Case Report

A Solitary Rosai-Dorfman Disease of The Talus: The First Case Report of Bisphosphonate Therapy with Imaging Follow-up

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Rosai-Dorfman disease (RDD) manifesting as a solitary osseous lesion especially of talus bone is rare. The authors reported a 31-year-old Thai man who had chronic left ankle pain and the biopsy of his talar lesion demonstrated emperipolesis, the typical histological feature of RDD. He was treated with curettage and adjuvant bisphosphonate and appeared to show improvement in clinical symptoms and radiological evidence. To the authors' knowledge, this is the first report of an intraosseous RDD lesion treated with bisphosphonate with imaging follow-up.

Keywords: Rosai-Dorfman disease; Talus; Solitary; Radiology; Bisphosphonate

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Rosai-Dorfman disease (RDD) is a unique entity of histiocytic proliferation or disorder of unknown etiology. It was first described in cases primarily involving lymph nodes by Rosai and Dorfman in 1969 under the name "sinus histiocytosis with massive lymphadenopathy"[1]. Later, involvement of different organs was reported, including solitary and multiple lesions or multi-organ lesions. However, RDD manifesting as a solitary intraosseous lesion with no lymphadenopathy is rare and accounts for less than 1% of all cases so far reported[2]. The skull is the most reported site of solitary intraosseous RDD[2,3]. To the authors' best knowledge, this presentation would be the fourth report of a solitary talus lesion in the English literature[3-5]. Moreover, it was also the first trial of bisphosphonate therapy for a symptomatic osseous lesion affected by RDD.

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Case Report

A 31-year-old Thai man presented with chronic pain of the left ankle for one year. The pain was mostly aggravated at night. He had no predisposing trauma or any significant history of medical illness. His vital signs were stable and afebrile. Physical examination showed tenderness and mild swelling of the left ankle with limited ranges of motion, especially in dorsiflexion. No associated lymphadenopathy was noted. Result of a complete blood count was within normal limits with white blood cell count at 6,950 cells/microliter, neutrophils 66.1%, lymphocytes 25.5%, eosinophils 2.2%, and basophils 0.7%. Serum C-reactive protein was 4.9 mg/L, which the normal range is 0 to 5 mg/L.

Plain radiographs and computed tomography (CT) of the left ankle demonstrated mixed osteolytic and sclerotic areas occupied the talus with cortical destruction (Figure 1). Magnetic resonance imaging (MRI) revealed an infiltrative, marrow-replacing process involving the entire talus with destruction of the cortex and extension into adjacent soft tissue (Figure 1). The initial diagnosis by radiologic perspective was inconclusive and possibilities for differential diagnosis included primary lymphoma, chronic osteomyelitis, osteosarcoma, and unusual metastatic cancer.

The biopsy of the lesion showed lymphocyte and plasma-cell infiltrations in the fibrotic stroma. Some foci demonstrated several large histiocytic

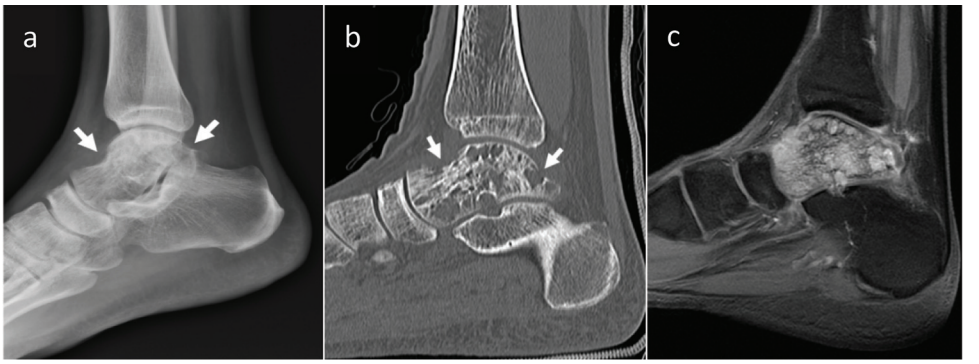

Figure 1. A 31-year-old male who presented with chronic left ankle pain. (a) Plain radiograph of left ankle in lateral view and (b) computed tomography in sagittal view, show multiple osteolytic lesions with some sclerotic areas involving the left talus with cortical destruction (arrows). (c) Magnetic resonance imaging, contrast-enhanced T1W fat suppressed image in sagittal view reveals an enhanced infiltrative marrow-replacing lesion involving the entire talus with several areas of cortical destruction with focal soft tissue extension.

