Intraneural granular cell tumor of the dorsal ramus of a thoracic nerve

Sir,

We present a rare case of granular cell tumor (GCT)

arising from the dorsal ramus of the thoracic nerve. A 23-year-old man presented with a 6-month history of tenderness in the right side of his back. A physical examination revealed a tender 10 mm × 10 mm mass that was smooth and firm and not fixed to the skin or underlying tissues [Figure 1]. Ultrasonography revealed a hypoechoic spindle-shaped mass beneath the superficial fascial layer. The shape and accompanying tenderness of the mass led to the clinical diagnosis of a schwannoma, and the mass was surgically excised. During surgery, we detected a fusiform mass continuous with a cord-like tissue running laterally, which was thought to be the cutaneous branch of the dorsal ramus of the eighth thoracic nerve. The mass could not be dissected from the cord-like tissue. The surrounding soft tissue was not involved. The mass and 2–3 mm of the cord-like tissue, proximally and distally, were excised.

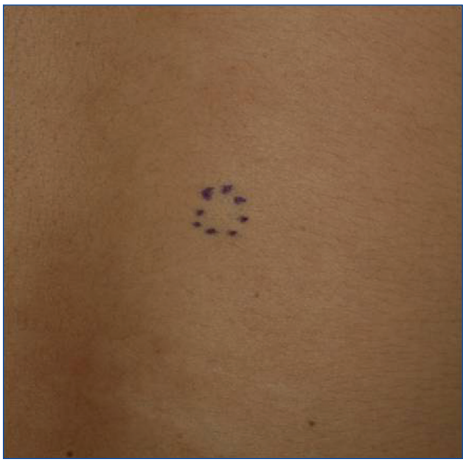

Figure 1: A tender 10 mm × 10 mm subcutaneous mass

Microscopically, the lesion was an elongated mass continuous with the peripheral nerve and surrounded by a thin layer of fibrovascular tissue. Most of the lesion was composed of a proliferation of round to polygonal cells with voluminous pink granular cytoplasm [Figure 2a]. The tumor had round nuclei with no atypia or pleomorphism. In a part of the lesion, the intact peripheral nerve was enveloped by and merged with tumor cells [Figure 2b]. Although the intact peripheral

