

CASE REPORT

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Extranodal Rosai-Dorfman Disease: A Solitary Lesion with Soft Tissue Reaction

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We report a case of extranodal Rosai-Dorfman disease (RDD) (sinus histiocytosis with massive lymphadenopathy) presenting with a solitary active lesion of the femur.

Key words: Rosai-Dorfman disease, skeletal, femur, MRI

INTRODUCTION

ROSAI-DORFMAN DISEASE (RDD), ALSO KNOWN AS sinus histiocytosis with massive lymphadenopathy (SHML), is a rare benign entity of unknown etiology.¹ Classically, the most clinical finding is painless, bilateral, massive cervical lymphadenopathy accompanied with inflammatory abnormalities of laboratory findings, and often weight loss and immunological abnormalities such as hypergammaglobulinemia.²⁻⁵

On the other hand, extranodal involvement of RDD also has been noted in 182 of the 423 patients since 1969.⁶ Skeletal involvement has been identified in less than 5-10% of cases and is usually multifocal at initial presentation.⁷ Patients presenting with a solitary extranodal lesion without additional clinical manifestations are rare.

We report a case of extranodal RDD manifesting as a solitary active lesion of the femur.

CASE REPORT

A 38-year-old woman presented with a persistent spontaneous pain in her right thigh. There was no palpable mass on her right thigh where she had tenderness. Physical examination revealed that limited motion was not present in her right hip joint. She did not have a

previous history of trauma or infection. Laboratory tests showed the following abnormal results: CRP 2.6 mg/dl, white cell count 10,000/ μ l, alanine aminotransferase 72 IU/l, and aspartate aminotransferase 50 IU/l. All other relevant laboratory values were within the normal ranges. Radiographs of the right femur revealed a 2 cm osteolytic lesion in the proximal metaphysis of the right femur with anterior and posterior cortical thinning (Fig. 1). There were no findings of periosteal reaction or intramedullary sclerosis. Computed tomographic (CT) scanning of the right femur showed cystic areas with shell, anterior and posterior cortical thinning, and poorly marginated osteolytic areas in the medulla (Fig. 2). There were no findings of cortical destruction or extraosseous extension of the lesion. Magnetic resonance (MR) imaging of the right thigh revealed a lesion measuring approximately 60 mm \times 35 mm located in the proximal metaphysis of the right femur. The lesion was heterogeneously low to intermediate in signal intensity compared with fatty marrow on T1-weighted spin echo (SE) MR images (Fig. 3). On T2-weighted fast spin echo (FSE) MR images, the lesions were heterogeneously intermediate to high signal intensity relative to muscle (Fig. 4). Areas with heterogeneously intermediate to high in signal intensity on T2-weighted FSE MR images corresponded to poorly marginated osteolytic areas in the medulla that were identified on CT. The lesion was diffusely and heterogeneously enhanced on Gadolinium-enhanced T1-weighted SE MR images with fat suppression (Fig. 5). There were also no findings of cortical penetration or extraosseous expansion on MR images. On Gadolinium-enhanced T1-weighted SE MR images with fat suppression, the gluteus medius muscle was heterogeneously enhanced, which corresponded to inflammatory reaction of the soft tissue (Fig. 5). Bone scintigraphy using hydroxymethylene diphosphonate (HMDP) revealed a solitary lesion of the proximal femur with prominent

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uptake that appeared much more located compared with the corresponding T1-weighted SE MR images (Fig. 6). CT scanning of the chest, abdomen, and pelvis failed to reveal any pertinent abnormalities. Considering the age of the patient, the radiological differential diagnosis included infection, metastatic disease, and hematologic malignancies. Biopsy specimens were obtained from osteolytic areas in the medulla identified on CT, and the histological diagnosis of RDD was made. Histologic examination showed a remarkable increase in the number of histiocytes and manifest emperipolesis (Fig. 7), in which normal lymphocytes were enveloped within the cytoplasm of histiocytes. The histiocytes strongly expressed CD68, S-100, and vimentin, but lacked CD1a expression. The surrounding lymphocytes were positive for CD3 and CD79a.

