

Rosai-Dorfman Disease Presenting as Large Pigmented Indurated Plaques

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Sir,

Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy, is considered a disorder of histiocytes with unknown aetiology (1–6). It fundamentally affects lymph nodes, preferentially the cervical ones, but the skin is one of the most common sites of extranodal involvement (3, 4). The cutaneous lesions of RDD are mainly papules or nodules, often with a xanthomatous, erythematous or purple appearance (2). We describe a case of RDD with hyperpigmented indurated plaques with hypertrichosis.

CASE REPORT

A 35-year-old Japanese man was referred to our university hospital with a 13-year history of pigmented lesions, mainly on the lower extremities. He had lost the sight of his right eye at the age of 5, of his left eye at the age of 9 and his bilateral auditory sense due to perceptive deafness at the age of 18.

Physical examination revealed palm-sized hyperpigmented indurated plaques with hypertrichosis on the chest, abdomen, back and bilateral arms (Fig. 1a). On the thighs and legs, they were coalesced into diffuse large indurated hyperpigmented plaques. The areas were warmer than non-affected areas. Several bean-sized brownish nodules were observed in the popliteal areas (Fig. 1b). There was no regional lymphadenopathy and he was afebrile.

Laboratory examinations showed increased platelet counts ($42.2 \times 10^4/\mu\text{l}$), increased erythrocyte sedimentation rate (ESR) (56 mm/h) and increased serum levels of soluble interleukin-2 receptor (724 U/ml, normal < 530). IgG, but not IgM, antibody for anti-viral capsid antigen (VCA) of Epstein-Barr virus (EBV) and anti-EBV-encoded nuclear antibodies (EBNA) were positive. Antibodies for EBV early antigen were negative. Serum and urine protein electrophoresis indicated no paraprotein or Bence Jones protein.

Cranial and cervical computed tomography showed



Fig. 1. (a) Palm-sized hyperpigmented indurated plaques with hypertrichosis on the arm. (b) The plaques were coalesced into a diffuse indurated hyperpigmented area on the thighs and legs with several bean-sized nodules in the popliteal area (arrow).

neither lymphadenopathy nor any mass suggesting cellular infiltration. Abdominal ultrasonography and bone marrow biopsy were normal.

Histological examination of the skin taken from the nodule at the popliteal area showed dense nodular infiltrates composed mainly of histiocytes. There were also infiltrates of lymphocytes and plasma cells, especially around the small vessels at the periphery of the lesion. No atypical lymphocyte was present. The skin specimen from hyperpigmented plaque showed basal pigmentation and less dense diffuse cellular infiltrates in the dermis composed of the same cells. Some histiocytes displayed emperipolesis of lymphocytes. Immunohistochemically, histiocytes were positive for S-100 protein and CD68, but negative for CD1a. In situ hybridization for EBV-associated latent small nuclear RNAs (EBER) was negative. Electron microscopic examination demonstrated no Birbeck granules. T-cell receptor gene rearrangement studies showed no evidence of monoclonal proliferation. He was diagnosed as having RDD. Since there was no evidence of vital organ involvement, all affected areas were treated with a topical corticosteroid (0.12% betamethasone valerate) for 2 years, but with limited effect.

DISCUSSION

RDD, or sinus histiocytosis with massive lymphadenopathy, is a benign, generally self-limited disease and is usually accompanied by fever, leucocytosis with neutrophilia, elevated ESR, or polyclonal or monoclonal hypergammaglobulinaemia (3–5).

Extranodal involvement occurs in 30–40% of cases. Skin, skeleton, salivary gland, central nervous system, eyes and upper respiratory tract can be involved (2–6). Our case had lost bilateral eye sight and auditory sense, but there was no evidence of infiltrates on cranial computed tomography. The eyes and ears may have been involved previously and the infiltration may have regressed spontaneously, because the bilateral involvement of eyes or ears is reported (7, 8) and clinical remission is sometimes observed in RDD (2, 5, 6). The diagnosis of RDD was made on the basis of the histological findings, the presence of emperipolesis, absence of Birbeck granules or individual cells atypia, and the result of immunohistochemical stainings. His cutaneous

lesions, large hyperpigmented indurated plaques with hypertrichosis, however, were not a typical presentation of the disease.

Previous reports have shown a wide range of clinical presentations, including psoriasiform generalized exfoliative dermatitis and eczema, granuloma annulare-like giant lesions, pustular and acneiform lesions and cystic nodules, red-orange papules with tenderness, multiple hyperpigmented panniculitic plaques, firm red-violaceous nodules with irregular surface surrounded by satellite papules, a pruritic papulosquamous rash with a positive Köbner phenomenon resembling guttate psoriasis and lesions with brownish pigmentation (2, 3). These indicate that RDD can manifest various cutaneous presentations.

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