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QUIZ SECTION

Solitary Tumour on the Neck: A Quiz

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A 70-year-old Japanese woman presented with a lesion on the neck that had increased in size gradually over the course of one year. The initial physical examination revealed a hard, asymptomatic, rose-pink tumour with a diameter of 2.5 cm on the right posterior neck (Fig. 1). No superficial lymphadenopathy was identified. She had no fever and results from routine laboratory tests were within normal range. Computed tomography detected no evidence of visceral malignancy or lymphadenopathy. Histological examination showed infiltrates of foamy histiocytes with large vesicular nuclei and abundant cytoplasm in the dermis (Fig. 2 a, b). Phagocytosis was found where the mononuclear cells seem to have been absorbed without being attacked or digested by enzymes (Fig. 2c). Immunohistologically, the phagocytic cells were positive for CD68 (Fig. 2d) but not for CD1a.

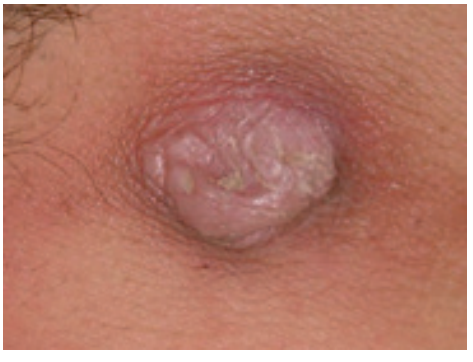

Fig. 1. (a) A hard, rose-pink tumour of 2.5 × 1.7 × 1.5 cm in diameter is observed on the right posterior neck.

What is your diagnosis? See next page for answer.

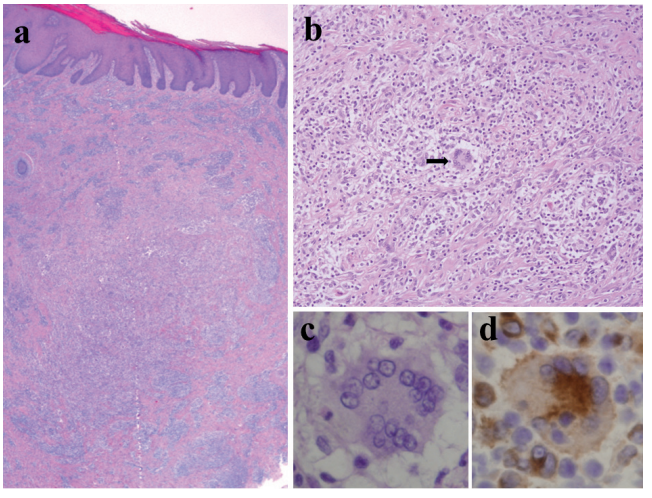

Fig. 2. (a) Histopathologically, numerous lymphocytes, plasmocytes and histiocytes infiltrate the dermis. (b, c, d) Characteristic emperipolesis (arrow) with positive immunohistochemical pattern of scattered CD68.

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ANSWERS TO QUIZ

Solitary Tumour on the Neck: A Comment

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Diagnosis: Cutaneous Rosai-Dorfman disease

Rosai-Dorfman disease (RDD) is a rare histiocytic proliferative disorder that is characterised by its histopathological features (1). RDD is currently considered to be a reactive proliferation of histiocytes rather than a malignant histiocytosis. The clinical presentation of RDD is usually with painless lymph node enlargement, which may reach massive proportions. Extranodal involvement is common with 43% of patients having at least one extranodal site of involvement (2). The skin is the most common extranodal site and it may be involved without nodal disease. Cutaneous RDD (cRDD) is a rare form of RDD that is limited to the skin, which is reported in approximately 3% of RDD cases (3). cRDD usually develops in middle aged or older individuals, and there is a greater female predominance than in systemic RDD (4). The clinicopathological diagnosis of cRDD can be challenging, with different clinical profiles from those of the nodal counterpart and occasionally misleading histological images. Histopathological manifestations are therefore vital to diagnose this rare disease. The lesions consist of infiltrates of foamy histiocytes, lymphocytes and plasma cells, and the characteristic feature is histiocytes with large vesicular nuclei and abundant cytoplasm in the dermis. Histiocytes frequently exhibit the active penetration of one cell by another which remains intact, a phenomenon called emperipolesis (5). The histiocytes are immunohistochemically positive for CD68 but negative for CD 1a (5). Evidence of emperipolesis provides a definitive diagnosis of RDD. Although some reports have indicated that RDD is triggered by viral infection, such as of human herpes virus, human immunodeficiency virus or Epstein-Barr virus, the aetiology of the disease is poorly understood (6). cRDD is a benign disorder with good prognosis when the disease is limited (6). Treatment options for cRDD include surgery, liquid nitrogen, radiation therapy, systemic glucocorticosteroid, and thalidmide regimens (7). In the present case, we performed total excision of the tumour because of the localisation of the lesion. Dermatologists should keep this rare disorder in mind when they encounter these characteristic pathological findings.

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