Case Report

Purely Cutaneous Rosai-Dorfman disease (CRDD) Co-Existed with Capillary Hemangioma Successfully Treated with Intralesional Corticosteroid

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Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy, is a benign idiopathic proliferative disorder of the histiocyte. Purely Cutaneous Rosai-Dorfman disease (CRDD) is a separated clinical entity without lymph node and organ involvement. The histologic features resemble RDD, but with dermal infiltration. This rare condition is benign and mostly self-limited. The authors report a 66-year-old Thai male patient, diagnosed as purely CRDD, with co-existing capillary hemangioma. In addition, we show that the treatment intralesional corticosteroid can produce the remission of the plaque and tumoral types of this condition.

Keywords: *Rosai-dorfman disease, Capillary hemangioma, Intralesional corticosteroid*

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Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy, is a benign idiopathic proliferative disorder of the histiocyte. This condition was first described in 1969 by Rosai and Dorfman. It is characterized by lymphadenopathy, predominantly on cervical lymph nodes. Constitutional symptoms such as fever, malaise, and myalgia are observed. Laboratory investigation reveals anemia, leukocytosis, increased erythrocyte sedimentation rate, and polyclonal hypergamma globulinemia. The diagnosis is based on histopathology revealing a diffuse infiltration of histiocytes with emperipolesis, the engulfment of inflammatory cells by histiocytes. The mononuclear histiocytes stain positively for S100 but negatively for CD1a.

Purely Cutaneous Rosai-Dorfman disease (CRDD) is a separated clinical entity from RDD[1]. This condition involves only the skin and does not affect lymph node and other organs. It is a rare disease and usually found in Asian, middle-aged female[2-4]. Etiology is still uncertain, but like other reactive conditions of lymphoid tissue, viral stimulus such as Epstein-Barr virus and human herpes virus-6 may be involved in the pathogenesis[3]. Laboratory findings are usually within normal range[3].

The authors describe a Thai male patient presenting with asymptomatic skin lesions. The way to reach definite diagnosis and treatment will be presented.

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Case Report

A 66-year-old Thai male presented with multiple asymptomatic papules and plaques, which gradually developed on the right flank for seven years. He was previously treated as chronic eczema with topical corticosteroid for many years, but the improvement was not seen. In contrast, some lesions progressed to nodular masses, and there was an increase in the number of papules. Examination revealed three discrete well-defined, erythematous, exophytic masses and erythematous infiltrated plaques surrounded by numerous papules (Fig. 1, left). No other systemic symptoms were observed.

Histopathology of a skin biopsy from the plaque on the right flank showed dense diffuse mixed inflammatory cell infiltration and foamy histiocytes with the presence of emperipolesis (Fig. 2). Immunohistochemistry staining was positive for S100 and negative for CD1a (Fig. 3). Systemic work up for

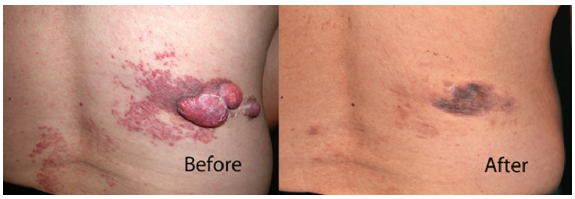

Fig. 1 3 nodules on top erythematous plaque at right flank (left) and the nodules resolute after intralesional steroid treatment (right).

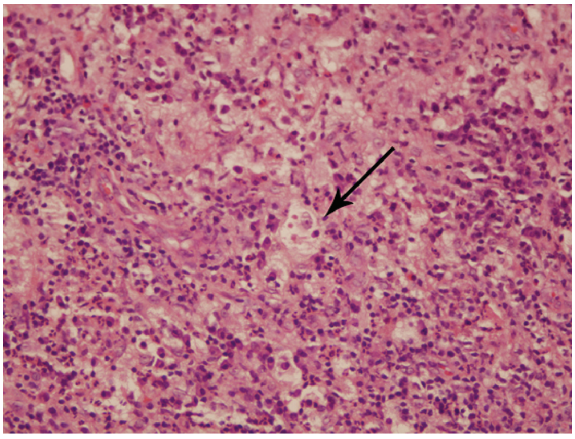

Fig. 2 Biopsy showed dense diffuse inflammatory cell infiltration and foamy histiocytes with emperipolesis (H&E x400).

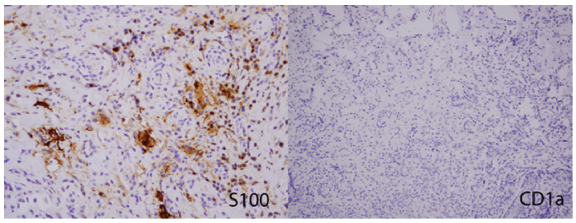

Fig. 3 Showed positive S100 (left) and negative CD1a (right).

