

# Extranodal Rosai–Dorfman disease in a carpal bone

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## ABSTRACT

We report a case of extranodal Rosai–Dorfman Disease (RDD) of the scaphoid in a 52-year old female. The patient presented with pain, swelling, and tenderness on deep palpation of the left wrist. Clinoradiological diagnosis was osteomyelitis or tenosynovitis and curettage was performed on the lytic lesion over scaphoid to procure tissue. Diagnosis was made by histomorphology supported by immunostaining. The patient was managed conservatively with resolution of the lesion.

**Key words:** Lymphophagocytosis, RDD, scaphoid bone, S-100 protein

## INTRODUCTION

**S**inus histiocytosis with massive lymphadenopathy (SHML), also known as Rosai–Dorfman Disease (RDD), is a rare hematopoietic disorder of primarily cervical lymph nodes, but it can also involve other nodal and some extranodal sites. The extranodal involvement occurs in 25% of cases.<sup>1</sup> Osseous involvement as the only manifestation is unusual.<sup>2</sup> We report a rare case of RDD involving scaphoid.

## CASE REPORT

A 52-year-old female presented with a 2-year history of dull aching pain in the left wrist. Clinical examination revealed a soft swelling over wrist [Figure 1] and there was tenderness in the scaphoid fossa on deep palpation. The range of motion (ROM) of the wrist joint was reduced to 55° in palmarflexion and 50° in dorsiflexion. There was no epitrochlear, cervical, axillary, popliteal, or inguinal lymphadenopathy, no hepatosplenomegaly, and no palpable abdominal mass. The white blood cell count was 5600/mm<sup>3</sup> (normal 4,500–13,000/mm<sup>3</sup>), with normal differential count. A mild normochromic, normocytic anemia was

noted, with hemoglobin of 120 g/l (normal 135–180 g/l) and a hematocrit of 0.35 (normal 0.41–0.53). Electrolyte levels and liver function test results were all within normal limits. The erythrocyte sedimentation rate (ESR) was elevated to 40 mm per hour (normal 0–15 mm per hour). The C-reactive protein (CRP) was also elevated to 50 mg/l (normal 0–10 mg/l). Serum protein electrophoresis showed the beta and gamma fractions slightly elevated to 15% (normal 6–12%) and 21% (normal 8–18%), respectively. Antinuclear antibody testing had a negative finding. Plain radiographs revealed a small lytic lesion over the scaphoid bone [Figure 2a]. The computed tomography (CT)/magnetic resonance imaging (MRI) were not initially advised due to financial constraints. The X-ray showed a lytic lesion over scaphoid with a clinoradiological diagnosis of osteomyelitis or tenosynovitis. The clinical diagnosis included osteomyelitis, infective tenosynovitis. A fine needle aspiration cytology (FNAC) was done from the lesion over scaphoid. Smears revealed scanty material comprising few lymphocytes, histiocytes, and plasma cells. Some of the histiocytes showed lymphophagocytosis. Curettage was performed by opening a dorsal 3 mm cortical window through the cartilage, on the lesion over scaphoid and the tissue procured was sent for biopsy.

The curetted specimen was received in the form of small bits of curette material measuring 0.8 cm × 0.5 cm × 0.5 cm. Histopathological picture was similar to that seen in lymph nodes. The histology section revealed a bit of bony tissue with lymphoid follicles having prominent germinal centers, admixed with plasma cells and histiocytes [Figure 3]. On higher magnification, the histiocytes were large with abundant pink cytoplasm and evidence of emperipolesis [Figure 3, inset]. The histiocytes were strongly immunoreactive for S-100 protein and negative for CD1a. From the above findings, a diagnosis of RDD was made. The detailed clinical examination and MRI of chest and

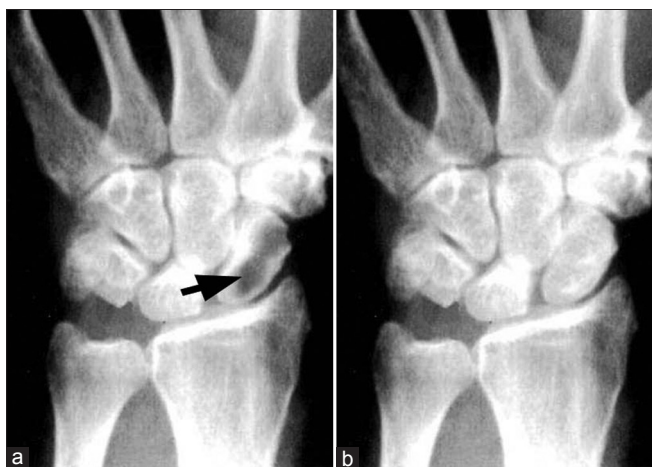
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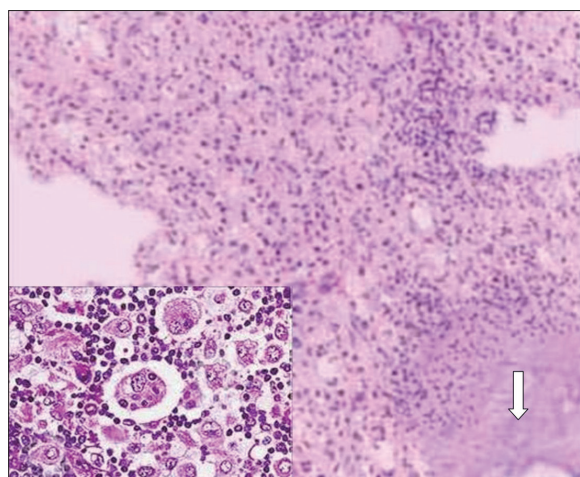
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**Figure 1:** Clinical photograph of patient showing swelling (black arrow) over the left wrist



