

Rosai-Dorfman Disease Presenting as a Primary Mesenteric Mass in an Elderly Patient: A Rare Case Report

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ABSTRACT

Rosai-Dorfman Disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare benign non-Langerhans cell histiocytic proliferative disorder that commonly presents with painless cervical lymphadenopathy in young adults. Extranodal involvement occurs in nearly 40% of cases; however, isolated abdominal disease is exceptionally uncommon and may mimic neoplastic lesions radiologically. This case report describes a rare case of mesenteric RDD presenting as an abdominal mass clinically and radiologically suggestive of a Gastrointestinal Stromal Tumour (GIST). A 66-year-old male presented with a 2-week history of abdominal pain and dysuria. Imaging revealed a progressively enhancing soft-tissue mass in the left lumbar region supplied by the superior mesenteric artery, favouring a diagnosis of GIST. Surgical exploration identified a highly vascular mesenteric mass attached to the small bowel. Histopathological examination showed marked lymphoid follicular hyperplasia with sinusoidal proliferation of large histiocytes exhibiting emperipolesis. Immunohistochemistry demonstrated positivity for S100, CD68, and Cyclin D1, while CD15 and CD30 were negative, excluding Hodgkin lymphoma. These findings confirmed the diagnosis of mesenteric RDD. This case highlights the diagnostic challenge posed by isolated abdominal RDD due to its non-specific clinical and radiologic features. Recognition of characteristic histopathological findings and immunophenotypic profile is essential for accurate diagnosis and appropriate management. RDD should be considered in the differential diagnosis of unexplained abdominal masses to avoid misdiagnosis and unnecessary aggressive treatment.

Keywords: Emperipolesis, Gastrointestinal stromal tumours, Histiocytosis, Immunohistochemistry, Lymphoid follicular hyperplasia, Mesenteric diseases, S100 proteins

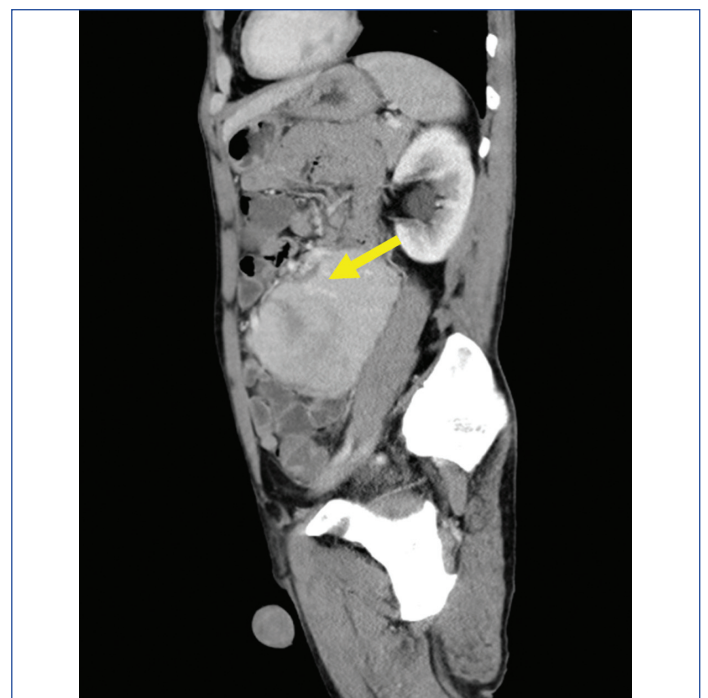
CASE REPORT

A 66-year-old man presented with a 2-week history of abdominal pain and dysuria. The abdominal pain was localised to the left lumbar region, insidious in onset, dull aching in nature, non-radiating, and progressively worsening. He had no significant past medical history. The patient also complained of constipation and painful defecation. There was no fever, nausea, vomiting, flank pain, loose stools, reduced urine output, difficulty in passing urine, haematuria, melena, or frank blood in stools. There was no history of loss of appetite or loss of weight. Past medical, surgical and family histories were unremarkable.

On examination, a firm, mobile, mildly tender mass measuring approximately 8×5 cm was palpated in the left iliac fossa. It was not attached to surrounding structures, and there was no local rise in temperature. Routine haematological and biochemical investigations, including Complete Blood Count (CBC), Erythrocyte Sedimentation Rate (ESR), C-reactive Protein (CRP), Liver Function Tests (LFT), and Renal Function Tests (RFT), were within normal limits.

