Case for diagnosis*

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CASE REPORT

A 72-year-old man presented erythematous and scaled plaque on the right neck for three months (Figure 1). The lesion grew progressively in size with no pain and pruritus. He had no symptom of fever, malaise or weight loss. Physical examination revealed no sign of lymph node enlargement in the bilateral parts of neck, axillary fossa and groin. Laboratory investigations indicated that the routine haematological and biochemical studies were normal. The skin biopsy specimen revealed diffuse inflammatory infiltrate consisting of lymphocytes, histiocytes, sparse population of neutrophils and plasma cells in the dermis (Figure 2A). Some histiocytes contained lymphocytes, neutrophils, plasma cells and RBCs within their pale and foamy cytoplasm (Figure 2B). Immunohistochemical staining revealed that histiocytes were positive for S-100 protein and CD68, but negative for CD1a (Figures 2C e 2D).

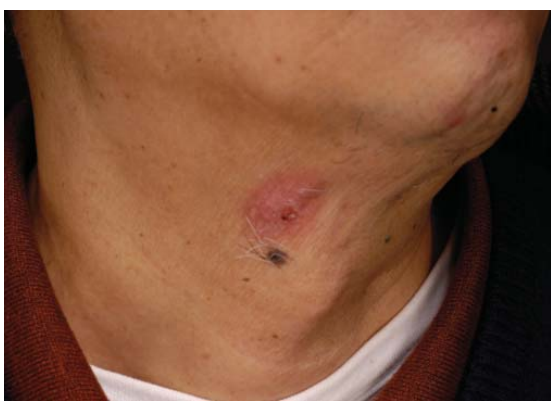

FIGURE 1: Erythematous and scaled plaque on the right neck

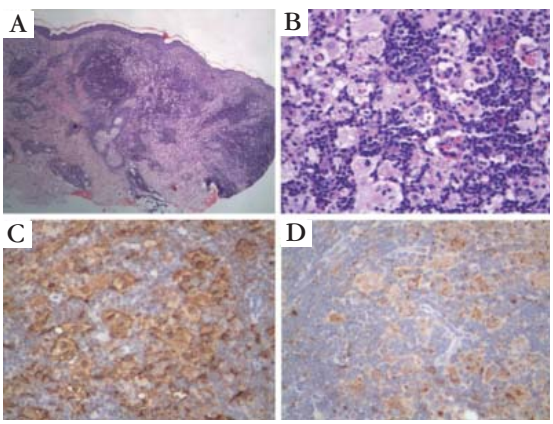

FIGURE 2: Histopathological features. **A -** Diffuse inflammatory infiltrate consisting of neutrophils, plasma cells, lymphocytes and histiocytes in the dermis (hematoxylin-eosin stain, original magnification×40). **B -** Cytophagocytosis of lymphocytes, neutrophils and RBCs (hematoxylin-eosin stain, original magnification×400). Immunopathological features. **C -** S-100 stains (original magnifications×400). **D -** CD68 stains (original magnifications×400)

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DISCUSSION

Based on the clinical and histopathologic character, the diagnosis was CRDD. Owing to local involvement, the lesion was exsected completely and remained asymptomatic with no signs of recurrence at the 9-month follow up.

RDD was first described by Rosai and Dorfman in 1969 as sinus histiocytosis with massive lymphadenopathy.[1] Classically, it presents as bilateral painless cervical and paratracheal lymphadenopathy, which can be massive, in 90% of the cases. Extranodal involvement is most commonly reported in the eyes and ocular adnexa, head and neck region, upper respiratory tract, skin and subcutaneous tissue, and central nervous system. Other sites include the gastrointestinal tract, salivary glands, genitourinary tract, thyroid, breast and testis. Clinically, it can be classified in two forms: an indolent cutaneous form without systemic symptoms of fever, hypergammaglobulinaemia and leukocytosis, and a nodal form with or without systemic symptoms.[2] The clinical course of most cases is a benign self-limiting course and can spontaneously regress without treatment and generally does not relapse. A few reported cases become progressive and require treatment secondary to life-threatening complications or organ dysfunction; prognosis is poor if there is involvement of kidneys, lungs or wide spread nodal dissemination.[3] The purely cutaneous form of the disease is very rare and usually presents as erythematous to brown papules, plaques, or nodules with no predilection for site.[4]

Histologically, CRDD is characterised by a bulk of inflammatory infiltrate consisting of lymphocytes and histiocytes in the dermis, with or without subcutaneous tissue involvement. The phenomenon of emperipolesis is the key point for differentiating RDD from other diseases. Immunohistochemically, the histiocytes show reactivity for S-100 protein and CD68.

The etiology of RDD is still unknown. It has been noted to occur sporadically, with occasional clustering, which suggests a genetic or an infectious component. It has been suggested that this disease is linked to a reactive disorder since it arises from circulating mononuclear cells and there is an increase in auto-immune antibodies in some affected individuals. Epstein-Bar virus and human herpes virus 6 have been proposed as the infectious agent and recently other diseases, including varicella zoster virus, cytomegalovirus, Brucella and Klebsiella, have also been implicated. Meanwhile, it has been postulated that the development of RDD may be driven by irregular cytokine expression.[3]

Various kinds of treatment, such as steroids, chemotherapy, radiation therapy, acitretin and surgery have been applied for this disease, with differing outcomes. Surgery is indicated as the best choice for dealing with the local disease and it often results in long-term remission.[5] In our case, surgical excision of the lesions had been shown to be effective with no relapse. But we must keep in mind that due to its benign course and spontaneous resolution in most cases, less aggressive options are employed whenever possible. ❑

Abstract: Cutaneous Rosai-Dorfman disease is a rare, lymphoproliferative disease. It is benign and self-limited, only involves skin and subcutaneous tissue and typically occurs as histiocyte-rich inflammatory infiltrates, manifesting as erythematous to brown papules, plaques, or nodules, without predilection for site. The authors describe a case of cutaneous Rosai-Dorfman disease in a 72-year-old man who presented erythematous and scaled plaque on the right neck for three months without systemic symptoms. Owing to local involvement, the patient received a surgery to exsect the lesion completely and remained asymptomatic with no signs of recurrence at the 9-month follow up.
Keywords: Histiocytosis, sinus; Neck; Skin Neoplasms

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