Cutaneous Rosai-Dorfman Disease: A Case Report

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Abstract

Cutaneous Rosai-Dorfman Disease (CRDD) is a rare form of Rosai-Dorfman Disease (RDD) or sinus histiocytosis with massive lymphadenopathy, which has a varied clinical presentation, an unknown etiology, and multiple treatment options which lack efficacy. We report a case of a 31-year-old African-American female (AAF) who presented with grouped skin-colored and pink papules and plaques within a hyperpigmented patch on her thigh, treated with topical, oral, and intralesional steroids with minimal improvement.

Case Description

History of Present Illness: A 31-year-old AAF presented with a slowly spreading pruritic rash on her right thigh for approximately 1 year. She had previously seen a dermatologist and was prescribed triamcinolone 0.1% cream and bactroban 2% ointment, though declined a biopsy at that time.

Medical History/Surgical History: Anxiety

Social History: Single, sexually active, nonsmoker, no alcohol or drug use

Family History: Eczema, hypertension

Medications: Triamcinolone 0.1% cream and bactroban 2% ointment

Physical Examination: Well-developed, well-nourished black female in no acute distress, alert & oriented x3, skin type V/VI. Grouped skin-colored to light pink papules and plaques within a hyperpigmented patch on the right medial thigh (Figure 1).

Laboratory Data: CBC w/diff revealed leukopenia

Studies: CT of chest/abdomen/pelvis was weil. Punch biopsy was negative for fungal, bacterial, or AFB culture.

Biopsy (Figure 2, 3, & 4): The biopsy specimen shows a dense dermal infiltrate of large histiocytes admixed with inflammatory cells composed predominantly of lymphocytes and plasma cells. The histiocytes within the inflammatory infiltrate have vesicular nuclei and abundant eosinophilic cytoplasm. Areas of emperipolesis (intact inflammatory cells within histiocytes) are noted. The large histiocytes are S100+ and are negative for CD1a.

Final Diagnosis: Cutaneous Rosai-Dorfman disease

Introduction

• Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a non-Langerhans cell histiocytosis.1
• There are two main forms of RDD: One form that affects the lymph nodes and in certain cases the extranodal organs, while the other is purely cutaneous RDD (CRDD).1
• CRDD is extremely rare and the etiology is unknown, though a number of viral and immune causes have been postulated.1
• Approximately 10% of RDD patients exhibit skin lesions, and in 3% it is contained solely in the skin.2
• CRDD presents with median age, 43.5 years, a female predominance (2:1), and most commonly affects Asian and Caucasian individuals.1

Diagnosis

• CRDD presents as solitary or numerous papules, nodules, and/or plaques.1
• Histopathology reveals emperipolesis, the presence of intact lymphocytes (or less often plasma cells, neutrophils, and red blood cells) within histiocytes.1,3
• Histiocytes stain positively for S100 protein, CD4, Factor VIIIa, and CD68 and negatively for CD1a.1,3

Discussion

• Both the clinical and histologic differentials are broad for CRDD. Differential diagnosis includes sarcoidosis, acne vulgaris, lupus vulgaris, granuloma annulare, vasculitis, hidradenitis suppurativa, malignant breast neoplasm, and other histiocytes.
• The most common site of lesions in CRDD is the face, with eyelids and malar regions frequently involved, followed by the back, chest, thigh, and shoulder.1,3
• Rarely CRDD may be associated with the involvement of other disorders, including bilateral uveitis, antinuclear antibody positive lupus erythematosus, rheumatoid arthritis, hypothyroidism, lymphoma and HIV infection.1
• CRDD may be self-limited, yet surgical excision, cryotherapy, local radiation, topical steroids, laser treatment, dapsone, thalidomide, isotreinoin, imatinib, and methotrexate have all been attempted in various case reports in the literature.1-7

Conclusion

• CRDD is an unusual clinical entity with varied lesions
• CRDD follows a benign clinical course, with a possibility of spontaneous remission or various treatments
• Further studies are required to confidently classify the etiology and variance between both RDD and CRDD.

References


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