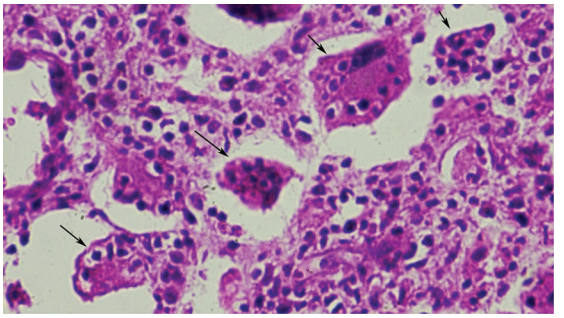

Figure 2. Histology of the lesion (H&E; original magnification ×400) shows numerous large histiocytes that have engulfed intact lymphocytes and plasma cells within the faint eosinophilic cytoplasm imparting so-called "emperipolesis" (arrows).

cells containing numerous intact lymphocytes and plasma cells in their cytoplasm, imparting so-called "emperipolesis" (Figure 2). Immunohistochemical stains revealed the large histiocytes were positive for S100 protein and CD163 but negative for CD1a and CD68 (Figure 3). Therefore, a diagnosis of RDD was entertained. The patient was initially treated with curettage, but his pain was not relieved. He was then given monthly intravenous injections of 4-mg zoledronic acid or bisphosphonate, for 18 months. The pain completely subsided after eight months of bisphosphonate therapy. Neither lymphadenopathy nor any other lesion developed during the entire follow-up period. Plain radiography and magnetic resonance imaging at 12 months after initiation of bisphosphonate therapy showed increased thickening of bone trabeculae with small areas of fatty marrow replacement in the lesion (Figure 4).

Discussion

RDD affecting bone either as primary or secondary forms is rare. To the authors' knowledge, there are no characteristic radiological features known to allow for definitive diagnosis of RDD, even though it is important to differentiate it from other mimicking lesions such as lymphoma, chronic osteomyelitis, osteosarcoma, and unusual metastatic cancer. Thus, radiologists and clinicians should be aware of this mimicry to avoid misdiagnosis of RDD. For correct diagnosis of RDD, histological evidence is mandatory. The key diagnostic feature is the presence of emperipolesis, such as presence of intact lymphocytes or plasma cells in the cytoplasm of histiocytes[1]. The supportive immuno-stains for this entity were typically positive for S100 and CD68[6]. According to the consensus recommendations for the diagnosis of this disease in 2018, the committee added further criteria of positivity of Fascin, CD14, and variable CD163 aside from positivity of S100 and CD68[7]. However, CD163 positive cases with variable CD68 similar to the case presented were occasionally reported but still in minority[8,9]. Some investigators mentioned that CD163 is a more specific marker of macrophage and monocyte differentiation than CD 68 in the sense that CD68 is an organelle-specific marker for lysosomes rather than a lineage-specific marker that is also expressed in fibroblasts and epithelial cells[10,11]. In addition, there were reports of CD68 negative but CD163 positive in other histiocytic lesions such as histiocytic sarcoma[12], atypical xanthogranuloma[13], atypical fibrous histiocytoma[10], Langerhans cell histiocytosis[10], and giant cell tenosynovial tumors[10].

