Case Report

iMedPub Journals
www.imedpub.com

Insights in Biomedicine
ISSN 2572-5610

2021
Vol.6 No.5:21

Cutaneous Rosai-Dorfman Disease Presented with a Solid Mass in the Thigh: A Case Report

Vouros Dimitrios* and Toutouzas Konstantinos

1st Propaeudeutic Department of Surgery, Hippokrateion General Hospital, Athens, Greece

*Corresponding author:
Dr. Dimitrios Vouros

jimsamiotis@hotmail.com or jimsamiotis91@gmail.com

1st Propaeudeutic Department of Surgery, Hippokrateion General Hospital, Athens, Greece

Tel: 306976721606

Abstract

Rosai-Dorfman disease (RDD) is a rare benign disorder of histiocytic origin It was first described by Rosai and Dorfman as sinus histiocytosis with massive lymphadenopathy in 1969. The disease mainly affects cervical lymph nodes and may express extranodal involvement. A purely cutaneous form of RDD has also been described. The etiology of the disease is unknown. Various mechanisms such as genetic, infectious, and inflammatory have been proposed. Various forms of treatment are suggested ranging from local steroids to surgical excision. We report the case of a young female who presented with a cutaneous lesion of the right thigh and was subsequently diagnosed with RDD involving the skin and the subcutaneous tissue.

Keywords: Rosai- Dorfman disease; Sinus histiocytosis; Emperipolesis; Surgery; Hematology

Received: May 05, 2021; **Accepted:** May 17, 2021; **Published:** May 24, 2021

Citation: Dimitrios V, Konstantinos T (2021) Cutaneous Rosai-Dorfman Disease Presented with a Solid Mass in the Thigh: A Case Report. Insights Biomed Vol.6 No.5:21

Introduction

Rosai-Dorfman disease (RDD) is a rare benign disorder of histiocytic proliferation that usually presents with bilateral cervical lymphadenopathy in children. We describe the case of a A 28-year-old female patient presented to our institution with a progressively growing lesion located in the external surface of the right thigh of 8 months duration.

Case Presentation

A 28-year-old female patient presented to our institution with a progressively growing lesion located in the external surface of the right thigh of 8 months duration. She had no fever, night sweats and loss of weight in the past. The patient had a medical history of Hashimoto disease with a recent neck ultrasonography with the presence of two thyroid nodules and absence of cervical lymphadenopathy and her medication was thyroxine with normal thyroid hormone levels. She had no history of previous operations. The patient was initially admitted to a vascular surgeon who performed a venous Doppler ultrasound of the lower limp with no signs of superficial or deep vein thrombosis and recognition of a solid mass with increased vascularity and local hypodense regions with absence of blood flow. A magnetic resonance imaging (MRI) done one month prior to presentation showed a poorly defined mass in the subcutaneous tissue of the external surface of the right thigh with a cephalocaudal diameter of 9.2 cm and transversal diameter of 4.5 cm extending from the internal edge of the skin until the fascia of the iliotibial tract with no signs of invasion. The mass had increased signal intensity on T2-weighted MRI with elevated contrast agent intake and restricted diffusion.

On physical examination, a palpable immobile painless mass was recognized. The skin over the mass had a bruishe-like distortion with absence of papules, plaques or nodules **(Figures 1, 2a and 2b)**. Laboratory tests were normal with no signs of leukocytosis, neutrophilia and normal serum tumor markers. Vitamin D levels were slightly decreased and parathormone (PTH) was elevated. An excisional biopsy was initially performed under local anesthesia. The histopathology report was non diagnostic. It reported invasion by lymphocytes and hystiocytes and was negative for melanoma and sarcoma. As this nonspecific inflammation was non diagnostic, a total excision of the mass was decided. A wedge resection of the lesion was performed. The skin and the fascia of the iliotibial tract were excised as well **(Figures 3a and 3b)**. A Redovac drainage system was placed and the skin was closed with interrupted nylon sutures. The patient had an uncomplicated postoperative course and was discharged the next day after removal of the drainage.

