

### **Case Report 3**

#### **Cutaneous Rosai-Dorfman Disease presenting as a flank swelling**

**Prof. Saleh M. AISalamah MBBS, FRCS**

Professor of Surgery and Consultant General & Laparoscopic Surgeon  
Department of Surgery, College of Medicine, King Saud University  
Head University Surgical Unit, King Saud Medical City, King Saud University, Riyadh, KSA

**Dr. Muhammad Abdullah FCPS, FRCS**

Senior Registrar  
Department of Surgery, College of Medicine, King Saud University  
University Surgical Unit, King Saud Medical City, Riyadh, KSA

**Dr. Reem Abdullah AISalamah MBBS**

Resident in Dept. of Surgery, King Saud Medical City, Riyadh, KSA

**Dr. Abdul Rehman Abdullah Al Hadlaq MBBS, MD**

Consultant Dermatologist  
Department of Dermatology, King Saud Medical City, Riyadh, KSA

**Ebtehal Sulaiman Al Mogbel MBBS, SBFM, ABFM**

Rotating Senior Registrar Family Medicine, Al Qassim University  
King Saud Medical City, Riyadh, KSA

**Dr. Ghadah Mohammed Al Yahya MBBS, MS**

Registrar in Dept. General Surgery  
King Saud Medical City, Riyadh, KSA

#### **Abstract**

Rosai-Dorfman disease (RDD) is primarily a disease of lymph nodes caused by accumulation of excessive histiocytes. The extra-nodal sites are simultaneously involved in almost half the cases. However, presentation with isolated extra-nodal sites is uncommon and disease confined to skin and soft tissues is a rare entity. We report RDD in a 33 year old man who presented as a flank mass and was a diagnostic challenge for the surgical team. This case report strengthens the evidence that RDD should be added to the list of rare causes of skin and soft tissue swellings and informs how to manage such patients.

**Key words:** Rosai- Dorfman Disease, Sinus Histiocytosis with massive lymphadenopathy, Extranodular Rosai-Dorfman Disease

#### **Corresponding Author:**

**Prof. Saleh M. AISalamah MBBS, FRCS**

Professor of Surgery and Consultant General & Laparoscopic Surgeon  
Department of Surgery, College of Medicine, King Saud University  
Head University Surgical Unit, King Saud Medical City, King Saud University, Riyadh, KSA  
P.Box 261283, Riyadh 11342, KSA  
Tel: +966 (11) 4671 585  
Fax: +966 (11) 4679 493  
Email: [smsalamah@hotmail.com](mailto:smsalamah@hotmail.com)

## Introduction

Rosai-Dorfman disease (RDD) was first described as a distinct clinic-pathological entity in 1969 by Rosai and Dorfman.<sup>(1)</sup> It is a benign disease characterized by over production and accumulation of histiocytes primarily in the lymph nodes of the body with cervical region being the commonest. In 43% of cases, extra-nodal sites are involved simultaneously and in only 23% does isolated extra-nodal RDD occur.<sup>(2)</sup> Various extra-nodal sites have been reported to be affected including skin, eye and adnexa, paranasal sinuses, genitourinary system, central nervous system, bone, breast, soft tissues and thyroid.<sup>(3)</sup> The skin and upper respiratory tract remain the most common sites of extra-nodal involvement, each accounting for approximately 11% of cases.<sup>(4)</sup> Extra-nodal involvement confined only to the skin is an even rarer entity.<sup>(4)</sup> We report an unusual case of extra-nodal RDD presenting as a soft tissue mass in the left lumbar region.

## Case Report

A 33 year old male presented with complaints of left flank swelling for 6 months duration which had progressively enlarged and now started to cause pain. The patient suffered from asthma but had no other symptoms on systemic inquiry. Clinical examination revealed stable vital signs with normal body temperature. Chest and neck examination was unremarkable. Abdominal examination showed a 7 x 3 cm oval shaped swelling at the left lumbar region involving the skin and subcutaneous tissues which was firm in consistency, non-mobile and non-tender. The overlying skin was thickened with nodularity and had yellow-brown papules with no discharge. Axillary and inguinal lymph nodes were not palpable. All haematological and biochemical laboratory work up was within normal limits. An ultrasound scan (US) followed by a Magnetic Resonance Imaging (MRI) scan of the region was done which showed 7.1 x 3.7 x 4.7 cm lesion extending to deep subcutaneous fat with surrounding infiltration and stranding producing fluffy margin but did not reach the ipsilateral flank or gluteal muscles (Fig 1). The pelvic bones and skeletal muscles showed no focal lesion or signs of involvement. The findings were suggestive of chronic inflammatory process but the possibility of a soft tissue tumour could not

be excluded. Hence, it was planned to excise the lesion under the general anaesthetic. Per-operatively a very firm swelling was found uniformly involving the skin and subcutaneous tissues until the deep fascia and was very vascular. The swelling was excised with adequate margins and the histopathology showed extra-nodal cutaneous RDD. The patient made an uneventful recovery but developed a residual nodule over the scar which revealed similar histopathology on punch biopsy. It was most probably a recurrence in a small area but that settled without any further treatment. The patient is currently under long term dermatology surveillance for further recurrence.