DISCUSSION

RDD is a rare condition of uncertain lineage, with histiocytic infiltration, often associated with sinus expansion and lymphophagocytosis of nodal lesions. RDD may occur in extranodal sites, often without any involvement of lymph nodes.

Since 1969, when Rosai and Dorfman first described RDD, more than 400 cases have been reported.⁶ Of these, extranodal involvement has been noted in approximately 40% of patients in the registry of RDD. Patients with extranodal RDD have a predilection for older age than those with nodal-based disease. Extranodal RDD is usually observed in the paranasal sinuses, soft tissue, bone, and orbits. Cutaneous and suprasellar lesions have also been reported.^{2,4}

Skeletal lesions are usually multifocal, and patients



Fig. 1. Front view of the radiograph shows an osteolytic lesion in the proximal metaphysis of the right femur with anterior and posterior cortical thinning. There are no findings of periosteal reaction or intramedullary sclerosis.

presenting with a solitary extranodal lesion of bone are rare. Foucar colleagues showed a frequency of bone involvement of less than 10%.⁶ Three of the nine patients lacked other sites of extranodal involvement and had isolated osseous disease at presentation. Some sporadic cases of osseous involvement have reported.⁶⁻¹⁴ Of these, the skull was reported as the most common site of involvement in patients who had a solitary bone lesion. Solitary involvement of the femur, vertebrae, sacrum, radius, ulna, tibia, talus, phalanges, index finger, and metacarpal bone have also been reported.⁷⁻¹⁴

The skeletal lesions of RDD are typically lytic and

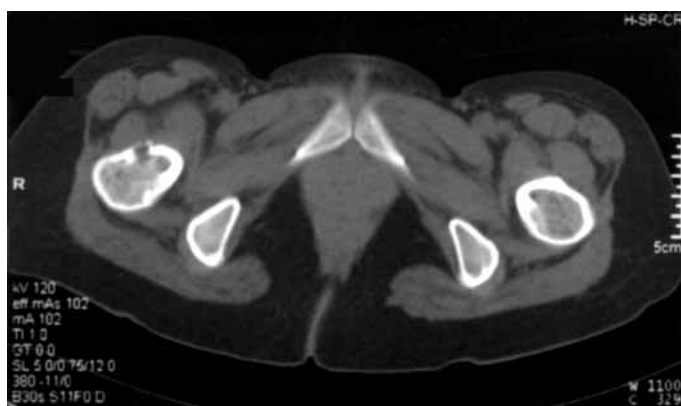


Fig. 2. Axial CT scan (A) and MPR image (B) of the right femur show cystic areas with shell, anterior and posterior cortical thinning, and poorly marginated osteolytic areas in the medulla.

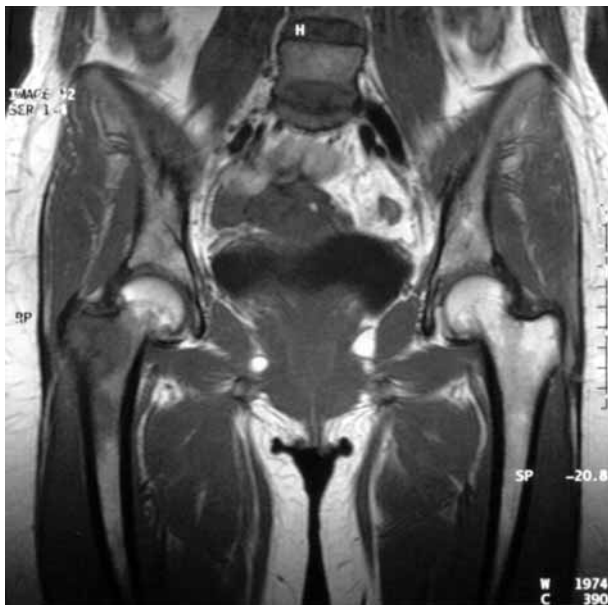


Fig. 3. Coronal T1-weighted spin echo MR image (1.5 Tesla: repetition time msec/echo time msec, 550/12) shows heterogeneously low to intermediate signal intensity compared with fatty marrow.



