
CASE REPORT

Imaging Features of Isolated Extranodal Rosai-Dorfman Disease in Iliac Bone

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ABSTRACT

We report a case of isolated extranodal Rosai-Dorfman disease occurring in iliac bone. Magnetic resonance imaging, computed tomography, and pathological features of Rosai-Dorfman disease are discussed.

Key Words: Histiocytosis, sinus; Lymph nodes; Lymphatic diseases; Thigh

中文摘要

髂骨孤立性結外Rosai-Dorfman病的影像特徵

林家昌、冼凱忻、徐波、黃雪雲

本文報告一例發生於髂骨的孤立性結外Rosai-Dorfman病，並討論Rosai-Dorfman病的磁共振成像、電腦斷層掃描和病理特徵。

INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare idiopathic histiocytic disease that was first described in 1969 by Rosai and Dorfman.¹ The disease is characterised by sinus histiocytosis with massive lymphadenopathy. Extranodal RDD is uncommon. Here we report a case of RDD involving the iliac bone of the pelvis.

CASE REPORT

In April 2014, a 43-year-old man presented with right-sided sciatica for 1 month after minor sprain of the lower back. He had a history of chronic low back pain. On physical examination, there was diffuse tenderness over the lower back. Lower limb power, sensation, and reflexes were all normal. No superficial or deep lymphadenopathy was detected. Complete blood cell

count, erythrocyte sedimentation rate, and liver and renal functions were unremarkable.

Plain radiographs of the lumbosacral spine showed no bony destruction. The patient was thought to have a prolapsed intervertebral disc and, therefore, non-contrast magnetic resonance imaging (MRI) of the lumbosacral spine was performed. Incidentally, a 2.0-x 1.4-cm T1-weighted isointense lesion was seen at the medial aspect of the right iliac bone. The lesion was mildly hyperintense in T2-weighted sequence (Figure 1). The lesion had a well-defined, hypointense T1 and T2 border suggestive of a sclerotic rim. No cortical break and no associated soft tissue mass were seen.

Computed tomography (CT) performed 6 months later

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showed a vague lytic lesion at the same site (Figure 2). However, no hyperdense border could be seen to match the hypointense border seen on the MRI scans. No associated bone expansion, endosteal scalloping, or

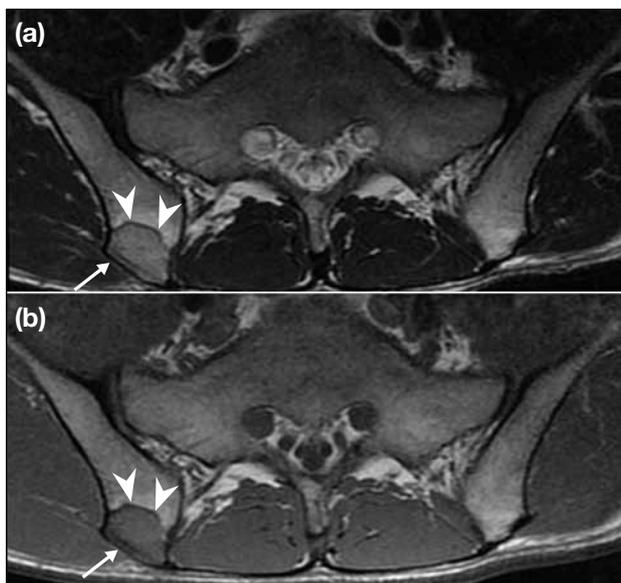


Figure 1. Magnetic resonance images of the patient with extranodal Rosai-Dorfman disease in the iliac bone. (a) T2-weighted and (b) T1-weighted images at the S1 level show a well-defined T2 mildly hyperintense and T1-isointense lesion at the posterior right ilium (arrows). There is a hypointense border in both sequences suggestive of sclerosis (arrowheads).



Figure 2. A computed tomography image in a narrow window shows a vague lytic lesion in the posterior right ilium (arrow). No hyperdense rim is seen to match the hypointense border seen in magnetic resonance images.

cortical destruction was seen. No internal septation or calcification was detected.

