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Pathology Page

Cellular-type neurothekeoma over the chest wall of a woman

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A 33-year-old woman with no underlying systemic disease visited our hospital with a tender, hyperpigmented skin tumor over the right lateral chest wall for 6 months. Initially, the lesion was a small hemorrhagic cutaneous papule. Perifocal hyperpigmentation developed progressively and the lesion gradually enlarged to 2 cm at its greatest diameter. On physical examination, a painful ill-defined mass-like lesion was palpated over the right lateral chest wall. It was a 1×2 cm^2 ovoid hyperpigmented skin patch with a 0.3-cm papule in the center (Fig. 1A). A simple excision was performed. After the operation, the patient recovered uneventfully and has had no local recurrence. The histological examination revealed lobulated solid nests and strands of large epithelioid cells with vesicular nuclei, minimal nuclear atypia, pale cytoplasm, and rare mitoses, with infiltration into the dermis (Fig. 1B). Immunohistochemically, the tumor cells were positive for vimentin and CD57 and negative for CD34, epithelial membrane antigen (EMA), human melanoma black 45, estrogen receptor, S-100, and cytokeratin. These features were consistent with a cellular variant of neurothekeoma.

Neurothekeoma is a type of benign nerve sheath neoplasm and presumably arises from small cutaneous nerves [1,2]. Neurothekeomas have been classified as myxoid, cellular, or mixed type. However, myxoid-type neurothekeomas have a relatively high local recurrence rate when managed by simple local excision and appear to be unrelated to cellular and mixed-type neurothekeomas. Nowadays, the term "neurothekeoma" should probably be reserved for cellular or mixed-type tumors. The myxoid-type neurothekeomas are considered nerve sheath myxomas [3].

Neurothekeomas encompass a spectrum of diseases with an uncertain histogenesis. They occur in the dermis and are composed of spindled and epithelioid cells arranged in a fascicular or concentric whorl pattern. There is no well-defined encapsulation. Neurothekeomas histologically differ from nerve sheath myxomas by their higher cellularity and presence of larger spindled or epithelioid cells with vesicular nuclei. Immunohistochemically, the tumor cells may express collagen type IV, calponin, smooth muscle actin, Leu-7, and S-100 protein, but not nerve growth factor receptor p75, glial fibrillary acidic protein, or CD34 [2].

Cellular-type neurothekeoma is a recently characterized variant of neurothekeomas [4]. The typical histology of a cellular-type neurothekeoma shows a lobulated dermal tumor composed of spindled and epithelioid cells, arranged in fascicles and nests, which are usually NK1/C3 positive, but lack immunoreactivity for S-100 protein, indicating they are not of Schwann cell origin [1,4]. They are also negative for EMA, indicating they are not of perineural cell origin either [2]. Histologically, most cases are poorly marginated. These tumors characteristically have a lobulated or micronodular architecture and are composed of nests and bundles of epithelioid to spindled cells with pale eosinophilic cytoplasm, often separated by dense hyaline collagen [4]. Most tumors show mild cytologic atypia in the form of nuclear variability and small nucleoli undergoing occasional mitoses. In the largest case series, the mean mitotic rate was three per 10 high-power fields [4]. Other histological features might include myxoid stroma (29%), marked stromal hyalinization (4%), osteoclastic giant cells (15%), notably pleomorphic cells (25%), and active mitoses (21%, $\geq$5 per 10 high-power fields) [4].

Cellular-type neurothekeomas tend to affect women more than men, usually in the second and early third decades of life, with a mean age at presentation of 15–21 years [1,2,4]. Hornick and Fletcher [4] reported that the mean size of cellular-type neurothekeomas was 1.1 cm (range: 0.3–6 cm; 90% < 2 cm) [4]. These tumors arose most often on the upper limb (35%) and head and neck (33%) [4]. About half of the tumors were limited to the dermis, and half involved the superficial subcutaneous tissue [4]. Complete

Conflicts of interest: none.

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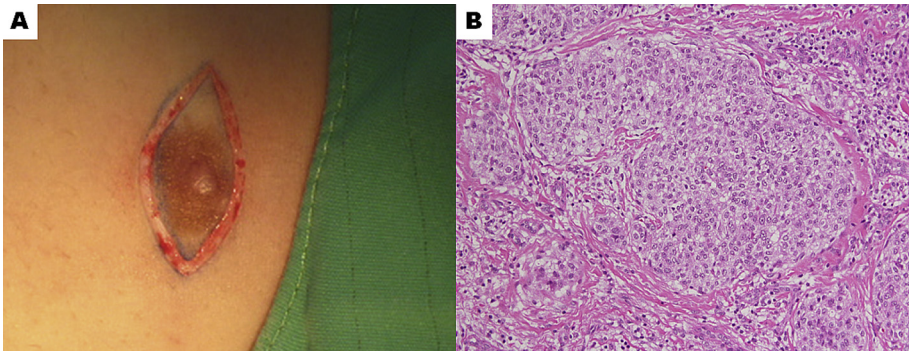

Fig. 1. (A) A 1 × 2 cm^2 ovoid hyperpigmented skin patch with a 0.3-cm papule in the center on the right lateral chest wall. (B) Histologically, there are lobulated solid nests and strands of large epithelioid cells with vesicular nuclei, minimal nuclear atypia, pale cytoplasm, and rare mitoses, with infiltration into the dermis (hematoxylin and eosin 200×).

surgical excision is the cornerstone of treatment. Recurrence is rare and only after incomplete excision [4].

Atypical features in cellular-type neurothekeomas have been reported, including large size (up to 6 cm), deep penetration (extending into skeletal muscle or subcutaneous fat, or both), diffusely infiltrative borders, vascular invasion, a high mitotic rate, and marked cytologic pleomorphism [1]. Although neurothekeomas with worrisome features raise concern about the biologic potential of these lesions, no recurrence has been reported after complete resection [1,4]. Atypical histologic features seem to have no clinical significance [4].

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