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When Histology Speaks: A Rare Case Of Soft Tissue Rosai-Dorfman Disease Masquerading As Hematoma In An Adolescent

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Abstract

Rosai -Dorfman Disease (RDD), also known as Sinus histiocytosis with massive lymphadenopathy, is an uncommon disorder of histiocytic overgrowth. While the classical site of involvement is lymphnode, 40% of the cases involve extranodal sites of which skin involvement is common. Extranodal RDD can pose significant diagnostic challenges due to its rarity, nonspecific clinical presentation, and its tendency to mimic infectious, inflammatory, or neoplastic processes both clinically and radiologically. Isolated soft tissue involvement is especially uncommon and may be mistaken for sarcoma, abscess, or hematoma, particularly when systemic signs are absent. In such cases, the diagnosis relies heavily on histopathological examination. Herein, we report a rare case of isolated extranodal RDD involving the soft tissue of the thigh in a 16-year-old patient, presenting without lymphadenopathy or systemic symptoms. This case highlights the importance of maintaining a high index of suspicion for RDD in the differential diagnosis of soft tissue masses and underscores the diagnostic value of histopathology and immunohistochemistry in confirming this unusual presentation.

Keywords: NIL

Introduction

We present a case of 16 year old patient presented with painful thigh mass for 6 months,with gradual increase in size. There was no history of trauma or fever. No other swellings/lymph nodes were noted on extensive physical examination. Clinical diagnosis was made as hematoma and FNAC was avoided. Radiological evaluation was inconclusive.Routine lab results were normal. Wide local excision was done. Gross examination grey yellow solid homogenous area measuring 6x4x3cm, surrounded by normal cutaneous areas. Microscopic examination showed skin with dense inflammation composed predominantly histiocytes exhibiting emperipolesis having intact lymphocytes, plasma cells, occasional neutrophils and red blood cells in their cytoplasm (Fig.1 and 2). Peripheral areas showed reactive fibrosis with focal collagen deposition. For confirmation, Immunohistochemistry for S100 was done (Fig.3). It showed diffuse cytoplasmic positivity in histiocytes exhibiting emperipolesis.Post operative period was uneventful and the recovery was successful.

Fig.1 & 2. showing histiocytes exhibiting emperipolesis with intact lymphocytes, neutrophils and red blood cells in their cytoplasm

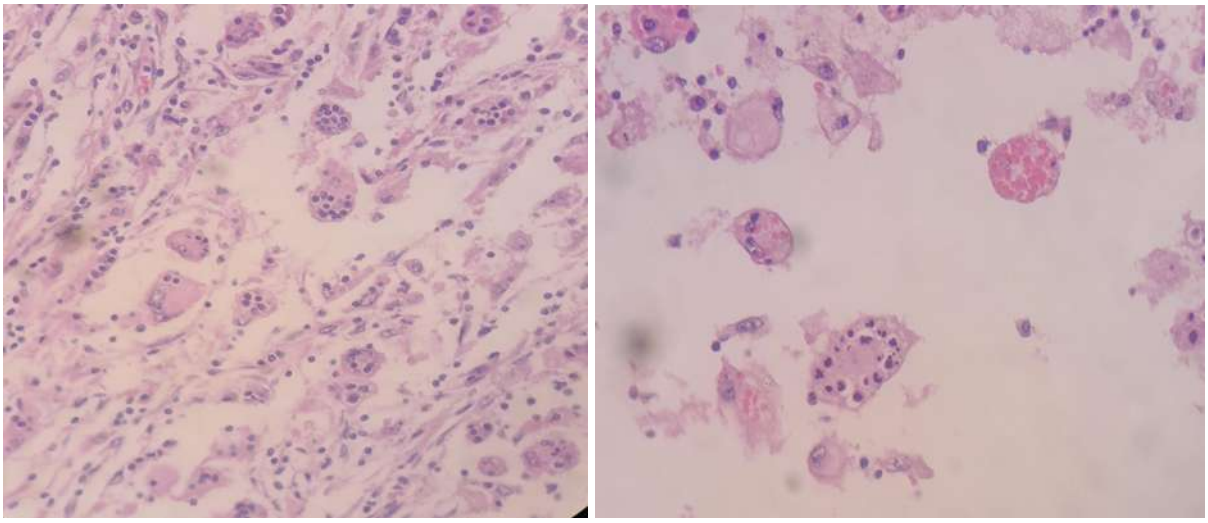
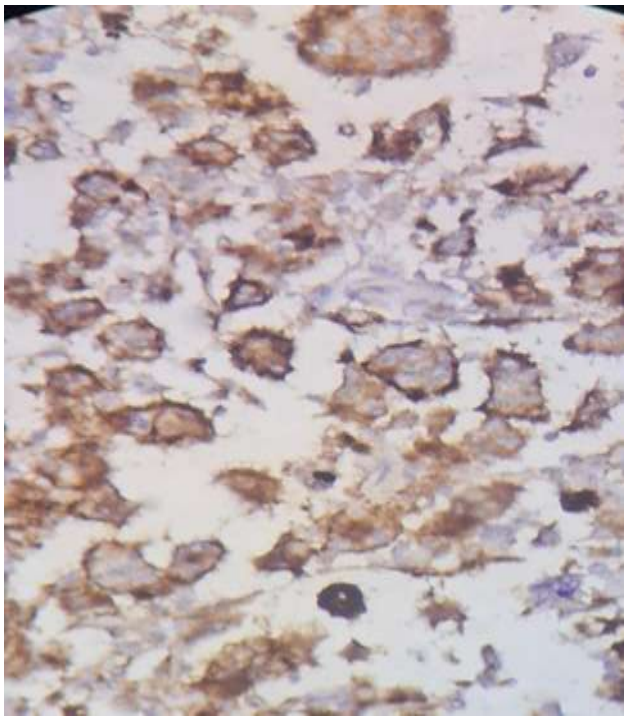


