Subcutaneous Sarcoidosis with Sarcoidal Polyneuropathy

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Sir,

We describe here a rare case of subcutaneous sarcoidosis with perineurial granulomas. A 70-year-old woman was referred to a neurologist in our hospital because of mixed paralysis of the limbs. A chest radiograph revealed bilateral hilar lymphadenopathy. The results of the electrophysiological tests revealed polyneuropathy. The tuberculin test was negative, following a previous positive response. The serum angiotensin-converting enzyme level was 27.7 IU/l (normal range 8.3–21.4 IU/l), but the results of other routine laboratory tests were within the normal limits. Since she was suspected to have sarcoidosis, she was referred to our department for an evaluation of cutaneous sarcoid lesions. She had noticed two nodules without any subjective symptoms on her lower extremities for the past several years. Physical examination revealed a 60×10 mm intramuscular hard nodule on her left leg and a 25×5 mm cord-like subcutaneous nodule that moved freely on her right knee (Fig. 1). Histologically, a biopsy specimen taken from the intramuscular lesion showed typical sarcoidal granulomas displacing adjacent muscle fibres. It consisted mainly of epithelioid cells, giant cells of Langerhans' type, and a few lymphocytes. On the other hand, the nodule taken from the lesion of the right knee showed a white-coloured long fibrous cord, running between subcutaneous fat tissue and aponeurosis of the knee. A histological examination showed many peripheral nerves with perineurial epithelioid granulomas in continuity with perivascular lymphohistiocytic inflammation (Fig. 2). A diagnosis of sarcoidosis with neuropathy was made based on these findings. The patient was treated with oral prednisolone at a dosage of 50 mg daily, and the related symptoms thereafter gradually improved.

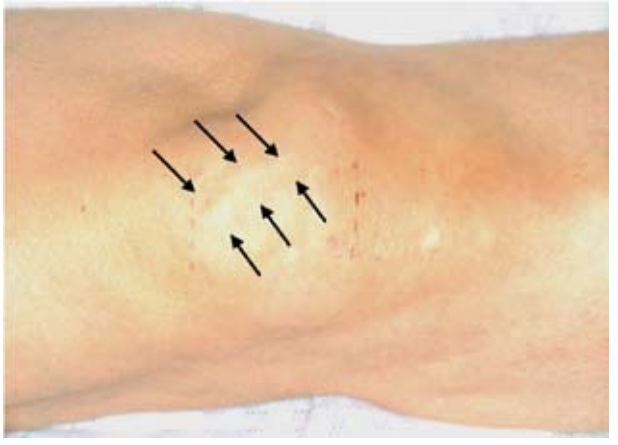

Fig. 1. A cord-like subcutaneous nodule on the right knee.

DISCUSSION

Sarcoidosis is a systemic granulomatous disease of unknown cause that commonly affects the lungs, lymph nodes, eyes and skin (1). Cutaneous sarcoidosis occurs in about 15% of such patients (2) and demonstrates a variety of clinical manifestations (3). The central or peripheral nervous system is affected in about 5% of cases of sarcoidosis, while peripheral nerve involvement is seen in about two-thirds of these cases (4). In the case described here, many peripheral nerves with epineural granulomas were seen in the lesion, which was probably related to sarcoid peripheral neuropathy. Although it is extremely rare to find nerve granulomas in nerve biopsy specimens from patients with sarcoid neuropathy (5), axonal degeneration with perineurial granulomas in a sural nerve biopsy has been reported to be associated with sarcoid neuropathy (6). In addition to nerve granulomas, vascu-

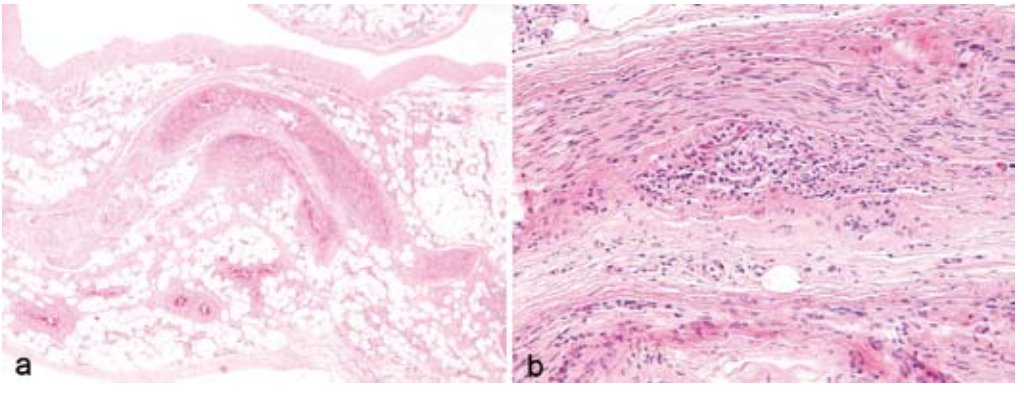

Fig. 2. Peripheral nerves with perineurial epithelioid granulomas in continuity with both perivascular lymphocytic and granulomatous inflammation (haematoxylin and eosin (a) ×100, (b) ×400).

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litis by granulomatous infiltration has been observed in some patients with sarcoid peripheral neuropathy (5), but was not demonstrated in the present case. Therefore, the neuropathy in this case was probably caused by compression from perineurial granuloma formation.

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