumor. Computed tomography results to determine organ involvement were negative. Computed tomography–guided core needle biopsy by interventional radiology was nondiagnostic, describing lymphoplasmatic infiltrate with fibrosis suspicious for chronic osteomyelitis.

Open biopsy was performed by orthopaedic surgery and the patient was noted to have what was seemingly purulence within cysts. A specimen was sent to the laboratory to rule out lymphoma. Osseous lesion was excised and repaired with cancellous chips. Pathology results revealed an inflammatory process (Figure 4)—a mixture of lymphocytes, plasma cells, and histiocytes. Upon closer examination, classic finding of emperipolesis was noted, confirming a diagnosis of Rosai-Dorfman disease. Emperipolesis, shown in Figures 5 and 6, is an uncommon biological process of nondestructive phagocytosis in which an engulfed lymphocyte remains intact within the cytoplasm of the histiocyte. The patient was managed conservatively. At 6 months postoperatively, her symptoms were slowly improving, she denied any new complaints, and her bony lesions were seen slowly consolidating on radiographs.

Discussion

Bone involvement occurs in <10% cases of RDD, with primary bone RDD being even more rare. Since its official classification in 1969, more than half of the approximately 600 RDD cases that have been registered presented in extranodal sites and with constitutional symptoms.[2,3] To our knowledge, few other reports published have identified

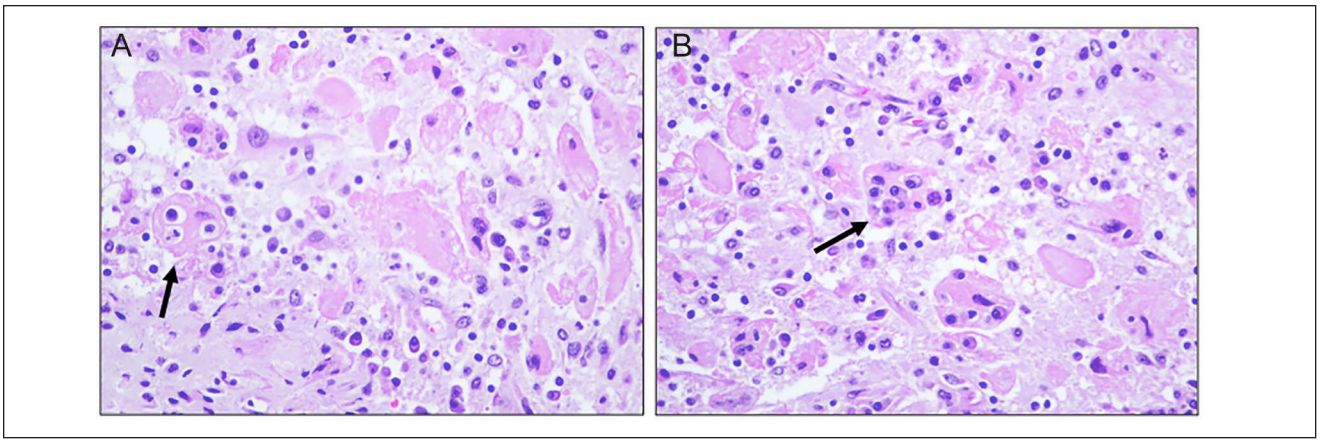

Figure 6. (A, B) Images of emperipolesis (arrow) with multiple inflammatory cells within the cytoplasm of a histiocyte (hematoxylin-eosin, original magnification × 40).

involvement of the foot and ankle, and no others have identified solely osseous manifestation in the distal tibia. Osseous manifestation of RDD in the talus of patients presenting with primary musculoskeletal complaints has been reported in the literature.[1,6,8] Moreover, 90% of patients with RDD will have complete remission with conservative treatment. Symptomatic treatment of lymph node debulking or osseous cyst curettage and packing is common. Prophylactic internal fixation has been reported as well. Death occurs in less than 3% of cases, seen in those with organ failure as a complication of RDD.[9] The mainstay of treatment is clinical observation.[7] Fairly recently in the literature, researchers have been documenting fine-needle aspiration cytology to diagnose osseous RDD without lymphadenopathy,[5,11] although in this case even large-needle biopsy was nondiagnostic. As with our patient, although isolated bone RDD is very rare, it can be a differential diagnosis in patients with musculoskeletal complaints in the foot and ankle.

Ethical Approval

Ethical approval was not sought for the present study because non-identifying/anonymous and archival data were used, and the case study exposed minimal risk to the patient.

Declaration of Conflicting Interests

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