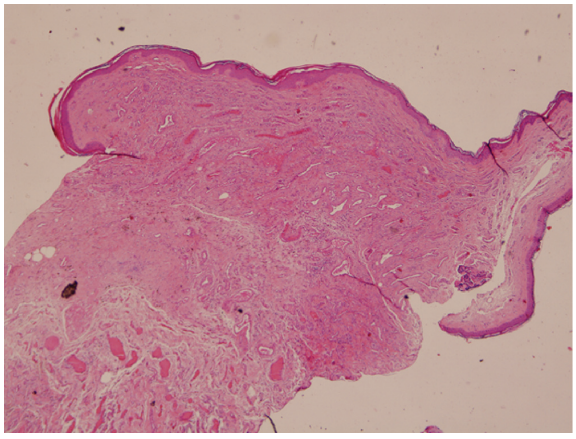

Fig. 4 Incidental finding capillary hemangioma (H&E x40).

hematologic involvement and serum electrophoresis were within normal limit. Abdominal ultrasound and eye examination were normal. Therefore, he was diagnosed as purely CRDD.

The authors decided to treat him with intralesional corticosteroid injection because of the enlargement of lesions and the unresolving lesions after years of topical therapies. After we treated him with intralesional corticosteroid 40 mg/ml every few months for three years, the nodular masses were significant flatten. The skin biopsy was retaken for a follow-up. Histopathology shows a similar pattern to the previous biopsy. However, there was proliferation of small to medium-sized blood vessels in the upper dermis compatible with capillary hemangioma, which was not seen in the first biopsy (Fig. 4). The diagnosis was purely CRDD co-existing with capillary hemangioma. The skin lesions have been resolved after intralesional corticosteroid injections for five years (Fig. 1, right).

Discussion

This 66-year-old Thai male patient was diagnosed as purely CRDD based on the histopathological result of histiocytes with emperipolesis and immunohistochemistry staining.

Clinical characteristic of CRDD has three main types[4]: papulonodular, indurated plaque, and tumor type. Cutaneous lesions are often asymptomatic and can be single or widespread lesions with no predilection site[2,3,6]. Differential diagnosis depends on clinical presentations, such as other cutaneous histiocytosis, as well as many other skin diseases such as lymphocytoma cutis, sarcoidosis, dermatofibrosarcoma protuberans, or pyogenic granuloma[3].

The histopathology renders the correct diagnosis[7]. It is characterized by a diffuse dermal infiltration composed of numerous histiocytes and inflammatory cells engulfed into the cytoplasm of histiocytes forming a characteristic of emperipolesis[2,3]. S100 staining is positive and CD1a staining is usually negative in contrast to Langerhan cell histiocytosis[2-4].

Prognosis of CRDD is variable. There are spontaneous regression within months, but lesions may persist for years[2-4,8]. Total excision may be useful for solitary and small lesions[8], while in multiple or widespread CRDD, treatment of choice are topical and

intralesional steroid, systemic steroids, methotrexate, retinoid, thalidomide, cryosurgery and radiation[1,2,8,9].

Our patient who presented with three clinical types showed clinical improvement from intralesional corticosteroid. It is possible that the clinical improvement resulted from the corticosteroid injections since the large lesions had not been resolved within many years before we started the injection therapy.

We present a case of CRDD with co-existed capillary hemangioma which was not seen in the first biopsy. Up to our knowledge, this coincidence has not been reported yet. The authors show the evidence that intralesional corticosteroid may produce the remission of the plaque and tumor types of CRDD. CRDD is a rare disease and is difficult to diagnose. The histopathology and special immunohistochemistry is needed for the definite diagnosis.

Potential conflicts of interest

None.

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ผู้ป่วย purely cutaneous Rosai-Dorfman disease (CRDD) ที่พบร่วมกับ capillary hemangioma ประสบความสำเร็จกับการรักษาด้วยการฉีดสเตียรอยด์: รายงานผู้ป่วย

วัลลภา อัศวเบ็ญจาง, สุเทพ จีระสุทัศน์, ปิ่นนรี ขัตติพัฒนาพงษ์, พัชรินทร์ จันทร์จำรัสแสง, ชินมนัส ตั้งจาตุรนต์รัศมี

Rosai-Dorfman disease (RDD) หรือ sinus histiocytosis with massive lymphadenopathy, เป็นเนื้องอกของ histiocyte เจริญเติบโตผิดปกติโดยไม่ทราบสาเหตุ purely cutaneous Rosai-Dorfman disease (CRDD) มีลักษณะทางคลินิกที่เป็นเอกลักษณ์ออกไปคือไม่มีอาการผิดปกติของต่อมน้ำเหลืองและอวัยวะอื่นร่วมด้วย แต่มีลักษณะทางพยาธิวิทยาของรอยโรคที่ผิวหนังคล้ายกันกับ RDD โรคนี้พบได้น้อยมากแต่การดำเนินโรคดี สามารถควบคุมโรคได้ ผู้นิพนธ์ได้รายงานผู้ป่วยชายไทย อายุ 66 ปี ที่ได้รับการวินิจฉัยว่าเป็น purely CRDD ที่พบร่วมกับ capillary hemangioma ผู้ป่วยตอบสนองต่อการรักษาด้วยการฉีดสเตียรอยด์ในผื่นรอยโรคชนิด plaque and tumor

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