A rare case of pure cutaneous Rosai-Dorfman disease

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Abstract

Rosai-Dorfman disease (RDD) is a rare but usually benign non-Langerhans cell histiocytic disorder which commonly affects lymph nodes and frequently present with bilateral painless enlarged cervical lymph nodes with systemic symptoms. Although extra nodal tissue involvement is seen in considerable proportion, purely cutaneous involvement without affecting lymph nodes is very rare and forms a diverse clinical entity. Histopathology with immunohistochemical staining remains gold standard in diagnosing this rare entity. Different treatment modalities have been tried in case reports and treatment response is variable. Here we report a case of pure cutaneous RDD in a 21-year-old young female who was referred for staged excision.

Introduction

Rosai-Dorfman disease (RDD) which is also known as sinus histiocytosis with massive lymphadenopathy is an uncommon benign non-Langerhans cell histiocytic proliferative disorder of unknown aetiology[1]. It commonly affects lymph nodes with or without extra nodal involvement and massive painless cervical lymphadenopathy is the usual presentation. More than 40% have extra nodal involvement, with the skin being the most frequently affected site[2]. However, purely cutaneous RDD without lymphadenopathy or internal organ involvement is rare and noticeable in 3% of reported cases[3].

Case report

A 21-year-old female presented to Colombo South Teaching Hospital, Kalubowila in 2018 September with gradually enlarging skin nodules over her right leg above the popliteal fossa (Figure 1), anterior chest in between her breasts (Figure 2) and left lower back (Figure 3) for three months duration. Initially the lesions appeared as small painless non itchy papules and progressively increased in size with occasional pain and surface ulceration. She denied any significant discharge. She did not have fever, malaise, loss of weight, loss of appetite, chronic cough, or night sweats. She declined a history of trauma or contact history of tuberculosis.

Examination revealed erythematous dome shaped nodule over right lower leg and erythematous indurated plaques with surface ulceration over the left lower back and anterior chest. Physical examination was normal without any evidence of pallor, lymphadenopathy and organomegaly.

Her full blood count, inflammatory markers, renal, liver profile and chest X-ray were normal. Skin biopsy revealed a dense infiltration of large histiocytes admixed with inflammatory cells with variable emperipolesis in the dermis (Figure 4). Immunohistochemical staining showed that the histiocytes were strongly positive for S100 and negative for CD1a.

Diagnosis of exclusive cutaneous RDD was made due to immunohistopathology findings and patient was referred to plastic surgical unit for staged excision. Unfortunately, she has defaulted follow up.

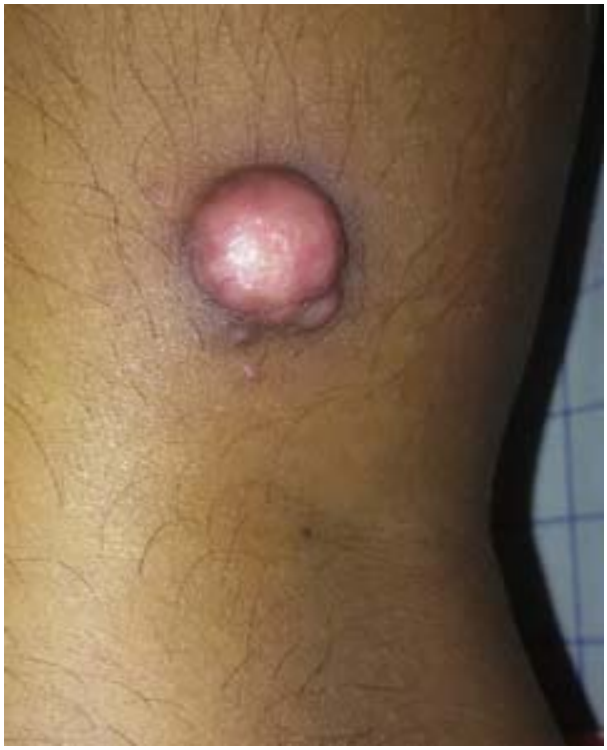

Figure 1.