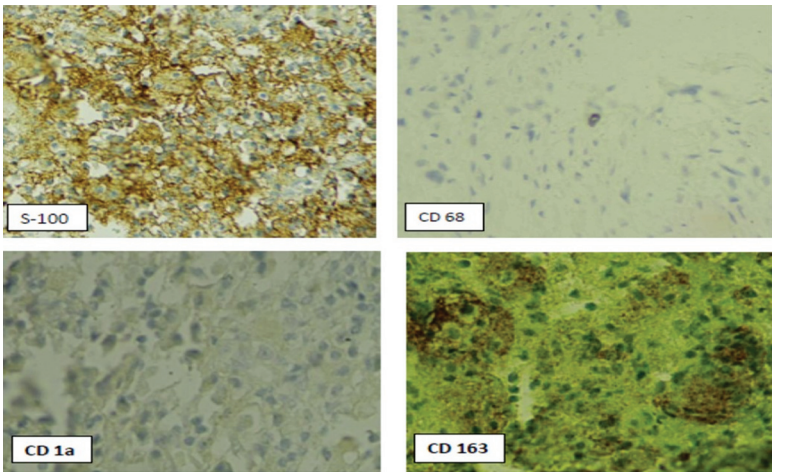

Figure 3. Immunohistochemistry shows large histiocytic cells in the lesion that are positive for S100 and CD163 but negative for CD68 and CD1a.

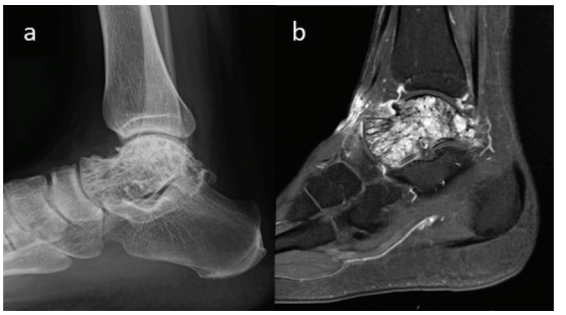

Figure 4. Follow-up imaging of the left ankle. (a) Plain radiograph in lateral view and (b) magnetic resonance imaging, contrast-enhanced T1W fat suppressed image in sagittal view demonstrates increased thickening of the bone trabeculae and minimally decreased extension and soft tissue component of the infiltrative marrow-replacing lesion in the talus.

The clinical course of RDD is often benign with most cases being self-limiting, and so far, no specific therapy has been recommended[3]. In general, systemic forms of RDD have been successfully treated by corticosteroid agents with or without cytotoxic chemotherapy. For symptomatic intraosseous RDD, bone curettage is the typical treatment for pain control and relief[14,15]. Owing to information regarding bone resorption induced by osteoclastic activity leading to expansive intramedullary lesions and cortical destruction in a series of 15 cases of intraosseous RDD reported by Demicco et al, the authors chose to try bisphosphonate for the present study case to inhibit osteoclastic activity and with the hope that it might also control pain and prevent future fracture[16]. After treatment, the patient showed improvement in both radiologic evidence and clinical symptoms, namely the pain completely subsided after eight months therapy of bisphosphonate. At the present time, it may not be able to conclude that the improvement was due to the treatment since most cases were self-limited.

In summary, the case report presented a rare primary RDD of the talus. The diagnosis was established histologically by presence of emperipolesis, positive S100 and CD163 proteins but negative CD1a. The patient was treated with bisphosphonate for 18 months and showed improvement in terms of clinical symptoms and radiologic evidence. To avoid misleading, the authors would like to emphasize that bisphosphonate should not be tried in general cases of RDD since the drug has not yet been approved for such lesion. It may be tried in only exceptional cases who have constraint in conventional treatment.

What is already known on this topic?

A solitary Rosai-Dorfman disease of the talus is rare and can mimic other bone pathology.

What this study adds?

This is the first trial of Bisphosphonate for pain relief in intra-osseous RDD.

Ethical approval and consent to participate

The study was approved by the Ethic Committee of Queen Savang Vadhana Memorial Hospital (IRB No.004/2021) and written informed consent for patient information and images to be published was provided by the patient.

Conflicts of interest

The authors have no conflicts of interest to declare.

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