BRIEF ARTICLES

Long Term Follow Up of Cutaneous Sinus Histiocytosis (Rosai-Dorfman Disease)

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ABSTRACT

Rosai-Dorfman disease is a benign histiocytic proliferative disorder of unknown etiology with cutaneous variants clinically presenting with painless cervical lympadenopathy, fever, leukocytosis and other systemic findings.[1] Although the skin is the most common extranodal site, rare purely cutaneous forms of the disease exist and diagnosing such cases rests solely on histopathologic findings.[2]

We report a case with a fifteen year follow up period of this uncommon disorder and describe its clinical course marked by multiple episodic recurrences.

INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy also known as Rosai-Dorfman disease was originally described in 1969.[1] It is a histiocytic proliferative disorder characterized by painless lymphadenopathy, fever, leukocytosis, elevated sedimentation rate and IgG polyclonal hypergammaglobulinemia.[3] Extranodal sites can be affected in 43% of cases with the skin and upper respiratory track most commonly represented.[4] The skin may be the only organ affected in about 10% of cases without lymphadenopathy and may represent a distinct clinical entity.[4,5]

CASE REPORT

A 65-year-old man presented with a reddish brown papule of the right shoulder of several weeks duration in November 2002. The lesion was asymptomatic and he denied any systemic symptoms. Full skin physical exam was unremarkable and no lymphadenopathy was identified. The patient's only pertinent past medical history included hypertension, hyperlipidemia, folliculitis and nonmelanoma skin cancer. A tangential scoop biopsy of the site demonstrated a nodular, diffuse mixed infiltrate composed of sheets of large tissue macrophages that contained abundant eosinophilic cytoplasm. Many of these macrophages contained phagocytized lymphocytes (Figure 1). The infiltrate was admixed with numerous lymphocytes and plasma cells. Eosinophils were not identified. Special stains demonstrated positive strong staining of the large macrophages with S-100 protein stain. In addition, the macrophages stained positive for CD-68 and negative for CD-1a. Special

stains for mycobacteria (AFB) were negative. The histologic changes together with the results of special stains suggested Rosai-Dorfman disease.

Initial workup included a negative serum immune electrophoresis, positive Epstein-Barr viral capsid antigen IgG but negative IgM. Complete blood count was normal, blood urea nitrogen was slightly elevated at 23 (reference range 7.0 to 20 mg/dl) as well as creatinine at 1.6 (reference range 0.5 to 1.5 mg/dl). The lesion resolved following biopsy but over the subsequent years of follow up similar localized lesions formed on the left upper arm (9/2003, 6/2007), right posterior upper arm (6/2007), mid posterior neck (7/2011), right neck (11/2015), left mid chest (11/2015), right mid back (4/2016), left mid lateral back (4/2017), and right chest (11/2017). All lesions were confirmed by biopsy and treated with localized excisional biopsies. In our patient, all new lesions were morphologically alike (reddish brown firm papules) and isolated (Figures, 2,3). During this long follow up period the patient has remained free of systemic symptoms and has never developed lymphadenopathy.

Figure 1. Emperipolesis or phagocytosis of intact lymphocytes by plasma cells (H & E; magnification x 400).