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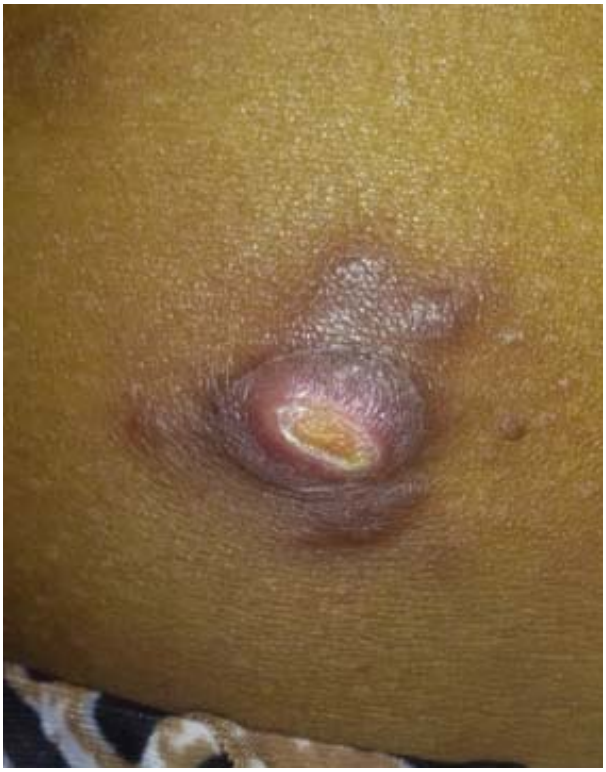

Figure 2.

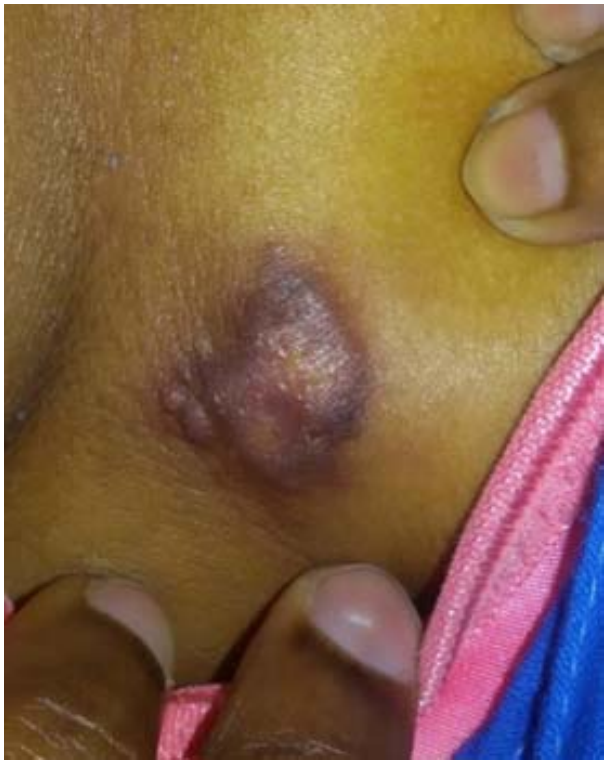

Figure 3.

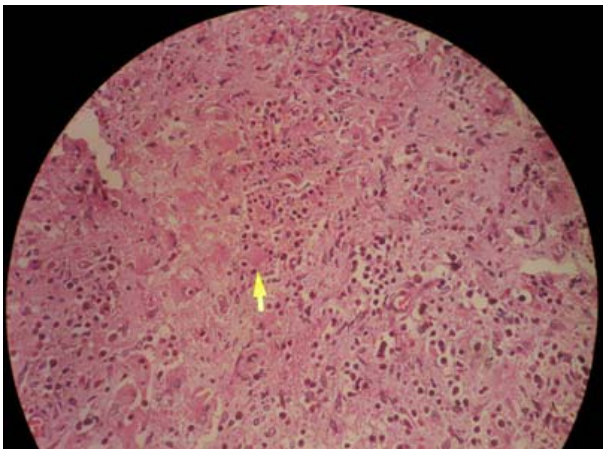

Figure 3.

Discussion

Although rare, pure cutaneous RDD can be presented with asymptomatic papules, nodules and plaques without lymph node or organ involvement. Histopathology and immunohistochemical studies are the corner stone in diagnosis. Dermal infiltrate of histiocytes with engulfed lymphocyte and inflammatory cells – emperipolesis is a characteristic feature. Immunostaining of histiocytes exhibit positive S100 protein, CD11c, CD14, CD33, and CD68 antigens and are CD1a negative[4].

Some cases are self-limiting, and the treatment options are variable and nonspecific which includes corticosteroids, dapsone, methotrexate, thalidomide, isotretinoin, cryosurgery, radiotherapy, and surgical excision[1].

Although rare, pure cutaneous RDD is increasingly being recognized and optimal management has yet to be elucidated.

References

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