Contrast-enhanced CT of the abdomen revealed a large, progressively enhancing soft-tissue mass in the left lumbar region supplied by the superior mesenteric artery [Table/Fig-1]. The radiological impression suggested a GIST. Diagnostic laparoscopy revealed a 10×9 cm mass arising from the small bowel border in the left lumbar region. It was highly vascularised, and no adhesions were noted. The rest of the bowel was normal. Tru-cut biopsy was taken for diagnosis, which was reported as reactive lymphadenitis on histopathological examination with no evidence of tumour or any specific pathology. Surgical resection of the mass was performed, along with resection of the proximal and mid-small bowel with re-anastomosis under general anaesthesia. Grossly, the mesenteric mass weighed 311 g and measured 11.6×9.4×7 cm in size representing the definitive

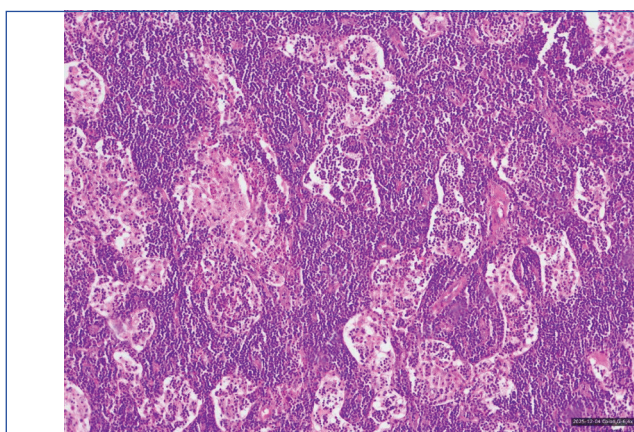
post-excision dimensions. The apparent difference reflects the distinction between intraoperative estimation and formal pathological measurement. The external surface appeared smooth. The cut section showed a well-circumscribed mass with grey-white homogenous areas abutting the capsule. No areas of necrosis, haemorrhage, or calcification were noted. The resected bowel sections were 76 cm in length and appeared unremarkable, and on cut section showed normal mucosal folds. No polyps, ulceration, growth or thinning of



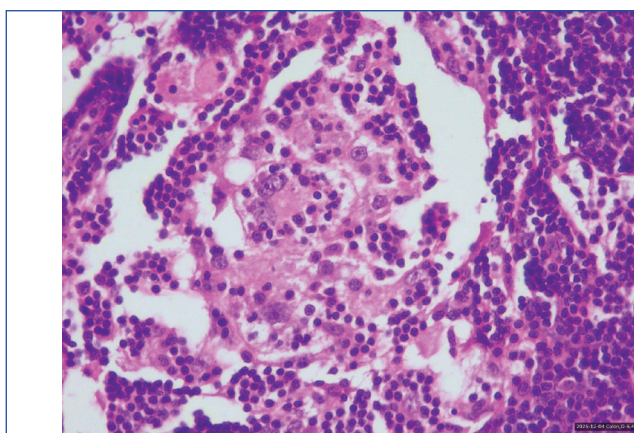
[Table/Fig-1]: Contrast-enhanced CT showing an enhancing soft-tissue mass in the left lumbar region (arrow).

the mucosa were identified. Ten lymph nodes were dissected and, on cut section, showed grey-tan areas.

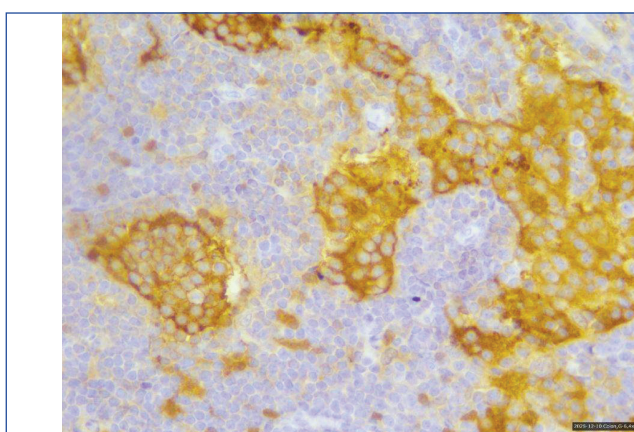
On microscopy, the section showed marked lymphoid follicular hyperplasia with sinusoidal accumulation of histiocytes with enlarged, large vesicular nuclei and abundant eosinophilic cytoplasm with prominent nucleoli, many showing emperipolesis with engulfed lymphocytes, plasma cells, and occasional neutrophils surrounded by reactive lymphoid follicles [Table/Fig-2,3]. Immunohistochemistry demonstrated positive staining for S100, CD68, and Cyclin D1 (nuclear) among the large cells [Table/Fig-4]. A differential diagnosis of Hodgkin lymphoma was considered in view of the mixed inflammatory background. CD15 and CD30 were negative, ruling out Hodgkin lymphoma. CD1a and Langerin were not performed because the lesion had characteristic morphology. The histopathological and immunohistopathological findings suggested a diagnosis of mesenteric RDD. The patient remained asymptomatic on eight months follow-up.



