Single Indurated Brown Nodulo-Plaque in a Young Male

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A 33-year-old man presented to our dermatology clinic with an erythematous to hyperpigmented, gradually progressive, painless plaque with grouped papules on right thigh for 4 months. It was asymptomatic without lymphadenopathy, fever, or weight loss. He denied history of trauma, and preceding or concurrent illnesses. His past medical history was unremarkable. On examination, there was an ill-defined, light-brown to skin colored plaque of size 10 × 12 cm over medial aspect of his right thigh. The plaque consisted of multiple, closely-aggregated, dark-brown hyperpigmented papules and nodules [Figure 1]. The patient was otherwise in good health and had no evidence of systemic disease.

Hematological and biochemical investigations, urinalysis, chest X-ray, and ultrasonography of abdomen were within normal limits. Skin biopsy from the nodular swelling revealed dense and diffuse collections of foamy histiocytes, lymphocytes, histiocytes, plasma cells, neutrophils, and few eosinophils in the dermis. In deep dermis, the infiltrate primarily composed of large foamy histiocytes with evidence of emperipolesis at few foci [Figure 2a-d]. The histiocytes were positive for S-100 and CD-68 on immunohistochemistry [Figure 2e and f]. A magnetic resonance imaging (MRI) of the thigh showed ill-defined homogenously enhancing subcutaneous swelling which showed higher signal to muscle on T1W with peripheral region of relatively high signal and central region of low signal intensity on T2W and STIR images [Figure 3].

What is Your Diagnosis?

Cutaneous Rosai-Dorfman disease

Discussion

The patient did not have any systemic involvement and the lesion was gradually progressive; hence, he was treated with 4 sessions of intralesional triamcinolone 10 mg/ml, every 3 weeks. There was partial improvement with treatment. The hyperpigmentation and nodular swellings improved significantly. The overall size of the lesion decreased and repeat MRI showed decreased signal intensity and reduction in the size of swelling.

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare, benign, self-limiting non-Langerhans cell histiocytosis of unknown etiology that was first described in 1969 by Rosai and Dorfman. This entity most commonly is limited to the lymph nodes; however, more than 40% of patients have extra-nodal involvement, with the skin being the most frequently affected site. About 10% of patients with RDD have skin lesions, and in 3% of cases, the disease is limited to the skin.[1] The features which differentiate cutaneous RDD from systemic RDD include later age of onset (median age, 43.5 years) and a female predominance (2:1) in cutaneous disease.[2] RDD has a median age of onset as 20.6 years and is slightly more common in males (1.4:1). Moreover, cutaneous RDD most commonly affects Asians and Caucasians while the majority of systemic RDD patients are Africans.[2]

Though the etiology of cutaneous RDD is unknown, its association with viruses (Epstein-Barr virus, human herpesvirus 6, and Parvovirus B19) and vaccination has been suggested. The polyclonal nature of the infiltrate in lesions

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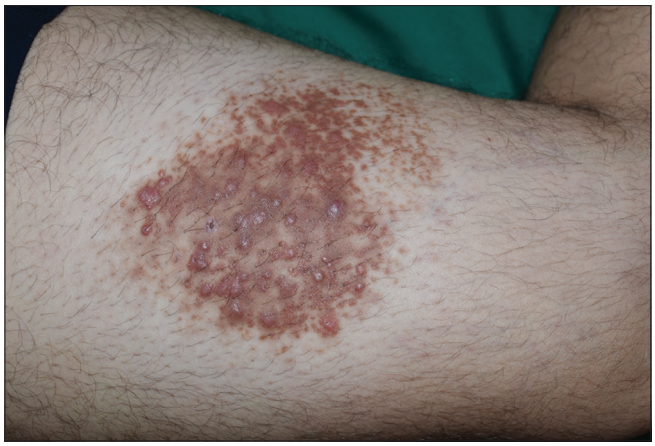

Figure 1: Right thigh showing brown colored swelling with multiple dark brown papules and nodules in the center and small papules in the periphery

and the clinical course of spontaneous remission over months to years suggest that cutaneous RDD is a benign reactive process rather than a neoplastic disorder.

Clinical features of cutaneous RDD are variable and include asymptomatic, solitary or numerous yellow-red to brownish papules, nodules, or plaques. In certain cases, the lesions may present as pustules, acneiform lesions, eruptive xanthoma like lesions and lesions mimicking psoriasis, vasculitis, and panniculitis.[3] Face is the most commonly affected site followed by the back, chest, thigh, flank, and shoulder. Our patient presented with only a skin lesion (subcutaneous swelling with papules and nodules) without systemic involvement. The differential diagnoses considered were sarcoidosis, Kimura's disease, lupus vulgaris, deep fungal infection, and non-Langerhans cell histiocytosis. Sarcoidosis usually presents as erythematous to brown-colored plaques and nodules and biopsy demonstrates non-caseating circumscribed lymphocyte-poor granulomas composed of epitheloid cells, histiocytes, and giant cells, which were not seen in our case. Kimura's disease presents with plaques or nodules and histopathology demonstrates an infiltrate composed of eosinophils and lymphocytes including lymphoid follicles along with abundant vascular channels in the deep dermis or subcutis. The other possibility of lupus vulgaris was less likely in view of lack of lymphadenopathy and scarring in the lesion. The lesions were less inflammatory for deep fungal infection. Histopathology also did not favor any infective pathology. The clinical and histopathological data in our case pointed toward the diagnosis of RDD. Therefore, a final diagnosis of cutaneous Rosai-Dorfman disease was made.

