Mediastinal ganglioneuroma with perineural cell differentiation. Report of a case

Zámečník M.[1], Chlumská A.[2,3], Ondriaš F.[1]

[1] Alpha Medical Pathology, s.r.o., Bratislava, Slovak Republic
[2] Šikl`s Department of Pathology, Faculty Hospital, Charles University, Pilsen, Czech Republic
[3] Laboratory of Surgical Pathology, Pilsen, Czech Republic

SUMMARY
An unusual case of ganglioneuroma with perineural cell differentiation is presented. The tumor was removed from the mediastinum in a 34-year-old male patient. Histologically, it contained neuroid bundles of bland spindle cells, scattered ganglion cells, and some foci of adipocytic metaplasia. Immunohistochemically, the tumor showed expected expressions of S100 protein, neurofilament protein and calretinin. In addition, many spindle cells were positive for perineural cell markers EMA, claudin-1, and GLUT-1. These cells were often arranged in an organoid fashion around the schwannoid bundles. This case indicates that the cells of ganglioneuroma can mature simultaneously towards both Schwann cell and perineural cell phenotypes.

Keywords: ganglioneuroma – perineurioma – EMA – claudin-1 – GLUT-1

Ganglioneuróm s perineurálnou diferenciáciou. Kazuistika

SÚHRN
Prezentovaný je prípad ganglioneurómu s neobvyklou perineurálnou diferenciáciou. Jednalo sa o tumor mediastína u 34-ročného muža. Histologicky obsahoval neuroidné zväzky blandných vretenovitých buniek, zrelé gangliové bunky a ložiskovú adipocytárnu metapláziu. Imunohistochemicky vykazoval tumor očakávané expresie S100 proteinu, kalretinínu a neurofilament proteinu. Prekvapujúcou bola pozitivita početných buniek na perineurálne markery EMA, klaudín-1 a GLUT-1. Jednalo sa často o bunky v organoidnom usporiadaní okolo S100-pozitívnych schwannoidných zväzkov. Prezentovaný prípad ukazuje, že elementy ganglioneurómu sa môžu diferencovať do fenotypu bunky Schwannovej i perineurálnej.

Kľúčové slová: ganglioneuróm – perineurióm – EMA – claudin-1 – GLUT-1

Cesk Patol 2012; 48(2): 94–96

Ganglioneuroma is a benign neoplasm composed of ganglion cells and neuroid spindle cells, occurring usually in adults, and most often located in the posterior mediastinum and retroperitoneum (1). It arises through maturation of neuroblastoma (2,3) or de novo (1). Ultrastructurally, the spindle cell component contains mostly Schwann cells (1). Rare ultrastructural studies have found, in addition to a Schwann cell population, some cells with the features of the perineural cell type (4,5). However, immunohistochemical expression of perineural cell markers such as epithelial membrane antigen (EMA), claudin-1 and GLUT-1 has not yet been described in this type of tumor to our knowledge and according to our literature search. Here, we would like to present a case of ganglioneuroma, in which a well-developed perineural cell component with the expression of perineural cell markers EMA, claudin-1 and GLUT-1 was found (6–8).

✉ ***Correspondence address:***
M. Zamecnik, MD
Medicyt, s.r.o., lab. Trencin
Legionarska 28, 91171 Trencin, Slovak Republic
tel: +421-907-156629
e-mail: zamecnikm@seznam.cz

MATERIAL AND METHODS

The tissue of the tumor was fixed in 10% formalin and processed routinely. The sections were stained with hematoxylin and eosin. For immunohistochemistry, the following primary antibodies were used: S100 (polyclonal, 1:400), alpha-smooth muscle actin (clone 1A4, 1:1000), desmin (clone D33, 1:3000), neurofilament protein (clone 2F11, 1:1000), GLUT-1 (polyclonal, 1:200), GFAP (polyclonal, 1:3000), EMA (clone E29, 1:700) (all from DAKO, Glostrup, Denmark), calretinin (clone 5A5, 1:100, Novocastra Lab., Newcastle upon Tyne, UK), claudin-1 (polyclonal, 1:50, Zymed, San Francisco, USA), CD34 (clone Qbend/10, 1:800, Novocastra Lab., Newcastle upon Tyne, UK).

Immunostaining was performed according to standard protocols using an avidin-biotin complex labeled with peroxidase or alkaline phosphatase. Appropriate positive and negative controls were applied.

CASE REPORT

In a 34-year-old male patient, a tumor of the posterior mediastinum detected on CT was removed surgically.

Grossly, the tumor tissue was obtained in four fragments which measured together 7.5x5x4cm. The fragments from the marginal