Fig. 4. Axial T2-weighted fast spin echo MR image (TR/effective TE: 4700/120). The lesions show heterogeneously intermediate to high signal intensity relative to muscle.



Fig. 5. Axial Gadolinium-enhanced T1-weighted SE MR image with fat suppression (779/12) shows diffusely and heterogeneously enhanced lesion. Also noted is heterogeneous enhancement of the gluteus medius muscle, which corresponds to inflammatory reaction of the soft tissue (arrow).



Fig. 6. Front view of bone scintigraphy using hydroxymethylene diphosphonate (HMDP) shows a solitary lesion of the proximal femur with prominent uptake (arrow).

intramedullary on radiography, with margins that may be either poorly or sharply defined, but rarely are sclerotic. One case presenting sclerosis of the cuboid on radiography showed lesion healing 12 months after the initial diagnosis.¹⁵ Complete disappearance of the lesion or the presence of a residual sclerotic focus has been considered to be a healing condition.¹⁶ However,

the pathologic diagnosis of our case was not similar to those of previous studies. Microscopic observations of biopsy specimens from the osteolytic lesion in the medulla revealed active disease without healing focus in our case. Microscopically, the lesion consisted of sheets of histiocytes, neutrophils, and lymphocytes. Although the lesion presented with slight fibrosis or

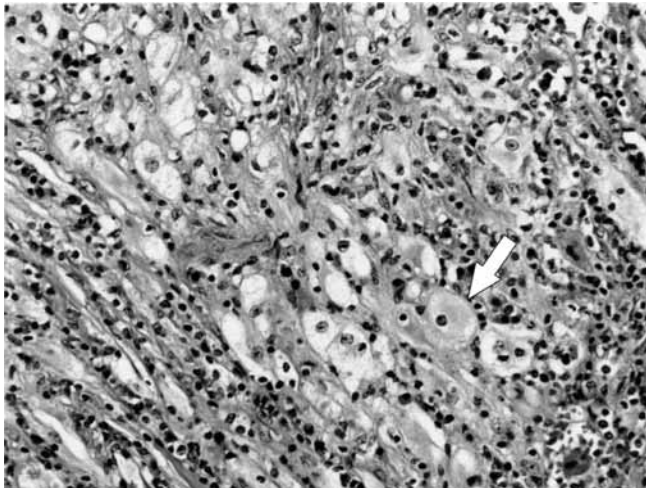


Fig. 7. Photomicrograph of the specimen (original magnification, $\times 100$; H-E stain) demonstrates remarkable histiocytes with emperipolesis (arrow).

xanthomatous focus in part, pathologic findings were different from those of healed lesions.

According to the rarity of extranodal RDD, studies describing CT and MR findings are limited. In our case, MPR images showed a lesion with cystic areas with shell, anterior and posterior cortical thinning, and poorly marginated osteolytic areas in the medulla. Most areas of the lesion showed heterogeneously high signal intensity on T2-weighted MR images. The lesion was diffusely and heterogeneously enhanced with extraosseous inflammatory reactions on contrast-enhanced MR images. The CT and MR imaging findings are similar to those of osseous lesion in other histiocytic disorders: Langerhans cell histiocytosis and Erdheim-Chester disease. Although the signal characteristics of MR imaging findings were nonspecific, corresponding MR images demonstrated extensive medullary involvement with contiguous reactive changes of the soft tissue.

Bone scintigraphy provides important information for detecting multiple lesions of RDD. However, recent imaging reports with special emphasis on RDD did present a solitary bone involvement without description of bone scintigraphic findings. Patients suspicious of RDD should undergo bone scintigraphy for screening because skeletal lesions are usually multifocal in patients with RDD. HMDP scintigraphy of our case revealed a solitary lesion of the proximal femur with prominent uptake.

Most patients require no therapy because the disease is usually self-limited and characterized by spontaneous regression. Patients who have osseous manifestations of RDD do not show any significant difference from the total group of RDD patients. In our case, the lesion persisted undiagnosed for six months and followed a course of stable disease without treatment.

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