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Inguinal Lymphadenopathy in an Adult Male with Rossi-Dorfman Disease in Saudi Arabia: A Case Report

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Abstract

Rosai-Dorfman disease (RDD) is a rare disorder marked by painless, massive lymph node enlargement, typically affecting cervical nodes. We report a case of a 46-year-old male with RDD diagnosed after inguinal swelling and surgical excision. Despite its rarity in this location, histopathological examination confirmed the diagnosis, highlighting RDD's varied presentation. Further research is needed to understand its mechanisms and enhance treatment options.

Keywords: Histiocytosis; Non-Langerhans-Cell; Lymphatic Diseases; Rosai–Dorfman; Saudi Arabia

Introduction

Rossi-Dorfman disease, also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML), is a rare and enigmatic non-malignant histiocytic disorder characterized by painless, massive lymphadenopathy, often involving cervical nodes. Herein, we present the remarkable case of a 46-year-old male who presented with a yearlong history of inguinal swelling, prompting surgical excision. Following histopathological examination, an unexpected diagnosis of Rossi-Dorfman disease was discovered, which is rare in this anatomical location. Here, we provide a detailed account of the clinical presentation, diagnostic workup, and therapeutic management of this unique case.

Case Report

A 46-year-old male, who was not known to have any past medical illness or surgical history, presented with a 1-year history of progressive left inguinal swelling without any signs of an inflammatory process, at presentation, or the period prior to that. He denied any history of weight loss, appetite loss, or night sweats. There was no family or personal history of malignancy. On physical examination, the patient generally appeared well. There was a left inguinal round non-tender swelling approximately 5 × 3 cm, firm, and fixed. No overlying skin changes were observed. Blood laboratory results were unremarkable. An excisional biopsy was performed. Microscopic examination revealed lymphoid tissue and distended sinuses containing histiocytes (Figure 1). There is a polymorphous population of lymphocytes along with abundant plasma cells. There was no evidence of granuloma or malignancy. The patient was doing well upon follow-up in an outpatient clinic.

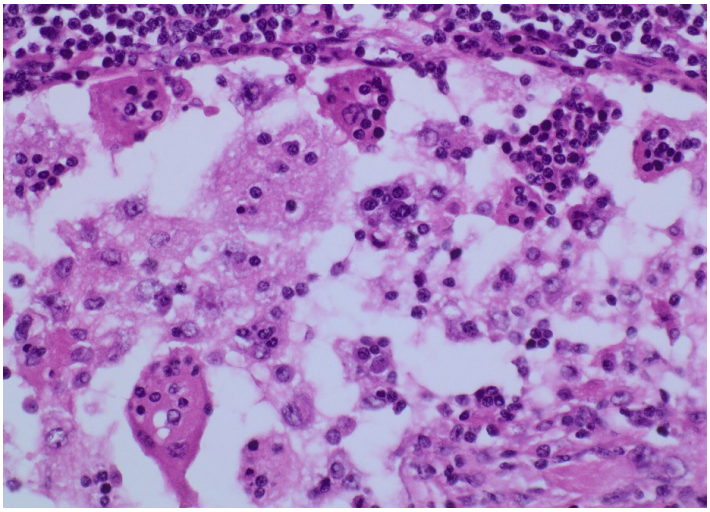

Figure 1: Lymphoid tissue and distended sinuses containing histiocytes.

Discussion

RDD is a rare disease with a prevalence of 1:200,000 and an estimated 100 new cases per year in the United States [1]. Although it has been reported up to the age of 74 years, it is more frequently observed in children and young adults (mean age, 20.6 years) [2]. RDD is more prevalent in people of African descent, whereas skin lesions are more prevalent in people of Asian descent [3].

According to the Histiocyte Society's classification system, the first cases of RDD are now regarded as the classic form of the disease: massive bilateral cervical lymphadenopathy accompanied by fever and leukocytosis, mostly affecting children and young adults, and frequently affecting African and male patients [4]. Nasal obstruction, epistaxis, and neck masses were the most typical presenting symptoms. Additionally, reported symptoms included vision loss, proptosis, eye tearing, hoarseness, and hyposmia [5-7]. RDD most frequently manifests as cervical lymphadenopathy. The head and neck, intracranium, bone, heart, skin, parotid gland, periodontium, orbit, thyroid, breast, and paranasal sinuses are just a few of the locations where extranodal involvement has been documented [8–10]. Although the inguinal, mediastinal, and axillary lymph nodes may also be involved, it is uncommon for RDD to localize to the retroperitoneal nodes [11].

Furthermore, it has been reported that RDD can manifest itself as unilateral inguinal adenopathy [12]. Nevertheless, lymphadenopathy can manifest in the inguinal, axillary, mediastinal, and other lymph nodes [13]. Lymph node involvement is usually bilateral and painless. Extranodal RDD is responsible for 25-40% of all cases [14,15]. RDD is classified as sporadic, familial, or cutaneous. The most prevalent type of RDD is sporadic RDD, which encompasses the typical nodal form, extranodal RDD, neoplasia-associated RDD, and immunological disease-associated RDD [4]. Classic RDD manifests as enormous bilateral painless cervical lymphadenopathy with fever, weight loss, and night sweats [16,17]. A study by Al-Maghrabi reported 17 patients diagnosed with RDD in Saudi Arabia in English literature. The study also reported distinctive features of RDD, including diverse histiocytic proliferation and pathognomonic emperipolesis, characterized by the presence of lymphocytes, plasma cells, and red blood cells enclosed within vacuoles in the cytoplasm of numerous histiocytes [18].

However, the underlying causes and mechanisms of RDD remain poorly understood. RDD can be hereditary or acquired through an infection. It has been proposed that this is the outcome of an immunological response to an infectious pathogen or an abnormal macrophage response to cytokines [19]. Although a direct connection has not been established, studies have linked RDD to viral infections, such as herpes viruses, Epstein-Barr virus, cytomegalovirus, and HIV [20]. RDD has also been associated with rheumatic conditions and malignancies [6].

Rosai-Dorfman disease is characterized by an increase in IgG4-positive plasma cells [15]. RDD is primarily diagnosed histologically based on the presence of proliferating foamy histiocytes with round vesicular nuclei and abundant pale cytoplasm. Lymphocytophagocytosis and emperipolesis are indicative of disease pathology [13]. Foucar, et al. documented a clinical trajectory characterized by stable disease in 54%, spontaneous regression in 21%, and progressive disease in only 1% of patients [13].

Although the best treatment is still being debated, surgical resection is generally curative. The prognosis is good with only a small chance of recurrence [18]. Steroids are the first-line therapeutic choice for individuals with RDD who require systemic treatment because they generate responses in both classical RDD and extranodal illness; however, the reliability and durability of these responses are unclear [14].

Conclusion

In conclusion, Rosai-Dorfman disease (RDD) is a rare and intriguing histiocytic disorder. Furthermore, RDD's clinical diversity extends to its rare manifestation as unilateral inguinal adenopathy, which can be encountered alongside lymphadenopathy in other regions. The underlying causes and mechanisms of RDD remain unclear, with suggested associations with viral infections, immunological responses, and various systemic conditions. Surgical resection is often curative for RDD, with a favorable prognosis and a low risk of recurrence. Steroids are the first-line therapeutic choice in cases requiring systemic treatment; however, the long-term effectiveness of such treatments remains uncertain. Further research and a comprehensive understanding of RDD's

etiology, pathogenesis, and treatment approaches of RDD are essential to enhance the management and outcomes of individuals affected by this rare and enigmatic disease.

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