The histopathology report revealed non-Langerhans

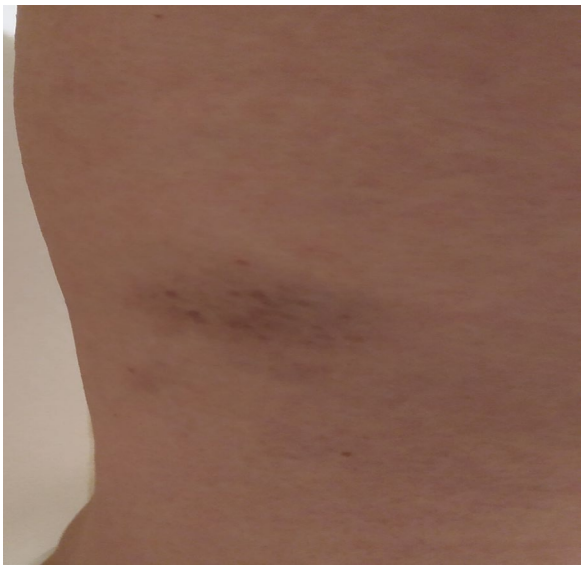

Figure 1 Clinical image showing the bruise-like distortion on the external site of the right thigh. The lesion was painless on physical examination.

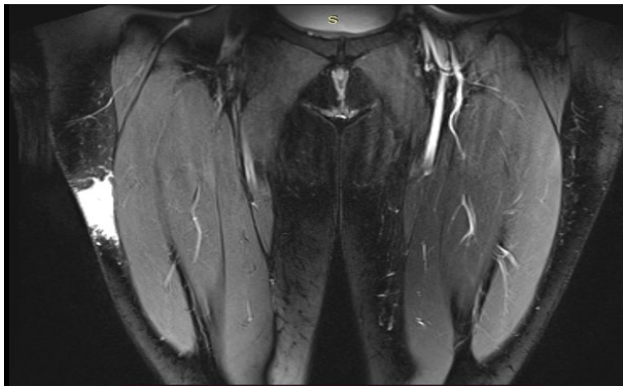

Figure 2a T2 weighted MRI coronal plane showing the lesion on the right thigh. The fascia of the muscle is shown with no signs of invasion.

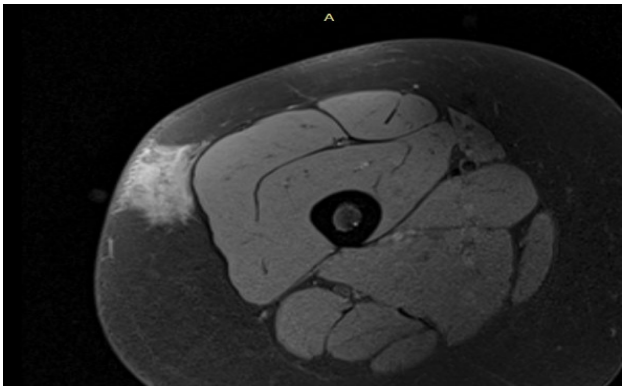

Figure 2b T1 MRI transverse plane of the lesion. A regional thickening of the skin above the lesion was noticed.

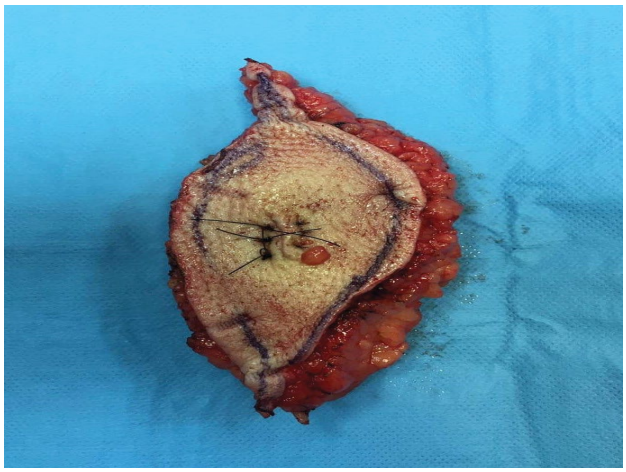

Figure 3a Macroscopic specimen including the skin. The interrupted sutures are from the site where excisional biopsy was initially performed.