[Table/Fig-2]: Photomicrograph showing dilated sinusoids containing large histiocytes surrounded by lymphoid aggregates (H&E, x200).



[Table/Fig-3]: Photomicrograph showing large histiocytes within the sinusoids demonstrating emperipolesis, with engulfed lymphocytes and neutrophils (H&E, x200).



[Table/Fig-4]: Photomicrograph demonstrating large histiocytes with positive S100 expression (Immunohistochemistry, x200).

DISCUSSION

The RDD is a rare non-Langerhans cell histiocytic disorder that involves the lymph nodes. It usually presents painless lymphadenopathy accompanied by constitutional symptoms such as fever, fatigue, and weight loss, occurring more commonly in young adults. The exact pathogenesis of RDD remains unclear [1].

Extranodal involvement is observed in nearly half of patients and may affect the skin, bone, soft-tissues, central nervous system, gastrointestinal tract, genitourinary system, endocrine system, orbit, and other organs [2]. Although, nodal disease is the most common presentation, isolated extranodal involvement can occur and may pose significant diagnostic challenges both clinically and radiologically. Abdominal RDD is particularly uncommon, with only a few cases documented in the literature [3,4]. Most commonly reported abdominal sites include the liver, pancreas, kidneys, retroperitoneum, and bowel [5]. Gastrointestinal tract involvement is rare, accounting for less than 1% of extranodal RDD cases [6].

Intra-abdominal RDD typically manifests as multiple mass lesions, often measuring less than 2.5 cm. In patients without a prior diagnosis of RDD, the imaging appearance is nonspecific, frequently necessitating histopathological evaluation for confirmation. However, in individuals with a known history of RDD, recognition of abdominal involvement in the differential diagnosis may prevent unnecessary invasive procedures and can help in deciding appropriate management.

Primary mesenteric RDD in the absence of nodal disease is exceedingly rare. Such lesions may closely resemble mesenchymal tumours, especially GIST, owing to their soft-tissue density, mass-like appearance, and proximity to bowel loops [6]. This radiologic overlap can complicate pre-operative diagnosis. In our 66-year-old patient, the large (11 cm) mass attached to the small bowel was initially interpreted as GIST on radiological imaging, similar to a previous report in which FDG-avid mesenteric or intestinal masses required surgical excision for definitive diagnosis [6].

Histologically, RDD is characterised by sheets of large histiocytes with abundant cytoplasm demonstrating emperipolesis within a background of mixed inflammatory infiltrate. Extranodal lesions are reported to share similar features but display increased fibrosis and sclerosis, fewer histiocytes, and less conspicuous emperipolesis compared with nodal disease. Immunohistochemistry is essential for confirmation [7,8]. The large histiocytes with emperipolesis show positivity for S100, CD68, and CD163, and negativity for CD1a, ALK, and langerin [1,2]. In present case, the resection specimen demonstrated lymphoid hyperplasia with sinus histiocytosis and prominent emperipolesis, suggesting a conglomerate nodal mass rather than a purely extranodal lesion.

While classic nodal RDD predominantly affects young males, abdominal RDD is more often encountered in older adults. The clinical course is variable; some cases have shown to undergo spontaneous remission, whereas large or symptomatic lesions, particularly those exceeding 10 cm, require surgical intervention. Recent consensus recommendations emphasise the potential role of targeted therapies, including MEK inhibitors, in cases harbouring MAPK pathway alterations, especially in multifocal or refractory diseases [2,7]. Nevertheless, complete surgical excision remains the treatment of choice for mass lesions such as ours. Long-term follow-up is advisable, as recurrence rates of up to 30% have been reported, although many patients achieve sustained remission following resection [2,9]. At eight months of follow-up, the patient remained asymptomatic with no evidence of recurrence.

Given the rarity of isolated abdominal RDD and its non-specific radiologic features, neoplastic pathology is frequently considered first in abdominal and pelvic masses [10]. In the present case, the initial biopsy suggested a diagnosis of reactive lymphadenitis. Definitive diagnosis was established only after surgical excision,

where identification of emperipolesis along with the characteristic immunophenotypic profile confirmed RDD.

CONCLUSION(S)

This case highlights a rare and atypical presentation of extra-nodal RDD. The absence of nodal involvement and its unusual abdominal location underscore the importance of considering RDD in the differential diagnosis of abdominal masses. Early recognition and accurate histological diagnosis are essential to avoid mismanagement. Additionally, this case emphasises the challenge posed by extranodal RDD due to radiological and clinical features with inflammatory and neoplastic conditions. The addition of appropriate immunohistochemical evaluation plays a pivotal role. Increased awareness of such rare presentations can aid pathologists and clinicians in preventing unnecessary aggressive management and ensuring optimal patient care.

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