To ascertain the nature of the lesion, CT-guided bone biopsy of the lesion was performed. Pathological sections showed cellular infiltrate in the bone marrow spaces (Figure 3). The infiltrate included large pale cells with rounded nuclei and abundant clear-to-pale eosinophilic cytoplasm (Figure 4). Emperipolesis was seen. The background was reactive mononuclear cells, including lymphocytes and plasma cells in the loose fibrous stroma. The bone trabecula showed degenerative and regenerative changes. No clonal proliferation was identified on immunohistochemical study for immunoglobulin light chain of kappa and lambda. The

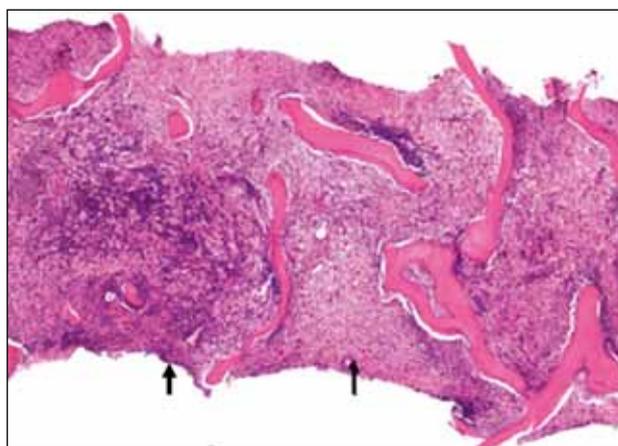


Figure 3. Pathological section shows cellular infiltrate in the bone marrow spaces (arrows) [H&E; original magnification, x 40].

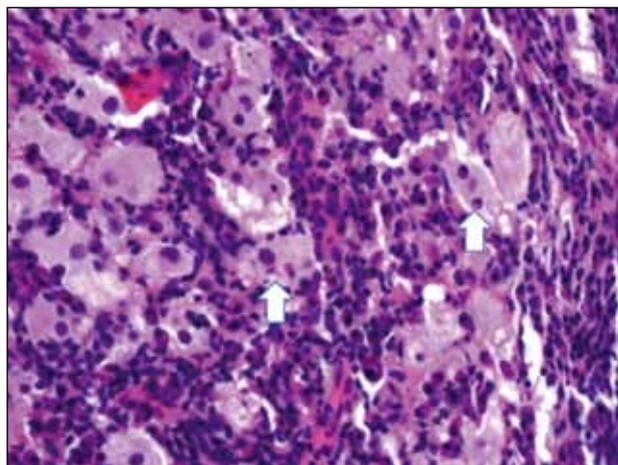


Figure 4. Pathological section shows large pale histiocytes with emperipolesis (arrows) in the background of mixed inflammatory infiltrate rich in plasma cells (H&E; original magnification, x 400).

large cells were positive for S100 protein (Figure 5). The features were compatible with extranodal RDD.

No surgical treatment was needed. Follow-up MRI with gadolinium contrast performed 15 months later showed that the lesion was enhancing (Figure 6). Increase in T2-hyperintense signal was observed when compared with the initial MRI study. A new cortical break was seen at the posterior part of the lesion. The size of the lesion was unchanged.

The patient opted for regular monitoring by MRI despite the possibility of disease progression, which had been explained.

DISCUSSION

RDD was originally described by Rosai and Dorfman in 1969 as a disease entity named “sinus histiocytosis with massive lymphadenopathy”.¹ Most of Rosai and Dorfman’s patients presented with bilateral painless cervical lymphadenopathy. RDD was classified as a benign tumour of undefined neoplastic nature by the World Health Organization in 2013.² It is a rare condition that usually manifests as nodal disease. About 2% to 10% of nodal RDD has simultaneous bone involvement.² Extranodal RDD has been described in about 43% of patients,³ with or without coexisting lymphadenopathy. Exclusive extranodal RDD, as in this patient, is rare. Extranodal RDD usually affects the skin, upper respiratory tract, central nervous system, and bone. Only 5% of extranodal RDD involves the skeleton.^{4,5} Demicco et al⁶ reviewed 15 cases of primary

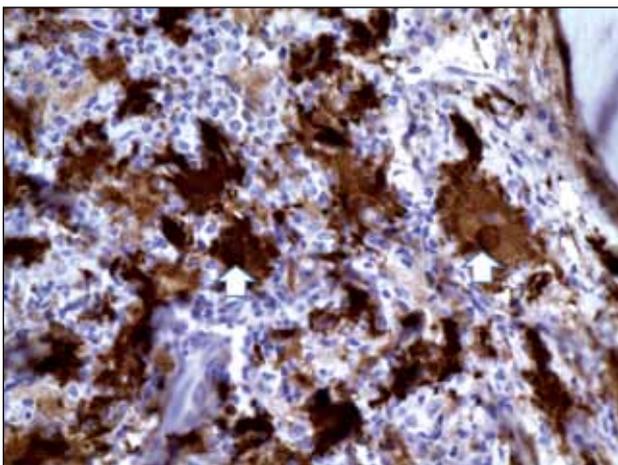


Figure 5. The large histiocytes are immunohistochemically positive for S100 protein (arrows) [original magnification, x 400].