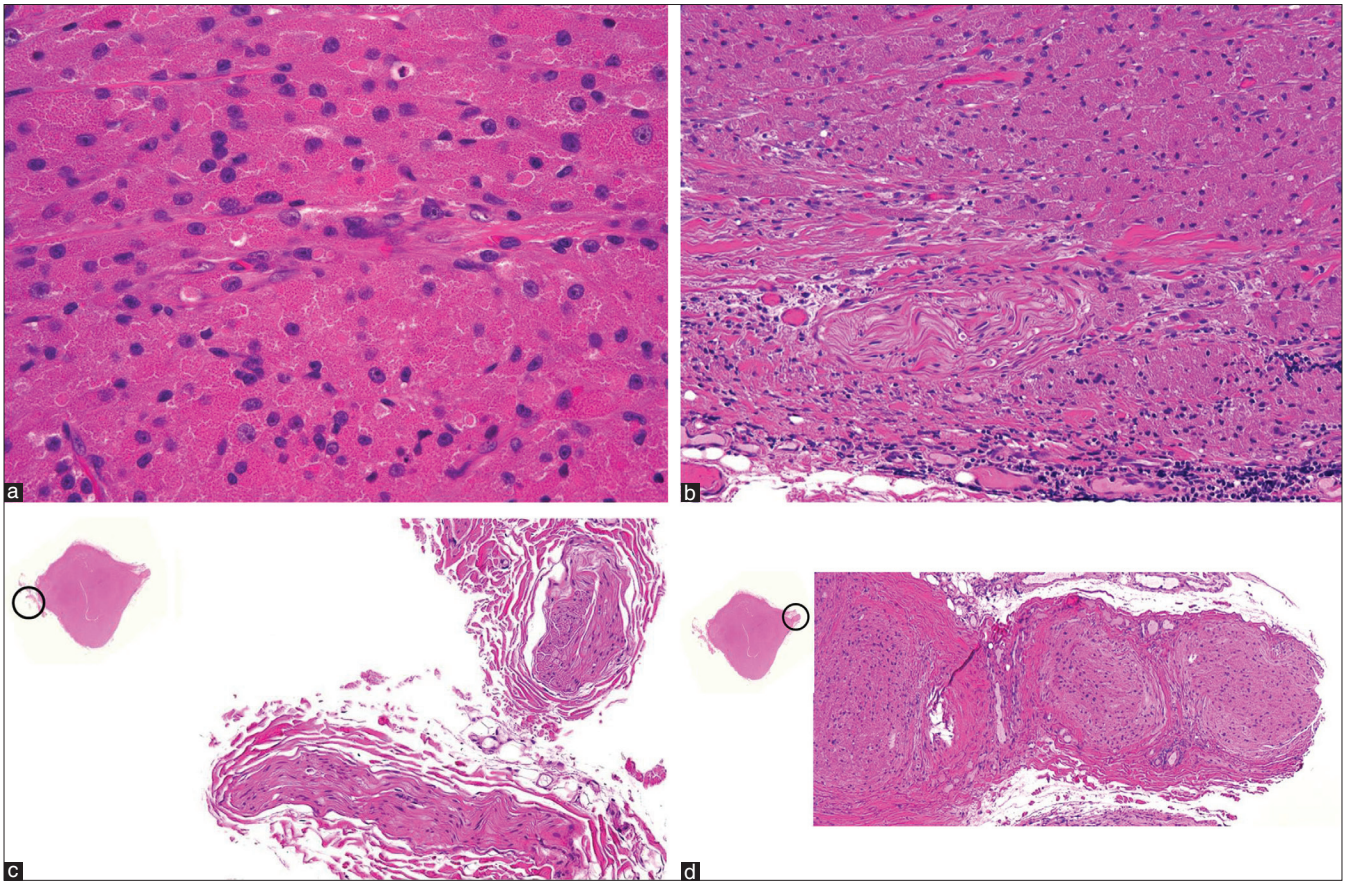

Figure 2: (a) The tumor was composed of round to polygonal cell proliferation with voluminous pink granular cytoplasm (H and E, ×300) (b) In a part of the lesion, the intact peripheral nerve was enveloped by and merged with tumor cells (H and E, ×100) (c) The intact peripheral nerve was observed at one end of the cord-like tissue (H and E, ×100 magnified image of the circle in the loupe magnification at the left) (d) The tumor cells extended to the other end of the cord-like tissue (H and E, ×40 magnified image of the circle in the loupe magnification at the left)

nerve was observed at one end of the cord-like tissue, the tumor cells extended to the other end [Figure 2c and d]. Many tumor cells were strongly positive for the S-100 protein, neuron-specific enolase and CD68. These findings were consistent with an intraneural GCT.

A wide local excision with 1 cm margins from the scar was performed because of a positive margin in the initial operation, and remnants of the tumor cells were not histologically observed. There was no recurrence or metastasis 6 months after surgery.

GCT is a relatively uncommon lesion and usually manifests as a solitary, painless nodule located in the dermis or subcutis, and less frequently in the submucosa. Although many reports suggest that these tumors originate from a Schwann cell, GCT of the peripheral nerve is extremely rare.[1] To the best of our knowledge, only 11 cases (including our case) of benign intraneural GCTs in soft tissue have been reported.[1] The average age of the patients was 28.6 years and the average longest diameter was 1.6 cm. The digital nerve was involved most often with four of the cases; moreover, nine of the 11 cases occurred in an upper extremity. All the cases had some symptoms or clinical findings of nerve compression, including tenderness, paresthesia and muscle weakness or a positive Tinel's sign.

The most effective treatment for GCT in nearly all extraneural cases is local excision with tumor-free surgical margins to prevent recurrence. However, in the peripheral nerves, the tumor tends to infiltrate the nerve trunks and is often unresectable from the nerve.[1] This is unlike a Schwannoma, which is well encapsulated and tends to easily separate from the peripheral nerve. Of the 11 intraneural GCT cases, enucleation could be performed in only two cases. Therefore, when a major peripheral nerve is entirely involved, segmental nerve resection and nerve reconstruction may be necessary.

Fanburg-Smith *et al*.[2] proposed that neoplasms that satisfied three or more of the six histological criteria (necrosis, spindling, large vesicular nuclei, increased mitotic activity, high nuclear-cytoplasmic ratio and pleomorphism) could be classified as histologically malignant. However, it is particularly difficult to accurately differentiate malignant GCTs from benign ones based on pathological findings alone, as metastases can occur in histologically benign tumors as well. Strong *et al*.[3] suggested that the size of the tumor (greater than 3 cm in diameter), rapidity of its growth and invasion of adjacent structures were clinically suggestive of malignancy. Our patient satisfied neither Fanburg's histological criteria nor Strong's clinical findings. However, close follow-up is necessary because malignant transformation of a benign GCT has been suggested and some patients have progressed to histological malignancy after recurrence.[4,5]

Our report would be helpful for better understanding of the intraneural GCT and the differential diagnosis of soft tissue tumors. Although they are relatively rare, intraneural GCTs should be included in the differential diagnosis of soft tissue tumors with neurological symptoms indicative of Schwannomas or nodular plexiform neurofibromas, as the surgical approach and prognosis of intraneural GCTs may be different from that of these tumors.

Yoshiyuki Nakamura, Yuka Iino, Yasuhiro Nakamura, Yasuhiro Fujisawa, Yasuhiro Kawachi, Fujio Otsuka

Department of Dermatology, Institute of Clinical Medicine, University of Tsukuba, 1-1-1, Ten-nodai, Tsukuba, Ibaraki 305-8575, Japan

Address for correspondence: Dr. Yoshiyuki Nakamura, Department of Dermatology, Institute of Clinical Medicine, University of Tsukuba, 1-1-1, Ten-nodai, Tsukuba, Ibaraki 305-8575, Japan.
E-mail: ynakamura-tuk@umin.ac.jp

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