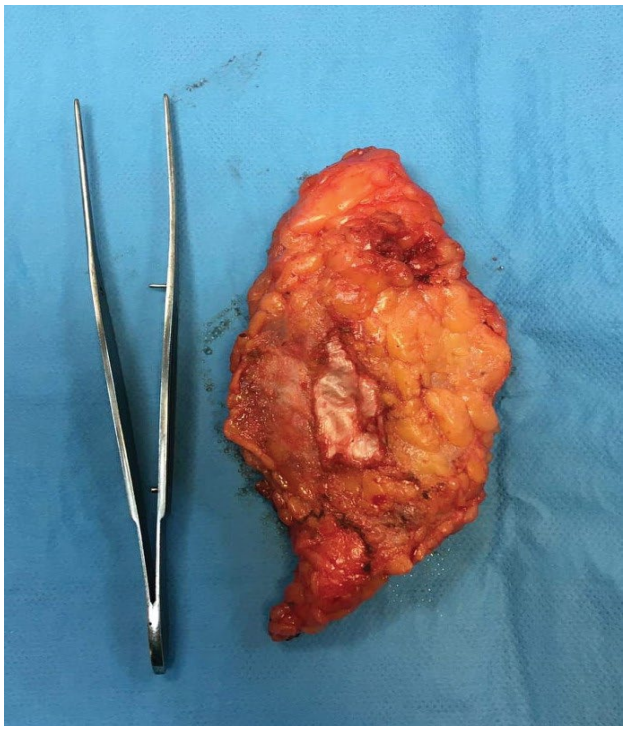

Figure 3b Macroscopic specimen of the skin, subcutaneous mass. The fascia of the underlying muscle was also excised based on the imaging findings.

histiocytosis of the skin and the subcutaneous tissue and specifically extranodular Rosai-Dorfman histiocytosis. The transverse dimensions of the lesion were 4 × 2 cm and the depth of the lesion in the subcutaneous fat was 3 cm. Mixed lymphoplasmacytic infiltrates with histiocytes was reported. On immunohistochemistry, it was found to be S-100 and CD163 positive. The cells were negative for CD1a, Factor XIII and mutated protein BRAF V600E. Nonstaining lymphocytes were recognized

in proximity of the macrophages suggesting intracytoplasmic phagocytosis, otherwise known as emperipolesis. Levels of IgG4 immunoglobulin and IgG/IgG4 ratio were normal. EBV detection using the *in situ* EBERs method was negative.

All surgical margins were negative and a R_0 resection was achieved. The patient was admitted to the Department of Hematology in our institution and further laboratory tests were performed. Specifically, virological tests revealed elevated EBVA, VZV and CMV IgG levels and the patient was negative for HbsAg and anti-HCV levels. Total IgG levels were slightly elevated. In an attempt to exclude nodular involvement and systematic disease, a PET-CT Scan was performed which was negative for lymph node involvement and other cutaneous lesions as well. Two months after the excision the patient remains disease free with no signs of local recurrence [1-3].

Discussion

Cutaneous Rosai Dorfman disease is a rare and unique form of histiocytopathy including a proliferation of histiocytes with macrophage properties in a polymorphous inflammatory cell background rich in plasma cells, lymphocytes and neutrophils [4]. The etiology of this disease remains unknown. Diagnosis is rendered based on same histological and immunophenotypical findings. Ahmed et al. published on 2019 a systematic review on cutaneous Rosai- Dorfman disease in an attempt to better define the clinical presentation, natural clinical course and prognostic significance of CRD based on published literature [3]. After systematic literature search, 263 patients in total of which 220 patients (84%) had purely cutaneous Rosai Dorfman disease in papers published up to May 2018. The mean age of patients was 47.1 years at the time of presentation. Females were 56% (121) of the patients. Caucasians comprised only 20% of the patients (33) as Eastern Asians were the majority of the cases reported (57%). Extremities, as in our patient, were the most frequent site of the disease (51.5%) with a single number of lesions reported in 86 patients (40%). Furthermore, dermis and subcutaneous tissue involvement was reported on 85 patients (32%).

