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Unusual presentation of extranodal Rosai-Dorfman disease: A scalp lump

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ABSTRACT

Rosai-Dorfman disease (RDD) is a benign and idiopathic proliferative histiocytic disorder that typically manifests with extensive cervical lymphadenopathy, fever, polyclonal gammopathy, and leukocytosis with neutrophilia. However, the purely cutaneous variant of RDD is exceptionally uncommon, occurring in only 3% of all RDD cases.

In this abstract, we present a rare case of RDD exhibiting a purely cutaneous form. The patient, a 40-year-old male, presented with a scalp lump. Diagnostic investigations, including histopathological examination, confirmed the diagnosis of RDD. No lymphadenopathy or systemic involvement was detected, highlighting the uniqueness of the cutaneous presentation.

The rarity of pure cutaneous RDD underscores the significance of reporting such cases to enhance our understanding of the diverse clinical manifestations of this condition. Awareness of this atypical presentation is crucial for accurate diagnosis and appropriate management, as the treatment approach for purely cutaneous RDD may differ from the more common systemic forms.

Through this case report, we aim to contribute to the existing medical literature on RDD, emphasizing the need for careful evaluation of skin lesions to include RDD in the differential diagnosis, even in the absence of lymphadenopathy or systemic symptoms. Further research and documentation of similar cases will undoubtedly advance our knowledge of this intriguing disease and aid clinicians in providing optimal patient care.

Keywords: scalp lump, lipoma, Rosai Dorfman disease, case report

INTRODUCTION

Rosai-Dorfman disease is a benign, idiopathic proliferative histiocytic disorder. Typically, it presents with extensive cervical lymphadenopathy, accompanied by fever, polyclonal gammopathy, and leukocytosis with neutrophilia [1].

However, the purely cutaneous variant of the disease is exceptionally rare, occurring in only 3% of Rosai-Dorfman Disease cases [1]. Currently, the treatment approach for cutaneous forms remains poorly defined.

In this report, we present a case of Rosai-Dorfman disease observed in its exclusive cutaneous form. The patient exhibited distinct skin lesions without involvement of lymph nodes or systemic symptoms.

PATIENT AND OBSERVATION

Patient Information: A 40-year-old male presented with a single 5 cm mass on the scalp, which had appeared three years ago (Figure 1). There was no history of trauma or previous surgery, and no relevant medical, surgical, or family history. The patient had no comorbidities.

Clinical Findings: The lesion was soft and non-tender upon palpation, fixed in relation to the deep plane.

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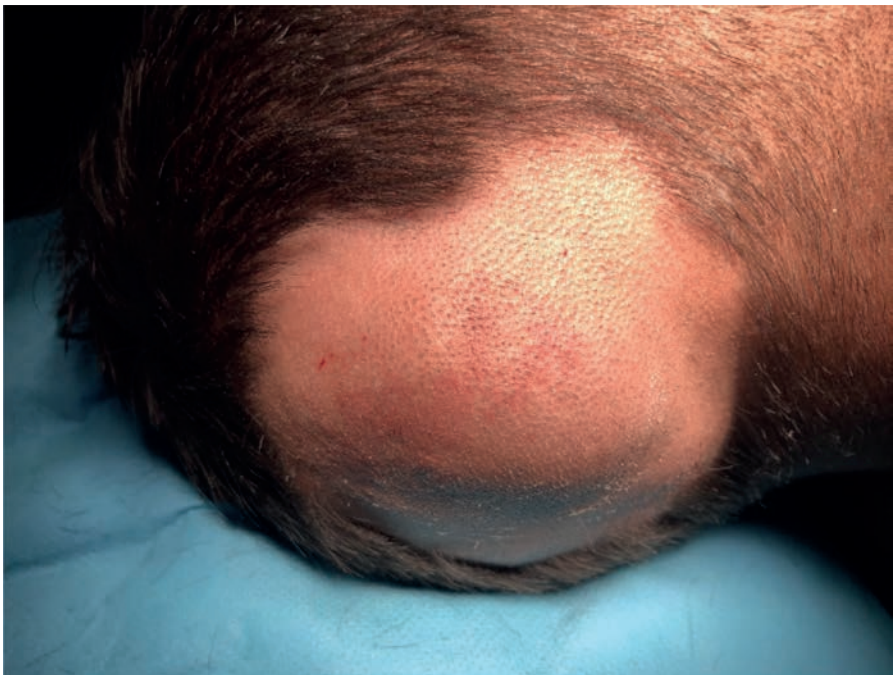

