A Well-defined, Non-encapsulated Tumor Composed of Delicate Spindle Cells in the Lower Extremity

Hsin Huang     Chao-Hong Liu     Shu-Rung Shei     Tzei-Yi Lin*

CASE REPORT

A 30-year-old woman had a slowly but progressively enlarging nodule on her right lower legs for more than 3 years. No neurological symptoms or signs were noted.

The detailed dermatological finding revealed a well-defined, smooth-contured, immovable, flesh-colored, and rounded nodule, with firm consistency, located in the deep skin (Fig. 1). A yellowish-white, ovoid mass measuring 1.4 x 1.2 x 1.0cm was excised.

Microscopically, it showed a well-defined, non-encapsulated nodule in the deep dermis. Proliferating spindle cells with delicate processes arranged in elongated bundles, interwoven fascicles, whorls, or loose storiform structures were seen. Intercellular "crackings" were also seen without significant onion-bulb formation nor discernible enlarged nerve twigs (Fig. 2). Neither mitosis nor tumor necrosis was noted.

These spindle tumor cells revealed immunohistochemically diffuse and strong cytoplasmic expression of EMA (Fig. 3), and nonexpression of S-100 protein, neurofilament, smooth muscle actin (clone 851), or desmin.

Fig. 1
A well-defined, immovable, and rounded nodule measuring 1.5 cm in diameter, with elastic to firm consistency, located in the deep portion of skin of the right lower leg.

Fig. 2
Spindle cells with delicate processes arranged in the interwoven fascicles forming loose storiform structures. Intercellular "crackings" were also seen. (H & E stain, X200)

Fig. 3
Diffuse and strong cytoplasmic expression of EMA. (EMA stain, X400)

From the Departments of Dermatology and Pathology,* the Hospital of China Medical University, Taichung
Accepted for publication: April 29, 2004
Reprint requests: Chao-Hong Liu, M.D., Department of Dermatology, the Hospital of China Medical University, Taichung, No. 2, Yuh-Der Rd., Taichung 404, Taiwan
TEL: 886-4-22052121 ext. 4430     FAX: 886-4-22064561
**DIAGNOSIS: Extraneural Perineurioma**

**DISCUSSION**

By knowledge to date, tumors of peripheral nerves are classified into schwannoma, neurofibroma, perineurioma, and malignant peripheral nerve sheath tumor (MPNST). There are two distinct forms of perineurioma: the intraneural perineurioma and the extraneural perineurioma. Both tumors are rare, and the incidence of extraneural perineurioma is less than intraneural perineurioma.

Lazarus and Thrombetta firstly presented perineurioma by its unique ultrastructure in 1978. Before their work, extraneural perineurioma had long been misdiagnosed as other tumor entity because it had no clear-cut clinical or histological criteria.

Extraneural perineurioma usually presents as a symptomless, solitary tumor ranges from 1 to 20 cm in diameter. It may be dermal, subcutaneous, or more deeply seated on the extremities. Other unusual location as the face and the kidney has also been reported.

Under light microscope, there are many different histological presentations, depending on the cellularity and the stromal variation, and different presentations may appear within a single tumor.

The most commonly seen histological pattern, as we presented, is composed of interwoven fascicles, loose whorls, and storiform arrangements. The constituent spindle cells resemble fibroblast, with more delicate, wave-like cytoplasmic processes that tangles. Intercellular "crack" is a commonly observed artifact. Small nucleoli are seen in elongated, curved, wrinkled, and flattened nuclei. In lesions with dense cellularity, small amount of stroma and storiform pattern are characterized and may be confusing in diagnosis. If the stroma is highly collagenized, the tumor cells usually dissect among and encircle collagen bundles, through the matrix. If the myxoid stroma predominates, the perineurial cells will be separated widely, showing the numerous delicate, interweaving cellular extensions more clearly.

The differential diagnosis of extraneural perineurioma includes dermatofibrosarcoma protuberans, fibrous histiocytoma, myxoid soft tissue tumors, giant cell of tendon sheath, neurilemmoma, neurofibroma, glomus tumor and meningo. The definite diagnosis of perineurioma depends on immunohistochemistry. Despite the variety in histological findings, the perineurial cells always show nonexpression of S-100 protein and expression of EMA. Ultrastructural study is not routinely performed, but if it is observed, the cells of perineurioma share the same features with normal perineurial cells with prominent pinocytic vesicles and scattered rudimentary basement membrane.

The treatment of choice of extraneural perineurioma is surgical resection. Our patient had no recurrence, and no neurocutaneous syndromes were noted thereafter.

**REFERENCES**