The histopathology report is necessary to establish the diagnosis of RDD and rule out pathologies such as sarcoma, melanoma [5]. The initial report of a nonspecific inflammation may delay the proper recognition and management of patients with RDD. Increased inflammation may cause ulceration, necessitating a histologic diagnosis to rule out a malignant process [6]. In our case, ulceration was not present. Cutaneous Rosai Dorfman disease has also been associated with different types of lymphomas such as marginal zone lymphoma at the same site that can either be synchronous or metachronous [5]. In those cases further therapies such as chemotherapy may be necessary. Another important factor for the proper categorization of the disease is whether Rosai Dorfman disease is related to the cutaneous IgG4 sclerosing disease [7]. The number of IgG4 positive plasma cells in our case was not increased. Emperipolesis, the classic phenotypic profile and the extent of histiocytic infiltration ruled out this pathology and confirmed Rosai Dorfman disease.

Surgical excision was performed in an attempt to establish a proper diagnosis and to ensure an acceptable therapeutic option. For that purpose, oncological principles were implemented and a wedge resection was decided. The mass was intraoperatively tried not to be seen and the underlying muscle fascia was also excised based on the imaging preoperative findings. R_0 excision was finally achieved. It seems that this treatment modality has the best outcomes in patients with cutaneous RDD as complete remission is achieved in most of the patients after surgical excision which is as high as 56% compared to other options such as chemotherapy, radiation, intra-lesional and oral steroid treatments [3].

Conclusion

Primary cutaneous RDD is a rare entity whose diagnosis remains challenging for clinicians and pathologists alike given its non-specific clinical and histologic presentation. Systemic work up is necessary in order to exclude systemic involvement. The role of immunohistochemistry is crucial in an attempt to establish a diagnosis and rule out other pathologies. Surgical excision seems to be vital with the best outcomes compared with other treatment options.

References

1 Rosai J, Dorfman RF (1969) Sinus histiocytosis with massive lymphadenopathy: A newly recognized benign clinicopathologic entity. Arch Pathol 87: 63-70.

2 Foucar E, Rosai J, Dorfman R (1990) Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Review of the entity. Diagn Pathol 7: 19-73.

3 Ahmed A, Crowson N, Magro C (2019) A comprehensive assessment of cutaneous Rosai-Dorfman disease Ann Diagn Pathol 40: 166-173.

4 Fayne R, Rengifo SS, Gonzalez I, Solorzano JL, Gonzalez D, et al. (2020) Primary cutaneous Rosai-Dorfman disease; a case-based review of a diagnostically and therapeutically challenging rare variant. Ann Diagn Pathol 2: 2-4.

5 Machan S, Medina C, Rodríguez-Pinilla SM, Suárez-Peñaranda JM, Castro Y, et al. (2015) Primary cutaneous marginal IgG4 lymphoma and Rosai-Dorfman's disease coexisting in several lesions of the same patient. Am J Dermatopathol 37: 413-418.

6 Lu CI, Kuo TT, Wong WR, Hong HS (2004) Clinical and histopathologic spectrum of cutaneous Rosai Dorfman disease in Taiwan. J Am Acad Dermatol 2: 2-4.

7 Kuo TT, Chen TC, Lee LY, Lu PH (2009) IgG4-positive plasma cells in cutaneous Rosai-Dorfman disease: An additional immuno-histochemical feature and possible relationship to IgG4-related sclerosing disease. J Cutan Pathol 36: 1069-1073.