FIGURE 1. *Subcutaneous nodule clinically suggestive of a lipoma*

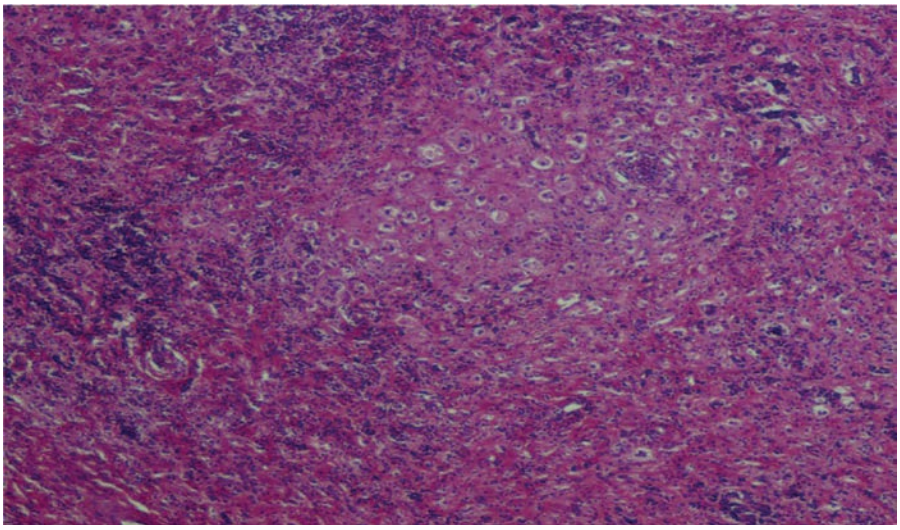

FIGURE2. *Tissue showing predominant histiocytes infiltration associeted with plasma cells, and lymphoid aggregates*

There were no associated systemic symptoms, such as fever, weight loss, or malaise, and no lymphadenopathy was observed during physical examination.

Timeline of the current episode: The mass had been evolving for three years.

Diagnostic assessment: All pertinent laboratory results, including serum protein electrophoresis, complete blood count with differential, and erythrocyte sedimentation rate, were within normal limits.

Diagnosis and therapeutic interventions: The patient underwent surgery, and a total excision of the mass was performed. Macroscopic appearance suggested a lipoma.

Histologic evaluation revealed a proliferation of tumor cells with a non-Langerhansian benign histiocytic appearance. Pathological examination of the mass showed tissue composed predominantly of histiocytes, plasma cells, and lymphoid aggregates (Figure 2). Histiocytes were observed phagocytizing lymphocytes, plasma cells, erythrocytes, and polymorphonuclear leukocytes without digesting them, a phenomenon termed emperipolesis (Figure 3). This histological pattern was consistent with Rosai-Dorfman disease.

Follow-up and outcome of interventions: The lesion was completely excised, and one year of follow-up showed no recurrence or development of new nodules. There were no indications of systemic involvement.

Informed Consent: informed consent was obtained from the patient to use this case.

