

Two Cases of Nerve Sheath Myxomas

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Nerve sheath myxoma(NSM) is a rare cutaneous neoplasm of uncertain histogenesis. NSM can be divided into two groups; NSM1(a myxoid NSM) and NSM2 (a cellular neurothekeoma). NSMs are characterized histologically by well-defined, lobular or plexiform dermal proliferation of stellate, spindle-shaped cells embedded in abundant myxoid stroma. A variable amount of nerve fascicles are entrapped within and around the tumor. Two variants of NSM share the same features to a certain degree but differ in cellularity and mucin deposition.

We report two cases of NSMs with different clinical and histopathologic features.
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Key Words : Nerve sheath myxoma, Cellular neurothekeoma

Nerve sheath myxoma (NSM) is a rare neoplasm of the dermis. NSMs can be divided into a small group, designated mature NSM (NSM1: myxoid NSM), and larger group (NSM2: cellular neurothekeoma) that includes both immature NSM (NSM2 A) and differentiating immature NSM (NSM2 B)¹(Fig. 1). In NSM1s myxoid micronodules that vary in size are loosely crusted in a fibrous matrix in the reticular dermis to form a multilobulated mass. In NSM2 A, fascicles of cells are arranged in dissecting patterns among the collagen bundles of the reticular dermis. In NSM2 B, some of the aggregates become locally expended to form broad fascicles and nodules in which both cellular and myxoid patterns are represented¹.

We report two cases of NSMs which could be classified into NSM1 and NSM2B.

CASE REPORT

CASE 1

A 25-year-old woman presented with a slightly tender, firm, skin-colored, 0.5×0.5cm-sized papule on the left hand (Fig. 2). She had had this mass for two months without change. The clinical diagnosis was dermatofibroma. The lesion was completely removed and submitted for microscopic examination.

Histopathologically, there were no changes in the epidermis and papillary dermis. The tumor was situated entirely in the dermis, and was well-defined with no membrane or capsule. It was divided into multiple distinct cellular lobules by thickened fibrous connective tissue (Fig. 3). Each lobule consisted of a few cells within an abundant mucinous background. The cells were spindle to polygonal in shape showing frequent intranuclear inclusions and eosinophilic plump cytoplasm. Mitotic figures were not seen. The myxoid stroma within the lobules stained positive with mucin stain. Immunohistochemically, the tumor cells were strongly positive for S-100 protein. A diagnosis of NSM1 was made based on these findings.

CASE 2

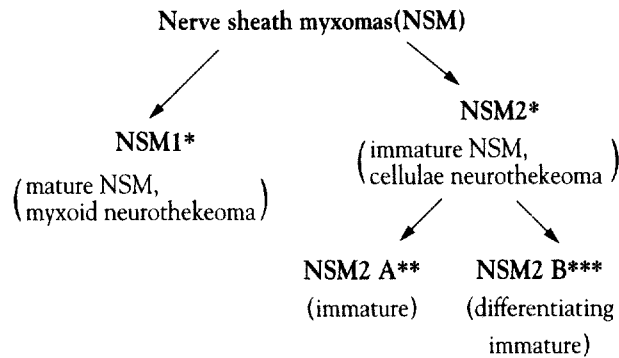
A 20-year-old man presented with slightly tender, skin-colored, 0.8 x 0.9 cm-sized papule on the side of

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- * myxoid and loosely cellular lesion that mostly appears micronodular
- ** cellular, or epithelioid and fascicular
- *** mixed mature (myxoid) and immature (epithelioid) patterns

Fig. 1. Schematic diagram of NSM classification.

Fig. 2. A 25-year-old woman presented with a slightly tender, firm, skin-colored, 0.5×0.5cm-sized papule on the left hand.

Fig. 3. Multiple distinct cellular lobules were divided by thickened fibrous connective tissue (H&E stain, ×40).

Fig. 4. A 20-year-old man represented with slightly tender, skin-colored, 0.8×0.9 cm-sized papule on the dorsum of the nose.

Fig. 5. Fascicles of cells are arranged infiltrating patterns among collagen bundles of reticular dermis (H&E stain, ×40).

Fig. 6. In areas showing the most abundant matrix, some of the cells are arranged in whorls(H&E stain, ×400).

nasal bridge (Fig. 4). Past and family histories were non-contributory. He noticed the papule one month before. Since then the lesion had slowly enlarged. Clinically, dermoid cyst, nasal glioma, pilomatricoma, and NSM were included in the different diagnosis. The whole mass was surgically removed.

Histopathologically, both cellular and myxoid areas coexisted. Some fascicles of cells had infiltrated among the collagen bundles of the reticular dermis (Fig. 5). In other areas, the cells were loosely spaced in a myxoid matrix (Fig. 6). Some tumor cells weakly stained for S-100 protein. They were positive for vimentin and negative for cytokeratin, desmin, actin, NSE. A diagnosis of NSM2 B was made.

DISCUSSION

NSM was originally defined by Harkin and Reed and was reintroduced by Gallager and Helwig under the designation of neurothekeoma. Various terms have been used for this tumor such as pacinian neurofibroma, bizarre cutaneous neurofibroma, cutaneous lobular neuromyxoma, and perineurial myxoma¹.

Clinically, a NSM1 is most common in middle-aged adults with a male-female ration of approximately 1:2. Asymptomatic, soft, skin-colored papules or nodules ranging from 0.5 to 1.0 cm in diameter are typically located on the face and the upper extremities, but can occur anywhere on the body¹. A NSM2 differs from the myxoid variant due to its earlier onset. The head is the favorite site. The lesions are firm, pink, red-brown papules or nodules, and some may produce symptoms³.

The histogenesis of NSM is still controversial. Ultrastructural studies support the view that NSM1 is primarily composed of Schwann cells and perineural differentiation and NSM2 is composed of undifferentiated cells with partial features of Schwann cells, smooth muscle cells, myofibroblasts, and fibroblasts. The similarities of cells seen in

the NSM1 and 2, along with the microscopic and immunohistochemical results, support the concept that NSM2 belongs within the spectrum of NSM, despite the fact that neural features are not fully expressed⁴.

Immunohistochemically, the S-100-positivity in the NSM1 and not in the NSM2 may be related to the cell origins. It may be that Schwann cells and perineural cells represent different stages of differentiation, both being derived from common precursor cells that may have transitional features and give rise to the NSM2 that does not express S-100 protein⁵. However the tumor of our second case stained weakly positive for S-100 protein. Its overlapped microscopic features and reactivity to antibody for S-100 protein may support the view that NSM1 and NSM2 can be regarded as the same spectrum.

NSM remains a puzzling entity because of the wide spectrum of its clinical, histogenetical, and immunohistochemical features.

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