NERVE SHEATH MYXOMA WITH BIDIRECTIONAL SCHWANNOMATOUS AND PERINEURAL DIFFERENTIATION

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Summary

A case of nerve sheath myxoma occurring in occipital region in 70-yr-old woman is presented. The tumor showed typical lobular and myxoid morphology. Immunohistochemically, it showed unusual coexpression of Schwann cell markers S100 protein and GFAP with perineural cell markers EMA and claudin-1. CD34+ fibroblast-like cells were scarce, and nerve axons were not found in the tumor. Clinical pathology and histogenesis of the lesion are discussed.

Key words: nerve sheath myxoma – perineurioma – schwannoma – epithelial membrane antigen – S100 protein

Súhrn

Myxóm nervového púzdra (nerve sheath myxoma) so schwannómovou a perineurálnou diferenciáciou

Prezentovaný je prípad myxómu nervového púzdra u 70-ročnej pacientky v okcipitálnej oblasti. Tumor mal typickú lobulárnu a myxoidnú morfológiu tejto zriedkavej jednotky. Neobvyklá bola difúzna koexpresia markerov Schwannových buniek S100 proteinu a GFAP a perineurálnych markerov EMA a claudin-1. CD34+ fibroblastické bunky boli málo početné a nervové axóny neboli v tumore nájdené. Diskutovaná je klinická patológia a histogenéza lézie.

Kľúčové slová: myxóm nervového púzdra – perineurióm – schwannóm – epitelový membránový antigén – S100 protein

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Nerve sheath myxoma (NSM) (6, 9) is a rare cutaneous/subcutaneous tumor that occurs mostly in middle aged adults on the extremities. Since its original description by Harkin and Reed (9), the lesion was reported under various names, including neurotheceoma (2, 7, 18, 24), cutaneous lobular neuromyxoma (10), myxomatous perineurioma (2, 22), bizarre cutaneous neurofibroma (14), myxoma of nerve sheath (2, 9), and dermal nerve sheath myxoma (19). This variability in nomenclature reflects well the doubts on histogenesis of the tumor. NSM is typically S100 protein positive whereas perineural cell marker EMA is absent or it stains only rare cells. Therefore, most of authors favor close relationship to schwannoma or to neurofibroma (2, 6, 9, 10, 14, 17-19, 21-23). We present an additional case of morphologically typical nerve sheath myxoma that shows, however, an unusual coexpression of Schwann cell and perineural markers. The case indicates that nerve sheath myxoma can posses bidirectional schwannomatous-perineural differentiation.

MATERIALS AND METHODS

The tissue of the excised tumor was fixed in 4% formalin and processed routinely. The sections were stained with hematoxylin and eosin and Bodian stain for nerve axons. Primary antibodies used for immunohistochemistry are listed in **Table 1**. Immunostaining was performed according to standard protocols using avidin-biotin complex labeled with peroxidase or alkaline phosphatase**.** Microwave antigen pretreatment was performed prior to applying the primary antibodies. Appropriate positive and negative controls were applied.

Table 1. Antibodies used in the study

Antibody	Clone	Dilution	Source
ASMA	1A4	1:1000	DakoCytomation
CD34	Qbend10	1:800	Novocastra Lab.
claudin-1	poly	1:50	Zymed
cytokeratins	AE1-AE3	1:200	Boehringer
desmin	D33	1:3000	DakoCytomation
EMA	E29	1:700	DakoCytomation
GFAP	poly	1:3000	DakoCytomation
NFP	2F11	1:1000	DakoCytomation
S100 protein	S1/61/69	1:50	Novocastra Lab

ASMA – alpha smooth muscle actin, EMA – epithelial membrane antigen, GFAP – glial fibrillary acidic protein, NFP – neurofilament protein, poly – polyclonal