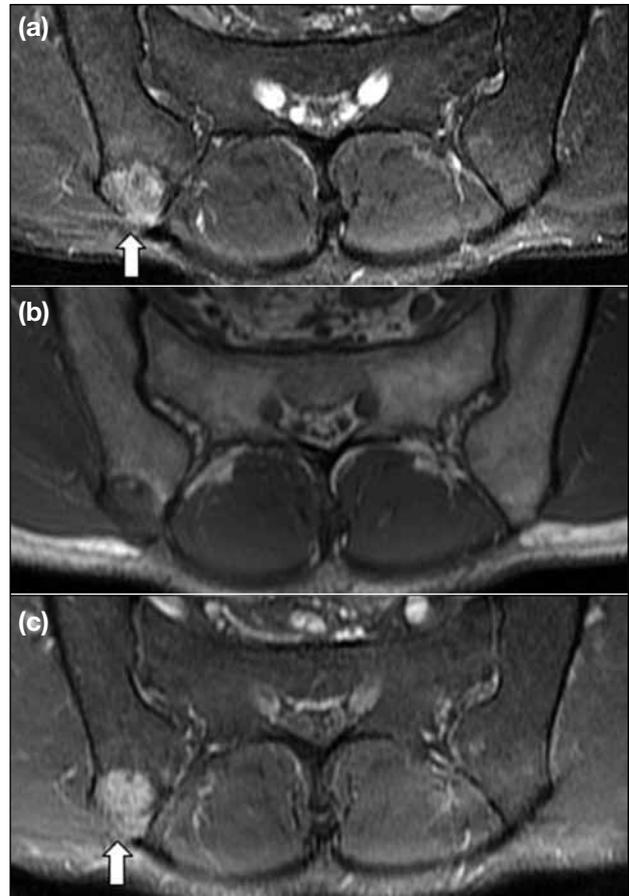


Figure 6. Follow-up magnetic resonance images taken after 15 months. (a) T2-weighted fat-saturated, (b) T1-weighted, and (c) T1-weighted post-contrast fat-saturated images show heterogeneous gadolinium enhancement. New cortical disruption is seen at the posterior part of the lesion (arrows) that is more T2-hyperintense than in the magnetic resonance image taken at presentation (Figure 1).

RDD of bone. The lesions most frequently occurred in the appendicular skeleton, especially the long bones of the upper and lower limbs. The skull, maxilla, and sacrum were rare sites of involvement.

RDD is considered to be a benign disease that usually resolves spontaneously. The prognosis is generally good for most patients. A minority of patients may have disease recurrence or metastases for which the cause remains elusive.

In Demicco et al’s series,⁶ 10 patients had radiographs taken and nine of them were lytic in nature. The margins of the lesions could be either well-defined or ill-defined. Sclerotic rims were present in some of the lesions.

Zhu et al⁷ have also studied the imaging characteristics of 13 patients with extranodal RDD. Six patients had CT performed, which showed that half of the lesions were mixed iso/hyperdense. The remaining lesions could be either homogeneously hyperdense or homogeneously hypodense. Five of the six lesions had bony destruction. On MRI, the lesions showed variable signal intensities in both T1-weighted and T2-weighted sequences. Gadolinium enhancement pattern also varied. Cortical break was not a consistent feature. The imaging features might change over time as in this patient.

Positron emission tomography/CT and bone scan may show hypermetabolic activity in the lesions. These imaging modalities have additional value in identifying synchronous lesions.⁸

There is no specific imaging characteristic that can lead to a confident diagnosis of RDD. Biopsy of the lesion and pathological examination are usually required to reach the diagnosis. Typical pathological findings include tissue fibrosis with marked expansion of sinus by histiocytes, plasma cells, and lymphocytes. Emperipolesis of intact lymphocytes and plasma cells by histiocytes can be present. Positive S100, CD163, and CD68 staining of histiocytes is a characteristic feature.^{2,9}

If the patient is asymptomatic, conservative treatment and close follow-up are usually adequate. Definitive treatment of RDD is surgical resection.^{10,11} Other treatment options include high-dose steroids, radiation therapy, chemotherapy, cryosurgery, and immunomodulatory agents.¹²

CONCLUSION

We have reported a case of RDD occurring primarily in bone. RDD is a rare disease and skeletal involvement is uncommon. We present this report to remind the readers

to include RDD in the differential diagnosis of focal osseous lesions. Imaging features of RDD can change over time as illustrated by this patient.

DECLARATION

The authors declare that they have no conflicts of interest.

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