Fig.3 Immunohistochemistry S100 highlighting the histiocytes with emperipolesis .



Discussion:

Rosai Dorfman Disease was regarded as a separate clinical entity from 1969 and has a prevalence of 1:2,00,000 in the United States[1,2] . Though it is benign and self limiting [3], a small proportion of cases show relapsing course. The primary site of involvement is lymph nodes in most cases but around 40% of the cases are seen in extra nodal locations. Next to lymph nodes, skin and soft tissue are commonly involves followed by nasal cavities and orbit [4].Although there have been studies linking it to viruses and reactive processes, the exact pathogenesis remains unexplored[5]. RDD is seldom suspected preoperatively. Sandoval-Sus et al.,quoted that extranodal involvement can occur without any lymphadenopathy, and in such cases, the disease may remain unrecognized until histologic examination

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[6].Histopathologically, RDD is characterized by the presence of large pale-staining histiocytes with intact inflammatory cells within their cytoplasm—a phenomenon known as emperipolesis. This is not to be confused with phagocytosis, as the engulfed cells remain viable. The histiocytes are immunopositive for S100 and CD68, and negative for CD1a, which helps distinguish RDD from Langerhans cell histiocytosis [4]. In our case, the histological hallmark of emperipolesis was clearly observed, and diffuse S100 positivity on immunohistochemistry confirmed the diagnosis.

Soft tissue RDD, particularly in young individuals, is a rare entity. A retrospective study by Pulsoni et al. observed that "cutaneous and soft tissue involvement tends to occur in a younger demographic and is often misdiagnosed as sarcoma, pseudotumor, or chronic abscess" [3]. . Radiological evaluation is usually inconclusive, reinforcing the need for surgical excision and histopathological diagnosis in ambiguous soft tissue masses. Although RDD is classified among histiocytic and dendritic cell neoplasms by the WHO, it is generally considered non-malignant. Recent studies have suggested clonal alterations in some cases, with mutations in MAPK pathway genes, including KRAS and NRAS, identified in a subset of patients [7]. However, the clinical relevance of these findings in isolated soft tissue RDD remains uncertain, and routine molecular testing is not currently warranted in the absence of atypical features. Therapeutically, most cases of RDD, especially localized extranodal ones, are self-limited and do not require systemic therapy. Surgical excision is often curative, particularly when the lesion is solitary and accessible, like in our scenario. This approach aligns with the findings of Komp et al., who emphasized that complete surgical excision is effective in cases of isolated extranodal RDD and the recurrence rate is minimal when margins are negative" [6]. Also, the site of extranodal involvement decides the prognosis. Involvement of kidney, liver or lower respiratory tract carries a poorer prognosis [8].

The differential diagnosis in soft tissue RDD is broad and includes both benign and malignant entities. Common differentials include nodular fasciitis, chronic abscesses, Langerhans cell histiocytosis, and inflammatory pseudotumors. These lesions can have overlapping clinical and radiologic appearances, often leading to misdiagnosis.None of the differentials show the characteristic emperipolesis. In cases where emperipolesis is not clearly made out, Langerhan cell histiocytosis may pose a difficulty in diagnosis. However the histiocytes which are immunopositive for S100 and CD68 are negative for CD1a, which helps distinguish RDD from Langerhans cell histiocytosis [3]. In our case, the histological hallmark of emperipolesis was clearly observed, and diffuse S100 positivity on immunohistochemistry confirmed the diagnosis.

Conclusion:

This case illustrates the importance of considering extra-nodal RDD in the differential diagnosis of isolated soft tissue masses, particularly in young patients. Histopathology and immunohistochemistry remain the cornerstone of diagnosis. Early recognition and complete surgical excision can lead to excellent outcomes in localized extranodal cases.Awareness of its rare extranodal presentations can prevent misdiagnosis and overtreatment, ensuring timely and appropriate management.

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