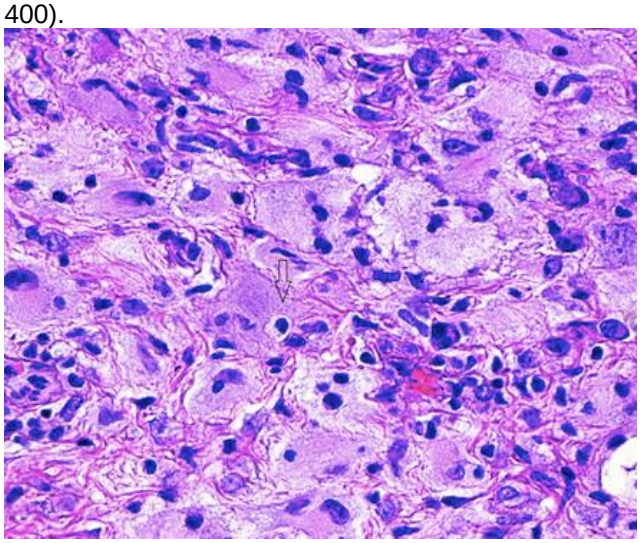


Figure 2. Left mid superior chest reddish brown papule

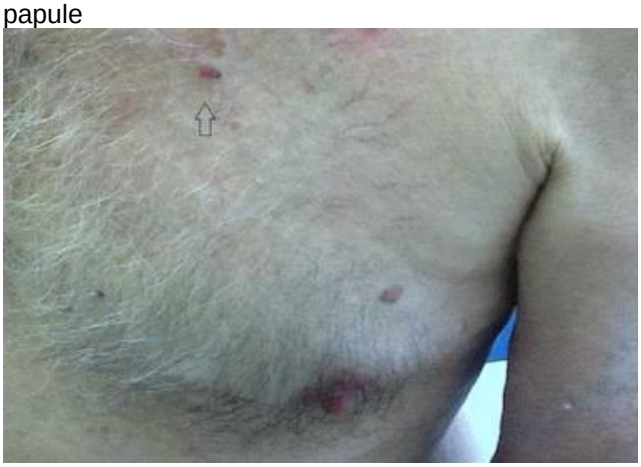
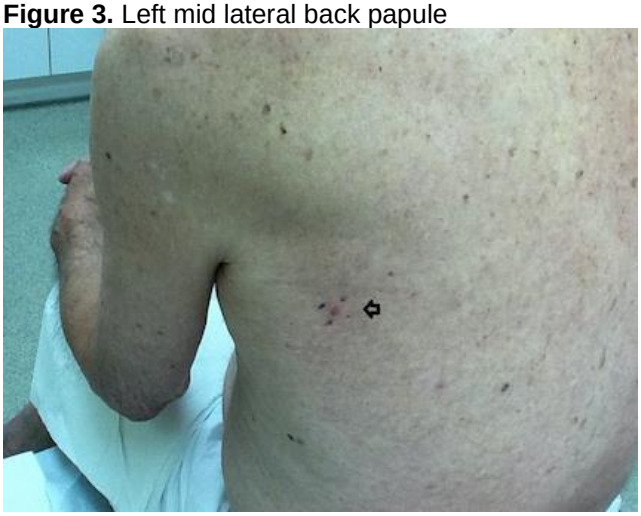


Figure 3. Left mid lateral back papule



DISCUSSION

A review of 22 cases with purely cutaneous Rosai-Dorfman disease documented an older age of onset (median, 43.5 years) with an increased female predominance and most commonly affecting Asian and Caucasian individuals.[6] The clinical morphology of the lesions is nonspecific. The condition is self-limited and of unknown etiology and lesions may be present for months or years and may spontaneously regress. On histology dense histiocytic infiltrates with large vesicular nuclei with prominent nucleoli and eosinophilic cytoplasm which stain positive for S-100,

CD-68 and stain negative for CD-1a are seen.[7] Emperipolesis or phagocytosis by histiocytes of lymphocytes, neutrophils, polymorphonuclear cells or plasma cells is a characteristic finding.[7] Various treatments with varying success have been attempted for cutaneous lesions including: cryotherapy, carbon dioxide laser, cortico-steroids (topically, orally and intralesionally), surgical excision, radiotherapy, dapsone, acitretin, imatinib and thalidomide.[8] A conservative treatment approach is often appropriate.[9] In the current case new lesions were treated with localized excisional biopsies successfully.

CONCLUSION

We present a case with a 15 year follow up of episodically recurrent cutaneous Rosai-Dorfman disease. Our case illustrates the fact that this disease can have a benign clinical course without ever developing lymphadenopathy or systemic symptoms. This case with its unique long term follow up period adds evidence that cutaneous Rosai-Dorfman disease is a distinct clinical disorder compared to cases of sinus histiocytosis with massive lymphadenopathy with secondary cutaneous involvement. Patients with cutaneous Rosai-Dorfman disease should be followed closely and managed conservatively in the absence of systemic symptoms.

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