## Discussion

RDD is also known as sinus histiocytosis with massive lymphadenopathy. It is classified as either nodal or systemic (cutaneous, respiratory and /or osseous) and typically presents with generalised lymphadenopathy and a polymorphic histiocytic infiltration of the lymph node sinuses.<sup>(5)</sup> Systemic RDD is more prevalent while the pure cutaneous form accounts for only 3% of RDD and involves only the skin and adjacent soft tissues without associated lymphadenopathy or systemic symptoms. The lesions of cutaneous RDD show no predilection for any specific location, sometimes it could be localized in the soft tissue and manifest as subcutaneous mass or panniculitis.<sup>(6)</sup> This occurrence of extranodal soft tissue RDD may masquerade as a sarcoma and challenge the clinical acumen of a surgeon where the diagnosis is very difficult without associated lymphadenopathy.

The presentation of our patient was similar to some findings of the previous reports with no features to suggest cutaneous RDD. Soft tissue RDD is particularly challenging to diagnose short of the excision biopsy because the characteristic cytomorphology is rarely obtained on fine needle aspiration cytology (FNAC) and the imaging modalities report a wide differential diagnosis. Although the diagnosis of classical RDD has being reported with FNAC by comparing the aspirates from the involved node and the extranodal site<sup>(7)</sup> but such has not been reported with pure cutaneous lesions. US and MRI can describe a detailed extent of the lesions but a diagnosis cannot be confirmed. The microscopic

hallmark is large histiocytes with abundant pale cytoplasm, round vesicular nuclei and showing lymphocytphagocytosis or emperipolesis (Figs 2 & 3). Extranodal cases are diagnosed easily if a classical nodal disease has been documented. However, in absence of diagnostic lymph nodes, a greater number of characteristic histiocytes with emperipolesis are required for extranodal disease to be defined. An Immunohistochemical diagnostic criterion of RDD is cytoplasmic positivity of histiocytes for S-100 protein. <sup>(8)</sup>The microscopic examination of the resected swelling of this patient revealed infiltration of lymphocytes, plasma cells and histiocytes with abundant eosinophilic cytoplasm and large non-cleaved vesicular nucleus with prominent nucleoli. These cells showed emperipolesis and strong immunoreactivity to S100 immunostain.

Considering the rarity of the lesions and preoperative diagnostic difficulties, surgical resection is probably the best option. In our patient the resected excision margins were

involved and the patient developed a residual nodule but it settled with observation. Due to the rarity of the disease and its self-limited course, no treatment protocol has been established for RDD. <sup>(9)</sup>In symptomatic patients without spontaneous resolution, surgical excision is usually performed. A varying response rate has been reported with steroids, alkylating agents and IFN $\alpha$  in symptomatic cases. <sup>(10)</sup>The role of radiotherapy is still poorly understood, with some reports describing full resolution while others showed no response. <sup>(5)</sup> Although cutaneous RDD presenting as a solitary soft tissue swelling has previously been published in the literature, this case report highlights that cutaneous RDD should be one of the uncommon differential diagnosis for any lesion presenting as soft tissue swelling with cutaneous involvement. This report also shows that a confirmed diagnosis is difficult to reach without an excision and histopathological assessment.

