Case Report

Rosai-Dorfman disease presented with multiple papules and nodules along with lymphadenopathy – a rare case report

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ABSTRACT:

Rosai- Dorfman disease is a benign proliferative disorder of histiocytes frequently observed after viral infection.

The disease usually presents with massive cervical lymphadenopathy. It usually appears in patients with 1st and 2nd decade of life as a febrile illness with lymphadenopathy, polyclonal hyper-globulinemia, leukocytosis, anaemia and elevated erythrocyte sedimentation rate. Other than skin, breast, kidney, thyroid, testis, and CNS are affected.

Male and black persons are specially susceptible. Skin lesions consists of isolated or disseminated yellow- brown papules or nodules or macular erythema with spontaneous remission.

We report such an uncommon entity in a 29-year-old lady presented with multiple papules and nodules over the right thigh for last 2.5 years. Histopathology shows sheets of large histiocytic cells having abundant eosinophilic cytoplas in the dermis.

Key Words: Rosai-Dorfman disease, Lymphadenopathy, histiocytes,

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Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare benign non-Langerhans cell histiocytosis. Rosai and Dorfman first described it as a clinicopathologic entity in 19691 . The etiology behind RDD remains unknown, but due to its sporadic nature, viral and autoimmune causes have been hypothesized. RDD may be limited to the lymph nodes or have extranodal involvement, which occurs in more than 40% of patients2. The most common site of extranodal involvement is the skin1. Cutaneous involvement without lymphadenopathy is extremely rare. To our knowledge, documentation in the literature is currently limited, with only few described cases of purely cutaneous RDD available. We present an additional unique case of cutaneous RDD and emphasize the challenging nature of accurate histopathologic diagnosis.

Case Presentation

A female of 29 years old married, muslim, normotensive ,non diabetic, housewife hailing from Cumilla with the complaints of multiple elevated lesions over the front of

the right thigh for last 2.5 years. Occasional pain & burning sensation over the affected area for same duration. On query she gave history of a slowly developing an elevated lesion over the front of right thigh associated with itching & burning sensation. A few months later, the area became hardened & the local temperature was increased.The papules were firm in consistency & not fixed with underlying structure. She took different medications consulting with local doctors but her condition didn't improve. Then she visited a dermatologist & clinically diagnosed as a case of leiomyoma.

On inspection of Integumentary system reveals multiple discrete or confluent, unilateral, some erythematous & some are flesh colored papules & nodules of different sizes & shapes over the extensor surface of the right thigh. Some nodules show telangiectasia & scales on hyperpigmented background. On palpation, these papules & nodules are firm in consistency , mildly tender, & the local temperature is raised, are not fixed with underlying structures. Regional lymph nodes are not enlarged . Mucous membrane , nail & hair reveals no abnormality Other systemic enquiry reveals normal findings She has no history of fever, weakness, cough, weight loss, lymphadenopathy, gastrointestinal symptoms, or similar lesions elsewhere. She has no history contact with TB patient. Her bowel & bladder habit are normal. General examination revealed normal findings. Her investigation reports including CBC, renal and liver function test, random blood sugar, lipid profile, thyroid function test, ECG, Echo cardiogram, ultrasonogram of whole abdomen, chest X-ray, montoux test revealed normal findings.

Findings were concerning for several differentials including but not limited to (Rosai Dorfman Disease) RDD, Langerhans cell histiocytosis, sarcoidosis, cutaneous lymphoma, Kaposi sarcoma, eruptive xanthoma, dermatofibrosarcoma protuberans, keloid, lupus vulgaris, keloidal blastomycosis, histoid leprosy and lymphangioma circumsciptum. Skin biopsy for Histopathology shows in epidermis there is mild hyperkeratosis. The dermis shows sheets of large histiocytic cells having abundant eosinophilic cytoplasm. Dermis shoes dense infiltration of chronic inflammatory cells including many plasma cells, Russells bodies and nuclear debri. No organism is found in Z-N & fite stained sections. Immunohistochemistry report shows CD 68: Positive in tumor cells, S 100 protein: Positive in tumor cells CD 1 alpha : Negative in tumor cells which is compatible with Rosai Dorfman Disease

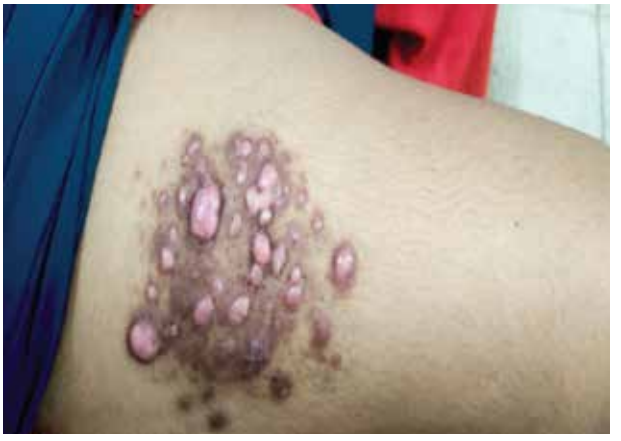

Figure 1: multiple ,unilateral, mildly pruritic, some erythematous & some are skin colored papules & nodules of different sizes & shapes overlying adherent scale over the extensor surface of the right thigh with hyperpigmented background. Papules & nodules are firm in consistency , mildly tender,

