

SHORT COMMUNICATION

Malignant Peripheral Nerve Sheath Tumour Arising From Solitary Sclerotic Neurofibroma

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Neurofibroma is a benign neoplasm of the peripheral nerve, which occurs as multiple tumours in neurofibromatosis type 1 (NF1) or as a solitary tumour with no association with NF1. Malignant change is sometimes reported in neurofibroma in NF1, but there are few reports of malignant transformation of solitary neurofibroma. We report here a case of malignant change in a solitary sclerotic neurofibroma following 3 recurrences.

CASE REPORT

The patient was a 77-year-old Japanese woman. In December 2006, a nodule appeared on her left buttock and was surgically excised. It was diagnosed as solitary sclerotic neurofibroma, and this case has been reported elsewhere (1). Histopathologically, the tumour was ill-defined. It recurred twice and was excised in October 2009 and November 2012, respectively. In May 2015, a subcutaneous nodule, 35 × 30 mm in diameter, which was highly movable on fatty tissue, appeared again on the scar after previous resections on the left buttock (Fig. 1). The nodule was excised with a 1-cm margin in August 2015.

Histopathologically, the tumour was ill-defined, and showed massive nodular growth, mainly in subcutaneous tissue (Fig. 2a). There was a relatively high density of neoplastic spindle cells, which were atypical in appearance (Fig. 2b), with large hyperchromatic nuclei, but no pleomorphism. There was a relatively high incidence of mitoses. The intralésional collagen bundles were thinner and were decreased in number, compared with the previous lesions of sclerotic neurofibroma. Distribution of small blood vessel and myxoid areas were also observed. In addition, there were areas identical to the previous lesion of sclerotic neurofibroma around the central lesion (Fig. 2c), which had patchy and chronic infiltration of lymphocytes (Fig. 2d).

Immunostaining revealed: CD34: focal positive; S100 protein: focal positive; epithelial membrane antigen: focal



Fig. 1. Clinical appearance of the tumour on the scar after previous resections on the left buttock.

positive; α -smooth muscle actin: negative; desmin: negative; calponin: negative; and anaplastic lymphoma kinase: negative (not shown).

Based on the above findings, the tumour was diagnosed as a low-grade malignant peripheral nerve sheath tumour (MPNST).

DISCUSSION

Sclerotic neurofibroma is a rare variant of neurofibroma. González-Vela et al. (2) reported only one case of sclerotic variant out of 340 cases of neurofibroma. The neoplasm is usually a well-circumscribed, flesh-coloured, slow-growing tumour. Histopathologically, the lesion contains many fibroblasts and abundant collagen bundles compared with the common type of

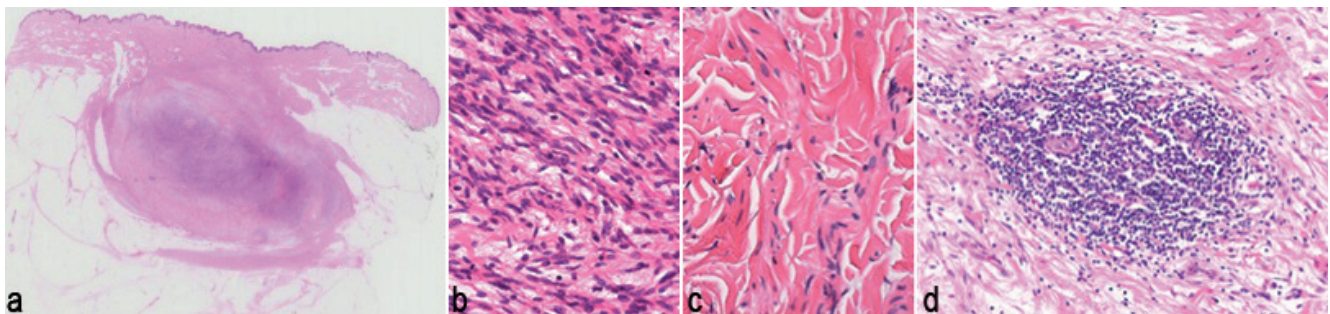


Fig. 2 (a). Overall histopathological view of the tumour. Note the central hypercellular lesion (haematoxylin and eosin; H&E × 20). (b) Proliferation of atypical spindle cells in the central lesion (H&E × 400). (c) Areas around the hypercellular central lesion. Note the characteristics of sclerotic neurofibroma (H&E × 400). (d) Patchy and chronic lymphocytic infiltration in areas of sclerotic neurofibroma (H&E × 100).

neurofibroma, and it is noteworthy that mast cells are located in the vicinity. These mast cells are thought to play an important role in the sclerotic change in the tumour (1).

Evans (3) reported 3 cases of solitary neurofibroma showing malignant change and summarized 13 previously reported cases. These rarely metastasized (2 cases), but had often recurred locally before malignant change (6 cases). Two of these cases recurred more than twice. Interestingly, Evans demonstrated that a focal to patchy and chronic lymphocytic infiltration was seen in the components of neurofibroma, as shown in our case.

It is known that pleomorphic adenoma, a benign tumour of the salivary gland, shows malignant change after incomplete excision (4). Although malignant change in the sclerotic variant of neurofibroma has not been reported previously, in the case reported here incomplete excision due to an ill-defined border caused 3 instances of recurrence, which probably resulted in malignant change, as has been shown in pleomorphic adenoma (4).

On the other hand, dermatofibrosarcoma protuberans (DFSP) is an ill-defined, low-grade mesenchymal tumour. Histopathologically, the grade of DFSP worsens after each recurrence (5). Based on these facts, it is possible that ill-defined sclerotic neurofibroma has a

high potential for low-grade malignancy, with the grade worsening after each recurrence.

Based on our findings, in order to reduce the risk of malignant change, ill-defined sclerotic neurofibroma should be excised with sufficient margins, and particular attention is required when patchy and chronic lymphocytic infiltration is observed in the tumour.

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