Perineural cell differentiation in ganglioneuromas. Report of 8 cases with immunohistochemical expression of perineural cell markers

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SUMMARY
Eight cases of ganglioneuroma were examined for a presence of perineural cell differentiation, using the immunohistochemical markers epithelial membrane antigen (EMA), claudin-1 and GLUT-1. The mean age of the patients was 42.3 years (range 26–68 years), six patients were females and two were males. Five tumors were located in the adrenal gland and 3 tumors in the retroperitoneum. Morphology of the tumors was typical, i.e., they were composed of neuroid spindle cell population and scattered mature appearing ganglion cells. Spindle cells positive for perineural cell markers claudin-1 and GLUT-1 were found in all lesions, at least focally. EMA+ cells were seen in 2 of 8 tumors. These perineural-type cells were often arranged in organoid fashion around the schwannoid bundles or around the vessels. Our findings indicate that perineural cell differentiation is commonly present in ganglioneuromas.

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

Perineurálna diferenciácia v ganglioneurómoch. Súbor 8 prípadov s imunohistochemickou expresiou perineurálnych markerov

SÚHRN
V súbore 8 ganglioneurómov bola zisťovaná prítomnosť perineurálnej diferenciácie pomocou 3 imunohistochemických perineurálnych markerov: epitelového membránového antigénu (EMA), klaudínu-1 a GLUT-1. Priemerný vek pacientov bol 42,3 rokov (rozsah 26–68 rokov), 6 tumorov bolo u žien a 2 prípady u mužov. Päť tumorov bolo v nadobličke a 3 v retroperitoneu. Morfológia tumorov bola typická, t.j. tvorená neuroidnou vretenobunkovou populáciou a zrelými gangliovými bunkami. Vretenovité bunky pozitívne na perineurálne markery klaudín-1 a GLUT-1 boli nájdené vo všetkých tumoroch. EMA-pozitívne bunky boli v 2 léziach. Bunky s perineurálnou diferenciáciou boli často usporiadané organoidne v periférii neuroidných fasciklov alebo okolo ciev. Náš nález ukazuje, že perineurálna diferenciácia je bežne prítomná v ganglioneurómoch.

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

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Ganglioneuroma is a benign tumor that occurs usually in adults, and it is most often located in the posterior mediastinum and retroperitoneum (1). It is composed of ganglion cells and neuroid spindle cells. Ultrastructurally, the spindle cell component contains mostly Schwann cells (1). Rare ultrastructural studies found, in addition to Schwann cell population, some cells with perineural-type features (2,3). Recently, we reported a case of ganglioneuroma with perineural cell differentiation proven with the immunohistochemical markers epithelial membrane antigen (EMA), claudin-1 and GLUT-1 (4). After seeing that case, we speculated that perineural cell differentiation can be more frequent in ganglioneuromas, and this prompted us to collect and examine additional cases. Now, we would like to present our series of 8 ganglioneuromas. Examining these tumors, we have found that perineural cell differentiation appears to be a common feature of ganglioneuroma, and that it can be demonstrated by currently used perineural cell markers – EMA, claudin-1 and GLUT-1 (5–7).

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MATERIAL AND METHODS

Eight cases with typical morphological and immunohistochemical features of ganglioneuroma were retrieved from our routine files. In all cases, the tumor tissue was fixed in 10% formalin and processed routinely. The sections were stained with hematoxylin and eosin. In all cases, available immunohistochemical slides showed typical immunophenotype of ganglioneuroma. The spindle cell component expressed the S100 protein, and the ganglion cells were positive for neurofilament protein and/or calretinin.

In all cases, we performed immunohistochemical examination for perineural cell markers on selected tissue blocks. The following primary antibodies were used: GLUT-1 (polyclonal, 1:200), EMA (clone E29, 1:700) (both from DAKO, Glostrup, Denmark), and claudin-1 (polyclonal, 1:50, Zymed). Immunostaining was performed according to standard protocols using avidin-biotin complex labeled with peroxidase or alkaline phosphatase. Appropriate positive and negative controls were applied.