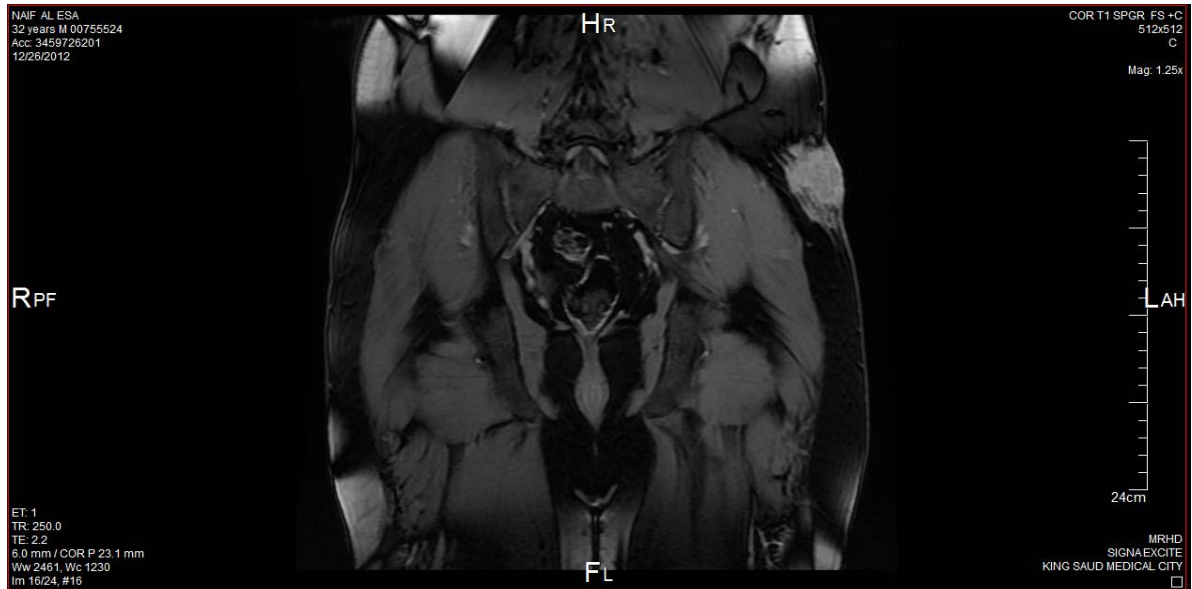
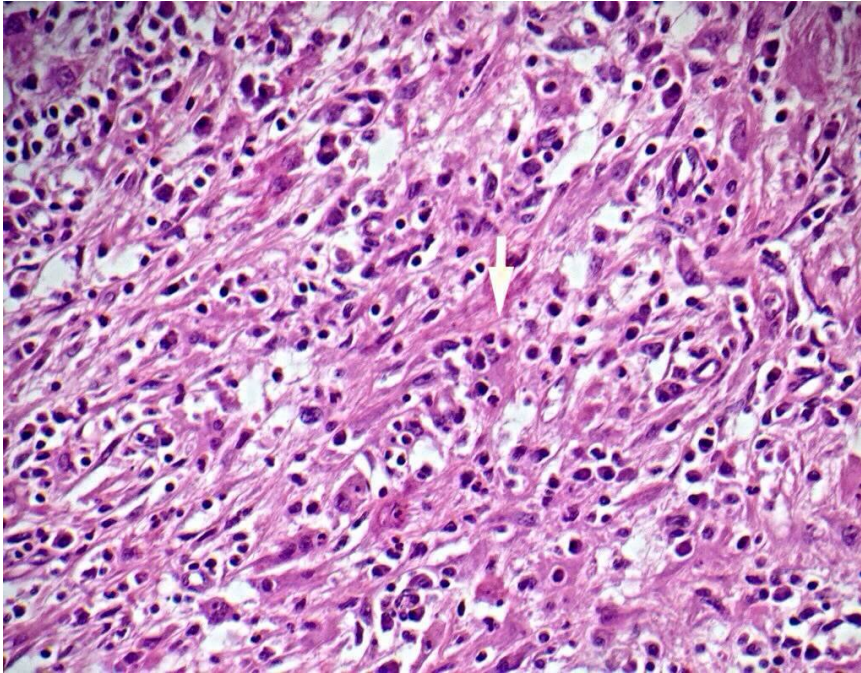
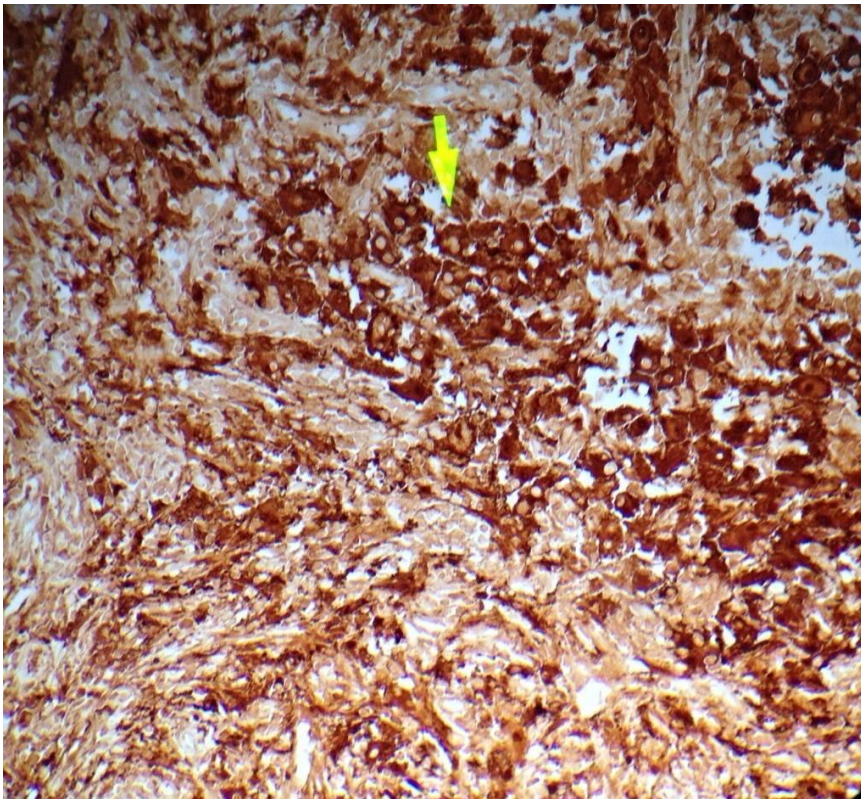


Figure 1: MRI showing the swelling in left lumbar region



**Figure 2: Histiocytes with evidence of engulfed intact lymphocytes (white arrow), H&E, X 400**



**Figure 3: Histiocytes with evidence of emperipolesis (yellow arrow), S100, X 400**



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