Multiple Cellular Neurothekeomas

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Multiple Cellular Neurothekeomas

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HISTORY OF PRESENT ILLNESS: A 69 year-old Caucasian female presents for evaluation of multiple lesions that have slowly developed on her bilateral dorsal hands and forearms for the last 30 years. The lesions are tender.

MEDICAL HISTORY/SURGICAL HISTORY: Hypertension, partial hysterectomy

FAMILY HISTORY: No family history of similar appearing lesions

MEDICATIONS: Metoprolol succinate ER, vitamin D 2000 units once daily, digoxin, topical conjugated estrogen, aspirin

CURRENT TREATMENT: Punch excisions

PHYSICAL EXAMINATION: Grouped 5-8 millimeter smooth, firm, pink, discrete papulonodules on bilateral dorsal hands and forearms. (Figures 1and 2)

BIOPSY: Advanced Dermatology Associates LTD and University of Washington (AD18-05392, 05/25/2018), Left lateral dorsal hand, left dorsal radial wrist, left lateral distal forearm, right ulnar wrist, right radial wrist: There are similar histologic features in all specimens which are notable for a fascicular, relatively uniform, proliferation of spindled and epithelioid cells with mild atypia in the dermis associated with a myxoid background. The proliferation is relatively well delineated. Immunohistochemical stains demonstrate the cells are negative for SMA. Mart-1, CK5/6, p63, CD31, HMB-45, CD2, EMA, S100, CD45RO, and CD34. CD163 and CD68 highlight background histiocytes. The cells are variably positive for Factor XIIIa, positive for NKI-C3, and weakly positive for NSE. PGP 9.5 is equivocal. A colloidal iron stain demonstrates moderate amount of dermal mucin deposition. (Figure 3)

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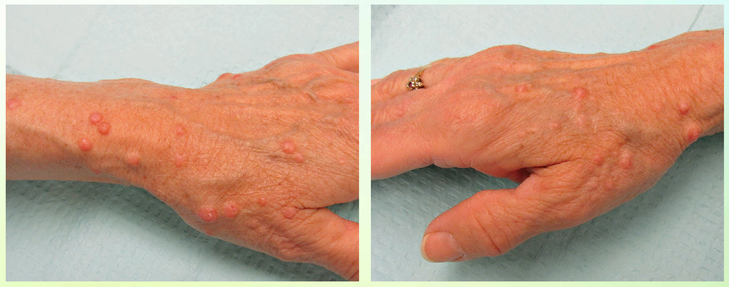
TABLE 1. IMMUNOHISTOCHEMICAL PROFILES OF MYXOID AND CELLULAR NEUROTHEKEOMAS.

Immunohistochemical staining pattern	Myxoid	Cellular
	S100 + Collagen type IV + Capsule EMA+	S100-Inconsistently stains for NKI/C3, PGP9.5, MITF, SMA, S100A6



Figures 1 and 2. Clinical appearance of multiple neurothekeomas.

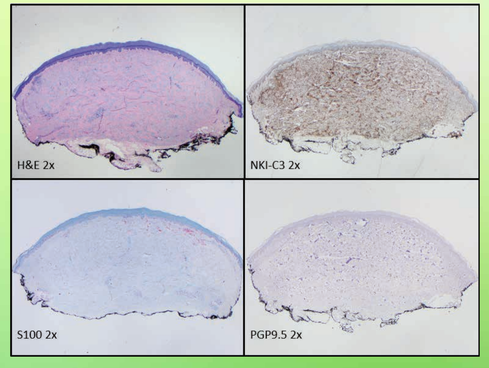


Figure 3. (H&E, 2x): A fascicular, relatively uniform, proliferation of spindled and epithelioid cells with mild atypia in the dermis associated with a myxoid background. (NKI-C3, 2x): Positive. (S100, 2x): Negative. (PGP9.5, 2x): Equivocal.

Neurothekeoma is a rare benign dermal neoplasm of debated lineage with consideration of fibrohistiocytic, neural, or smooth muscle derivation. Neurothekeomas most commonly occur in young females and develop as a solitary, slow growing, asymptomatic papulonodule on the head or neck. Multiple or agminated neurothekeomas are an unusual clinical presentation that have rarely been described.

Histopathologically, neurothekeomas are divided into three subtypes based on the degree of cellularity and mucinous stroma: myxoid, mixed, and cellular. All subtypes are poorly circumscribed plexiform dermal neoplasms that may extend into the subcutis. Myxoid neurothekeomas have abundant stromal mucin with few spindled and epithelioid cells in between fibrous septae. Cellular neurothekeomas are made up of fasciculated spindled and epithelioid cells in a stroma with minimal mucin. The immunohistochemical profile varies depending on the subtype (Table 1). Immunohistochemistry also helps exclude other histopathologic differential diagnoses including melanocytic, fibrohistiocytic, and Schwann cell tumors.

The varied histopathologic appearance and immunohistochemical staining patterns of neurothekeomas are hypothesized to be due to their existence on a morphologic spectrum directly related to nerve sheath maturation. Cellular neurothekeomas representing an undifferentiated variant and myxoid neurothekeomas representing a well differentiated variant. However, others believe that the varied immunohistochemical expression is secondary to their different cytomorphology.

Complete excision of neurothekeomas is both diagnostic and therapeutic. Although neurothekeomas may recur following incomplete removal, benignity is maintained and there are no reports of metastasis. Lastly, no known syndrome has been described in patients who have developed multiple neurothekeomas, but due to the rarity of this condition, it is suggested to follow these patients long term.