Case Report

Extra Nodal (Cutaneous) Rosai Dorfman Disease

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Abstract: Rosai-Dorfman disease is an extremely rare disorder characterized by proliferation and accumulation of histiocytes in lymph nodes, usually in the cervical region. In approximately 43% cases, this accumulation occurs in extra nodal sites such as the skin, nasal sinuses, central nervous system, kidney and gastrointestinal tract. We report such a case of extra nodal (cutaneous) Rosai Dorfman disease in a young female patient.

Key Words: Rosai-Dorfman disease, histiocytes, lymph nodes, extra nodal, skin

Introduction:
Rosai Dorfman disease, also known as Sinus histiocytosis with massive lymphadenopathy (SHML) was first described by Destombes in 1965 and then subsequently categorized as a distinct clinicopathological disorder in 1969 by Juan Rosai and Ronald Dorfman (1). It is a benign, self-limiting disease of unknown etiology occurring mostly in young patients, characterized by proliferation and accumulation histiocytes in lymph nodes or extra nodal sites such as skin, nasal sinuses, central nervous system, kidney and gastrointestinal tract. The usual presentation is a lymph nodal swelling in the cervical region associated with constitutional symptoms such as fever and elevated ESR with leukocytosis. Altered immune responses and infectious agents are thought to play a role in its occurrence. The hallmark of the disease is emperiploesis, which is the phagocytosis of lymphocytes or erythrocytes by histiocytes and this is required for the diagnosis (1).

Case Presentation
This is the case of a 28-year-old South Indian female who presented to Government Wenlock Hospital, Mangalore with a painless lump on the left anterior aspect of the arm for 2 years, which was progressively increasing in size. She had no preceding history of trauma, fever or any similar swellings elsewhere. She denied any constitutional symptoms or neurological deficits. She had no associated history of weight loss. On examination, it was a firm, well-defined swelling on the ventral aspect of the left arm, 6x4cm in size which was mobile in all planes, present 4cm below the acromion process and 10cm above cubital fossa of the left arm, with no associated skin changes, discharge or erythema over the swelling. The skin could not be pinched over the swelling and the swelling was not attached to any deeper structures, which was suggestive of a cutaneous swelling. There was no associated lymphadenopathy. Other systems examined were normal clinically and on imaging. Preoperatively, a full blood count was done which revealed a normocytic normochromic anemia and elevated ESR with leukocytosis. Fine Needle Aspiration of the left upper arm in the superficial plane. Features were likely A) Inflammatory lesion likely to be fungal/parasitic. B) Inflammatory cell rich benign/intermediate grade with soft tissue neoplasm – Benign neural neoplasm/Inflammatory myofibroblastic tumor.

The patient underwent wide local excision including excision of the involved skin which was done under general anesthesia and intra-op findings revealed a firm swelling in the...
subcutaneous plane with adhesions to the overlying fat but not involving the underlying muscle.

On cut section, it was a well circumscribed solid grey white homogenous lesion with specks of hemorrhage and few yellow areas abutting all resected margins. Histopathology revealed dense cellular infiltrate involving dermis and subcutaneous tissue made up of aggregates of lymphocytes and sheets of large, pale histiocytes with large round vesicular nuclei arranged in a nodular, lobular pattern, with emperipolesis in some. At places the lymphocytic nodules contained numerous plasma cells along with few neutrophilic micro abscesses and stromal fibrosis seen in some places. These features are suggestive of cutaneous (extra nodal) Rosai Dorfmann disease. Immunohistochemistry studies revealed the specimen to be positive for CD68 and S100.

Figure 1: Histopathology examination of left anterior arm mass demonstrating characteristic emperipolesis and immunohistochemistry CD68 and S100 positive

Postoperatively there was no discernible nerve damage with resulting loss of power or cutaneous sensation.

In 2 months of follow up, there was no evidence of recurrence at the site of lesion and no swellings elsewhere on the body.

Discussion
Rosai Dorfman disease is a class II histiocytosis, a rare inflammatory non-neoplastic disease with a worldwide distribution and 80% cases occurring in children and young adults in the 2nd and 3rd decade of life with a slight male predominance of 58% (2). It is not usually life threatening and most often does not require therapy apart from surgical excision which is considered curative; but it may undergo exacerbations and remissions. It is of unknown etiology, but studies reveal that Human Herpes Virus 6, Parvovirus B19 and Ebstein Barr Virus to have a role in the pathogenesis (1). Clinical features of CRDD are variable and can include asymptomatic yellow-red to brownish nodular papules or plaques with facial skin being most commonly affected (5). There is no real use for further workup such CT imaging for spread in CRDD (1).

Due to the self-limiting nature and the rarity of the disease, a definite treatment protocol has not been established thus far. However, for patients with CRDD such as ours, excision of the mass has proved to be curative in most cases with negligible rates of recurrence. The proposed treatment algorithm for other forms of the disease is shown in Fig 2.

Figure 2: Treatment algorithm for Rosai–Dorfman disease. (1)

Conclusions
Rosai Dorfman disease is a rare non-malignant histiocytic disorder which can be of nodal or extra nodal type. In our patient it was of the extra nodal cutaneous type, which was managed with surgical excision. The patient is on monthly follow up subsequently to watch out for recurrence at the site of excision or elsewhere.

References