Though the clinical manifestations are variable and non-specific, making early diagnosis of cutaneous RDD challenging, it has the characteristic histopathology and immunohistochemistry. Histopathology typically shows a dense dermal infiltrate of histiocytes with large vesicular nuclei and abundant cytoplasm, accompanied by

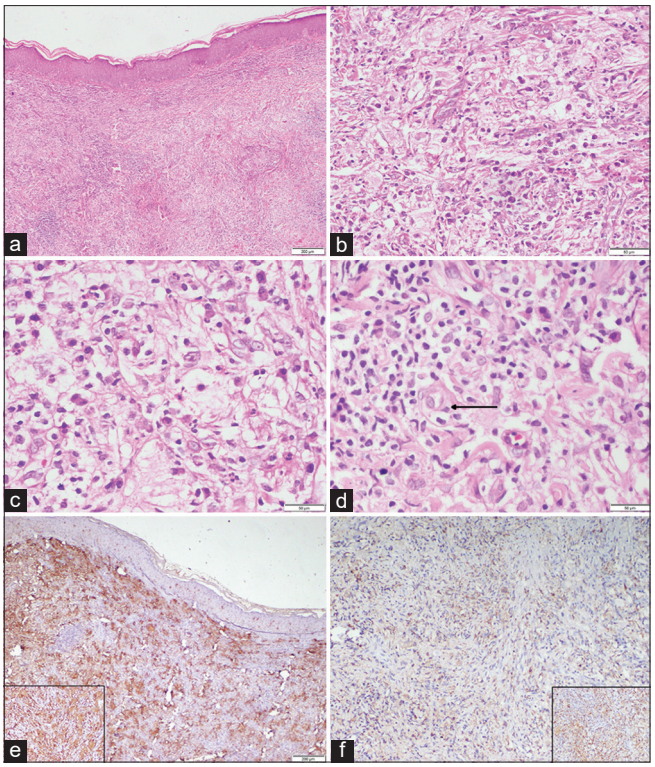

Figure 2: (a) Skin biopsy showing dense and diffuse infiltrate in the superficial, mid, and deep dermis (H and E, 40×), (b) close-up view showing the infiltrate comprising of foamy histiocytes, plasma cells, lymphocytes, histiocytes, and few neutrophils and eosinophils (H and E, 100×), (c) large histiocytes with abundant foamy cytoplasm and large vesicular nuclei (H and E, 200×), (d) higher magnification revealing emperipolesis (H and E, 200×), (e) immunohistochemical staining showed that the histiocytes were positive for S100 protein (IHC, 100 × with inset 200×), (f) immunohistochemistry positive for CD68 protein (IHC, 100 × with inset 200×). H and E, haematoxylin and eosin; IHC, immunohistochemistry

lymphocytes, plasma cells, few neutrophils and eosinophils. Some histiocytes have within their cytoplasm, the engulfed, intact inflammatory cells, i.e. lymphocytes, plasma cells, or neutrophils (emperipolesis). IHC is positive for S100, CD11c, CD14, laminin 5, and lysozyme. CD68 is variably positive, and CD1a is usually negative.[4]

Cutaneous RDD usually has a favorable prognosis with a self-limiting course; however, the condition may be associated with other disorders including bilateral uveitis, lupus erythematosus, rheumatoid arthritis, hypothyroidism, lymphoma, and HIV infection. Due to esthetic reasons and in relapsing cases, a variety of treatment modalities have been used, such as cryotherapy, surgical excision, irradiation, corticosteroids (topical, oral, intralesional), dapsone, thalidomide, methotrexate, isotretinoin, and imatinib with complete to partial response.[5] There are no well-designed studies to draw definitive conclusions regarding the management of this disease. Though our patient had a recent onset and skin-limited disease, he was concerned as it was progressive. Given the localized nature of disease, we treated him with intralesional triamcinolone and achieved good response without local or systemic side effects.

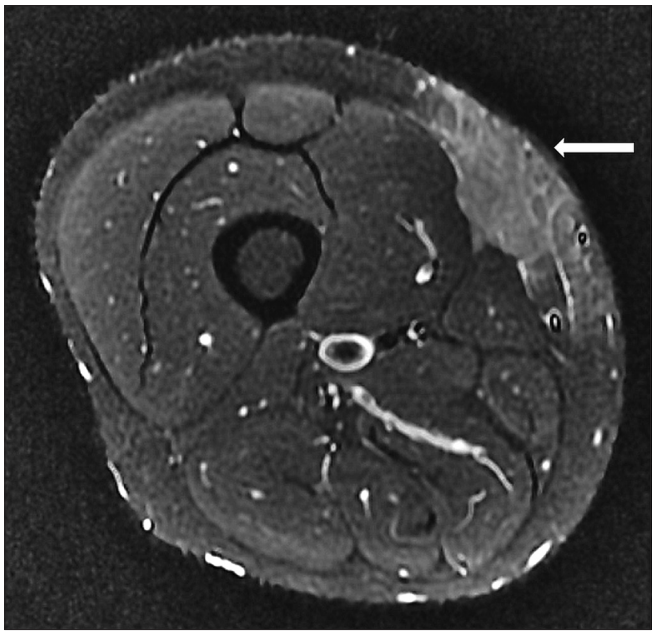

Figure 3: Magnetic resonance imaging showing homogenously enhancing subcutaneous swelling

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Nil.

Conflicts of interest

There are no conflicts of interest.

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