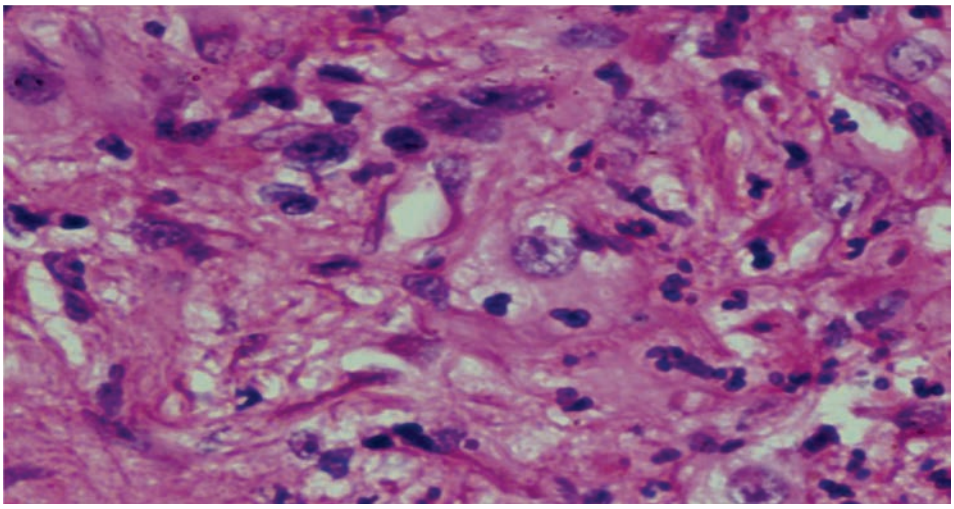

FIGURE 3. *Histiocytic cells with emperipolesis*

DISCUSSION

Rosai-Dorfman disease is a rare acquired condition that mainly affects young adults. Its clinical manifestation typically involves painless bilateral cervical lymphadenopathy, and less commonly, inguinal or axillary lymph nodes, accompanied by fever [2].

The purely cutaneous form of the disease is uncommon, and only one study in the literature has identified 22 cases with a purely cutaneous presentation [2]. Clinically, the skin lesions can resemble lipomas, making the diagnosis challenging. In such cases, histological examination combined with immunohistochemistry (IHC) is essential to confirm the diagnosis, as it reveals the characteristic feature of emperipolesis, with histiocytes expressing the proteins S100 and CD 68 [3] [4]. Although our case did not show associated biological abnormalities such as polynuclear neutrophilic leukocytosis, hypergammaglobulinemia polyclonal, increased sedimentation rate, or anemia [2] [5], these abnormalities may be present in other cases.

While the course of the disease is generally mild, it can exceptionally become life-threatening if lymph nodes compress nearby organs, particularly in the ENT region, or if there is systemic organ involvement, such as the liver or kidney, or associated hemolytic anemia [5]. However, in cases of pure cutaneous Rosai-Dorfman disease, spontaneous resolution within months or years is common, with intermittent relapses followed by remission.

The treatment for this disease remains poorly standardized due to its rarity, and there are no randomized comparative studies. Systemic treatment (chemotherapy, corticosteroid therapy, surgery, radiotherapy) may be considered in cases of lymph node or systemic involvement, with varying results depending on the studies [5].

CONCLUSION

In conclusion, Rosai-Dorfman disease is a rare acquired condition that predominantly affects young adults. The typical presentation involves painless bilateral cervical lymphadenopathy, and in rarer cases, inguinal or axillary lymph nodes, accompanied by fever. The purely cutaneous form of the disease is even more uncommon, and its clinical resemblance to lipomas can pose diagnostic challenges.

The prognosis of the disease is generally favorable, with spontaneous regression of cutaneous lesions over time. However, severe complications may arise if lymph nodes or systemic organs are affected.

Due to the rarity of this disease, treatment options remain poorly standardized, and there are no randomized comparative studies. Management depends on the presence of lymph node or systemic involvement, and may involve approaches such as chemotherapy, corticosteroid therapy, surgery, or radiotherapy.

Continuing to document and report cases of this disease is crucial to enhance our understanding and management of this rare condition. Further studies are needed to elucidate optimal therapeutic approaches and their effectiveness in different forms of Rosai-Dorfman disease.

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MDT: conceptualization, methodology and software; MDT and WF: data curation, writing-original draft preparation; MDT and WF: visualization, supervision, writing, reviewing and editing of manuscript; AJ and HB: validation.
All the authors have read and agreed to the final manuscript.

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