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REVIEW ARTICLE

SOFT TISSUE ROSAI: DORFMAN DISEASE OF THE SACRAL REGION, A VERY RARE DISEASE: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT: INTRODUCTION: Rosai–Dorfman disease (RDD) is a rare proliferative histiocytic disorder of unknown etiology. RDD typically presents with generalized lymphadenopathy and polymorphic histiocytic infiltration of the lymph node sinuses; however, occurrences of extranodal soft tissue RDD may rarely occur. We are presenting a case that presented as a subcutaneous swelling. **CONCLUSION**: RDD is a rare inflammatory lesion that should be considered in the differential diagnosis of a soft tissue tumor. Differentiation of extra nodal RDD from more common soft tissue tumors such as soft tissue sarcoma or inflammatory my fibroblastic tumor is difficult and usually requires definitive surgical excision with histopathological examination. The optimal treatment for extra nodal RDD is surgical excision and is usually curative.

KEYWORDS: Rosai Dorfman, soft tissue swelling.

INTRODUCTION: Sinus histiocytosis with massive lymph adenopathy (SHML) is a class II histiocytosis first described as a unique clinicopathologic entity by Rosai and Dorfman in1969.[1] Although lymph nodes are more commonly involved, any organ may be affected – thus the term RDD has been adopted in place of SHML.[1]

Although cutaneous involvement in RDD is common, purely cutaneous disease is rare and not well documented.[2] RDD typically presents with generalized lymphadenopathy and polymorphic histiocytic infiltration of the lymph node sinuses; however, occurrences of extranodal soft tissue RDD may rarely occur when masquerading as a soft tissue sarcoma.[1] Extra nodal RDD lesions may occur with or without lymphadenopathy and may be solitary or multiple,[3] involvement occurs in up to 43% of cases.[4]

CASE REPORT: A 34 year old lady presented to surgical OPD with swelling just above the sacral bone lateral (left) to midline for 3 months. The swelling was accidently noted and there was no change of size of the swelling during this time. It was not associated with any other swellings. On examination the size swelling was 1*1 cm with smooth surface but irregular shape, hard in consistency, well defined edges and freely mobile.

A thorough examination didn't reveal any other swelling or lymphadenopathy. FNAC showed histocytic proliferative lesion with significant emperipolesis. Routine blood investigations were within normal limits. A chest X-ray and USG abdomen was taken which was also normal. An excision biopsy was done. Intra-operative finding were the same and the color of the lesion was pale white similar to that of a lymph node. Biopsy was suggestive of Rosai Dorfman disease which was confirmed by immunohistochemistry. Patient has no complaints in a two month follow up.

DISCUSSION: Rosai–Dorfman disease was first described as a distinct clinic-pathologic entity by Rosai and Dorfman in 1969.[1] RDD is of unknown etiology, although viral agents such as human

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herpes virus-6 and Epstein–Barr virus are proposed to have a causative role.[2] Extra nodal involvement in RDD has been reported in a wide range of organs, skin and nasal sinuses are the most commonly affected organs,[2] but isolated soft tissue RDD is rare.[4]

In the largest series of RDD cases, 43% (500 patients) had extra nodal disease, while only 2.6% patients had the typical morphologic picture of soft tissue RDD without detectable lymphadenopathy[5]. While RDD is seen predominantly in males,[2] soft tissue Rosai-Dorfman disease is a rapidly evolving, mostly solitary and non-recurrent lesion in middle-aged females.[7] Soft tissue RDD presentation is with non-specific features, leading to misdiagnosis as benign inflammatory and fibro histiocytic lesions as well as lymphoma and malignant fibrous histiocytoma.[5,9]

Recurrence is common among isolated soft tissue RDD without lymph node involvement[5]. Cases were reported were it presented as breast swelling with or without axillary lymph nodes causing diagnostic confusion.[8] However, the pathogenesis of RDD is still poorly understood but is likely multifactorial, with infection and immune mediated disorders proposed as leading etiological factors.[1,3]

The diagnosis of RDD is generally made on histological grounds, while immunohistochemistry may be needed for diagnostic confirmation.[5] Treatment includes clinical observation without treatment when possible as spontaneous resolution of adenopathy is observed.[6] In symptomatic RDD surgical excision is preferred.[2] For isolated soft tissue RDD, a wide local excision is often recommended as recurrence is possible.[5] In the presence of vital organ compression and/or extra nodal localization with important clinical signs, surgical de bulking may be necessary. Radiotherapy has shown limited efficacy, while chemotherapy is in general ineffective.[6]

CONCLUSION: RDD is a rare inflammatory lesion that should be considered in the differential diagnosis of a soft tissue tumor. Differentiation of extra nodal RDD from more common soft tissue tumors such as soft tissue sarcoma or inflammatory my fibroblastic tumor is difficult and usually requires definitive surgical excision with histopathological examination. While the optimal treatment for extra nodal RDD is surgical excision which is usually curative, sometimes a wide excision is needed.

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