

# Angiofibroma-like perineurioma. Report of a case

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## SUMMARY

We report an unusual perineurioma with numerous vessels, showing a strong similarity with angiofibroma. A 2,5 x 2 x 2 cm subcutaneous/dermal tumor occurred in 58-ys-old male in the left brachial region. Histologically, it was composed of haphazardly arranged bland spindle cells and it contained prominent vasculature. In rare foci, the tumor cells showed thin bipolar processes and an onion-like perivascular whorling pattern. Immunohistochemically, expression of perineural cell markers EMA, claudin-1 and CD34 was limited to perivascular foci and to rare cells among the vessels. In addition, the tumor expressed CD10 diffusely. Our finding indicates that diagnosis of perineurioma should be considered also by tumors with an "angiofibromatous" morphology. Especially soft tissue angiofibroma, which often express EMA (perineural cell marker), shows a strong resemblance to angiofibroma-like perineurioma.

**Keywords:** perineurioma – angiofibroma – soft tissue – EMA – claudin-1

## Perineurióm podobný angiofibrómu. Kazuistika

### SÚHRN

Prezentujeme prípad perineuriómu s neobvykle bohatou vaskularizáciou, ktorá spôsobila podobnosť s angiofibrómom. Šlo o dermálny/subkutánný tumor ľavého ramena u 58-ročného muža. Tumor mal rozmery 2,5 x 2 x 2 cm. Histologicky bol tvorený blandnými fibroblastoidnými bunkami usporiadanými zväčša nepravidelne, len s ojedinelým špirálovitým radením okolo ciev. Malé množstvo buniek malo bipolárne tenké výbežky. Cievky predstavovali druhú prominentnú zložku lézie. Imunohistochemicky bola zistená expresia perineurálnych markerov (EMA, kladín-1 a CD34) v perivaskulárnych bunkách a v malej časti buniek vzdialených od ciev. Tumor bol ďalej difúzne pozitívny na CD10. Náš nález ukazuje, že diagnózu perineuriómu je potrebné zvažovať aj pri bohato vaskularizovaných léziách s „angiofibroma-like“ morfológiou. Demonštrovaný prípad perineuriómu sa veľmi podobal na recentne popísaný angiofibróm mäkkých tkanív (u ktorého bola často pozorovaná aj expresia EMA).

**Kľúčové slová:** perineurióm – angiofibróm – mäkké tkanivá – EMA – kladín-1

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Soft tissue perineurioma is a benign tumor composed of perineural fibroblasts (1). It occurs most often in adult patients, in superficial soft tissue of extremities and trunk. The cells of perineurioma are arranged in a storiform, whorled or short fascicular pattern (1–3). Prominent vascularization does not belong to previously described morphology of perineurioma. Recently, we have seen a case of perineurioma which contained numerous vessels, and which therefore showed a strong similarity to angiofibroma.

## MATERIAL AND METHODS

The tumor tissue was fixed in Bouin solution, subsequently in 10% formalin, and then it was 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), GLUT-1 (polyclonal, 1:200), EMA (clone E29,

1:700), CD35 (Ber-MAC-DRC, 1:50) (all from DAKO, Glostrup, Denmark), claudin-1 (polyclonal, 1:50, Zymed, San Francisco, USA), CD34 (clone Qbend/10, 1:800), CD10(clone 56C6, 1:50) (both from Novocastra Lab., Newcastle upon Tyne, UK), pancytokeratin (AE1/AE3/PCK26, prediluted), vimentin (V9, prediluted), CD99 (O13, prediluted), CD21 (2G9, prediluted) (all four from Ventana, Illkirch, France). Immunostaining was performed according to standard protocols using avidin-biotin complex labeled with peroxidase or alkaline phosphatase. Microwave antigen pretreatment was used for immunoreactions with claudin-1 and CD10. Appropriate positive and negative controls were applied.

## CASE REPORT

In 58-year-old patient, the subcutaneous/dermal tumor grew slowly for 5 years. Recently, a small superficial ulceration developed on the surface. The tumor was excised and submitted for examination. **Grossly**, the 2,5 x 2 x 2 cm dermal/subcutaneous nodule was unencapsulated, and of rubbery consistency. Its cut surface was of a homogeneous glistening appearance. **Histologically**, the tumor contained bland appearing spindle cells in fibromyxoid stroma and prominent vasculature. The spindle cells were usually arranged haphazardly or in ill-defined fascicles (Fig. 1A). In rare perivascular areas, they created vague perivascular whorls (Fig. 1B). In these foci, the cells showed thin bipolar processes, whereas in

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