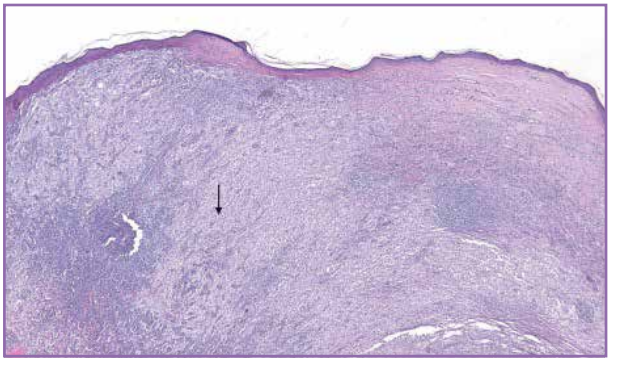

Figure 2 : A nodular proliferation of non-lymphod histocytic infiltrate consisting of eosinophilic appearing histocytes containing multiple nucleoli on H&E stain with magnification × 20

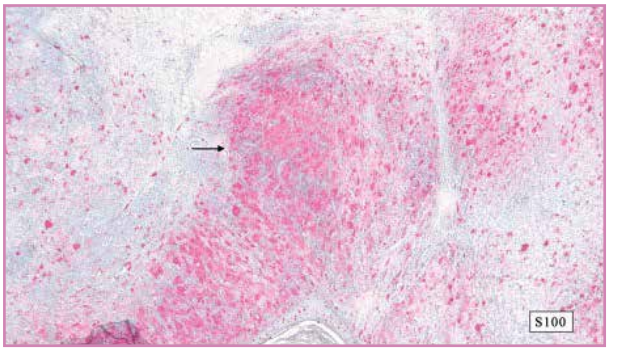

Figure 3: the histocytic infiltrate demonstrated positivity for S-100 with magnification × 20

Discussion

Compared to patients with systemic RDD, cutaneous RDD presents at an older mean age of about 43.5 years, is more common in White and Asian females, and has no lab abnormalities. Cutaneous manifestations are non-specific with asymptomatic red-brown to yellow papules, nodules, or plaques of varying size that can be confined or disseminated.. The most common sites of involvement are thought to be the extremities 3. The diagnosis of primary cutaneous RDD is often difficult and significantly delayed due to the non-specific nature of its clinical and histologic features 4 We present a unique case of cutaneous RDD as our patient was over 20 years older than the mean age of diagnosis, asian female;. A large series of cutaneous RDD was described by Kong et al. in their clinical and histopathologic study of 25 patients in China 5. Of the 39 skin lesions that were observed, the extremities were noted to be the most commonly involved, followed by the trunk and the face. The most common clinical presentation was the papulonodular type (79.5%), followed by the indurated plaque type (12.8%) and tumor type (7.7%). Unique histologic cases in this cohort were the identification of fibrosis in four patients, xanthomatous change in two cases, and coexistence of localized Langerhans cell histiocytosis in one patient. Follow-up was observed in 22 patients over 2-55 months where they found that surgical excision was the most exclusive effective treatment. Additionally, Ahmed et al. performed a review of the 220 patients with purely cutaneous RDD published to date and found the majority of cases presented with multiple lesions (60%) that remain present for an average timespan of 19 months 3. Histopathology of cutaneous RDD typically shows a dermal infiltrate of large polygonal histiocytes with abundant pale to eosinophilic cytoplasm admixed with an inflammatory infiltrate 6. Emperipolesis, which is the presence of intact inflammatory cells like lymphocytes, plasma cells, and neutrophils within the cytoplasm of histiocytes, is characteristic of the disease. The histiocytes stain positively for S100 and CD68 but are negative for CD1a 4. Diagnostic histologic challenges may include increased foamy cells effacing the epidermis, vascularity, stromal reaction, inflammation leading to ulceration which may be mistaken for xanthomatous lesions, hemangiomas/vasculitis, fibrous soft-tissue tumors, or malignant process respectively.

The recommended management of RDD is close observation, as it is usually an indolent, benign self-limited condition. If treatment is desired, surgical excision is considered most effective for both solitary disease and multifocal disease. Improvement has been observed with topical, intralesional, or systemic corticosteroids, cryotherapy, radiation, retinoids, methotrexate, high-dose thalidomide, vincristine, dapsone, and imatinib with mixed results.

Conclusions:

Diagnosing cutaneous RDD is challenging due to the variations in its clinical and histologic presentation.. This case also emphasizes the challenging nature of accurate clinical and histopathologic diagnosis. Key features of histopathology include emperipolesis and dermal infiltrate of histiocytes that stain positively for S100 and CD68 and negatively for CD1a. Management involves observation although surgical excision, and several treatment options are available if therapy is desired.

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