**Figure 2:** Plain radiograph of hand showing a small lytic lesion over the scaphoid bone before treatment (a); no lesion over the scaphoid bone after conservative treatment (b)



**Figure 3:** Photomicrograph showing bits of bony tissue (arrow) with lymphocytes arranged in follicular pattern and histiocytes (H and E  $\times 100$ ); inset shows histiocytes with evidence of emperipolesis (H and E  $\times 400$ )

abdomen was done to rule out the possibility of any nodal involvement. The findings on radiographs, MRI and CT were negative to rule out involvement of CNS, lung and liver. The patient was subjected to conservative treatment of immobilization of affected hand by scaphoid cast for 6 weeks and a short course of analgesic for 1 week. A repeat X-ray and ESR and CRP tests were done every 3 months.

Six months prospectively, the patient was pain free, and 9 months prospectively, the ESR, CRP values, and X-ray [Figure 2b] were normal.

## DISCUSSION

SHML (RDD) was first described as a condition of unknown etiology involving the lymph nodes, but it was soon recognized that the disease process may involve other organ systems, with the skeleton being the fifth common site.<sup>3,4</sup> Among the extranodal sites, the head and neck region including the sinuses, orbit, and ear are commonly involved.<sup>5</sup> Rare extranodal sites include the soft tissue, skin, upper respiratory tract, gastrointestinal tract, breast, bones, and the central nervous system.<sup>5</sup> About 5% of extranodal cases involve the bone; they are usually associated with extraosseous manifestations.<sup>4,5</sup> Primary solitary osseous involvement is very uncommon. RDD with primary solitary involvement of triquetrum was reported by Sir Young Loh *et al.*<sup>2,5</sup> We report a case of RDD of the scaphoid bone.

Radiologically, these lesions of small bones are typically lytic.<sup>3,6</sup> Clinically, they may be confused with osteomyelitis and other inflammatory conditions. Typical histomorphology supported by immunostaining for S-100 protein and CD1a can help us in arriving at a diagnosis of RDD.<sup>4,6</sup> The differential diagnosis includes osteomyelitis, lymphoma, metastatic carcinomatous deposits, and infective tenosynovitis (nonspecific and tubercular).<sup>5,7</sup> RDD shows dilated sinuses filled with lymphocytes and numerous histiocytes with intact lymphocytes within their cytoplasm, a feature known as emperipolesis or lymphophagocytosis. The germinal centers may be hyperplastic, or even sparse to absent. In lymphoma, the above architecture is effaced. Osteomyelitis usually shows chronic and acute inflammatory cells in a necrotic background. Infective synovitis contains synoviocytes, acute and chronic inflammatory cells. Tubercular synovitis contains tubercular granuloma. Metastasis is ruled out by absence of malignant epithelial cells.<sup>7</sup>

RDD is self-limiting in most patients.<sup>7</sup> Patients with RDD without vital organ involvement should be followed closely without any active therapy. Patients with systemic symptoms or those with sudden enlargement of nodes may be treated with prolonged course of low-dose prednisone.<sup>8</sup>

For patients with vital organ compression, like central nervous system, lung, or liver, surgery and high-dose corticosteroids should be tried first, but radiotherapy may be needed in resistant cases or wherever surgery is not feasible.<sup>5,8</sup> RDD in small bones is usually managed by conservative treatment with prolong followup.<sup>7,8</sup> Role of radiotherapy and chemotherapy is on trial in case of extensive lesions.<sup>9</sup> Although the etiology of RDD is unknown, the leading hypothesis is that it is of infectious origin.<sup>1,10</sup> It has been suggested that RDD may be linked to Parvovirus infection, as Parvovirus B19 capsid proteins have been isolated in some cases.<sup>10</sup>

We present this case to raise awareness of the disease and to include it in the differential diagnosis of lytic lesions of carpal bones.

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