SD – IAP č. 764

LEKÁRSKA FAKULTA
Univerzita Komenského
v Bratislave

LUCIA KRIVOŠÍKOVÁ

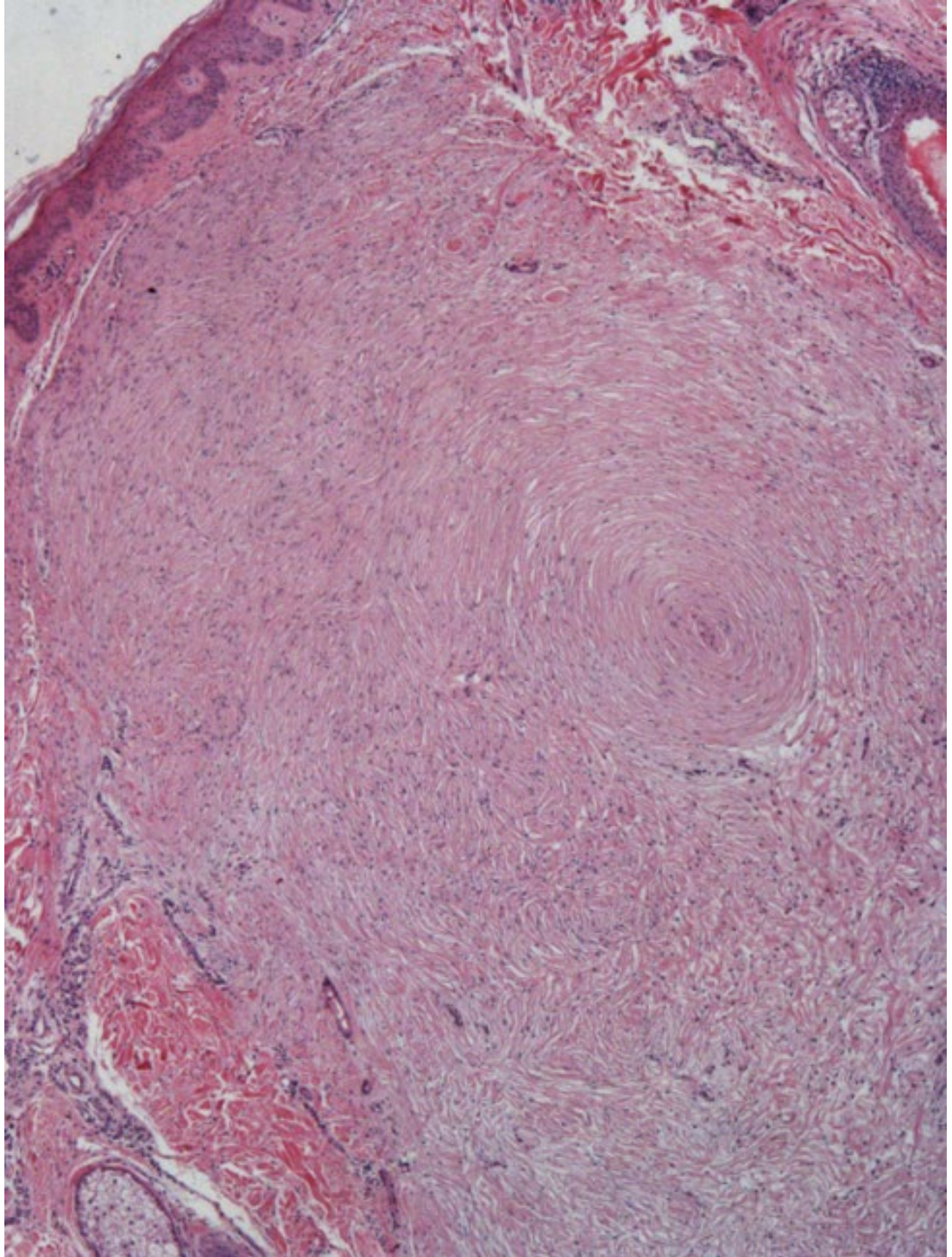
ÚSTAV PATOLOGICKEJ ANATÓMIE

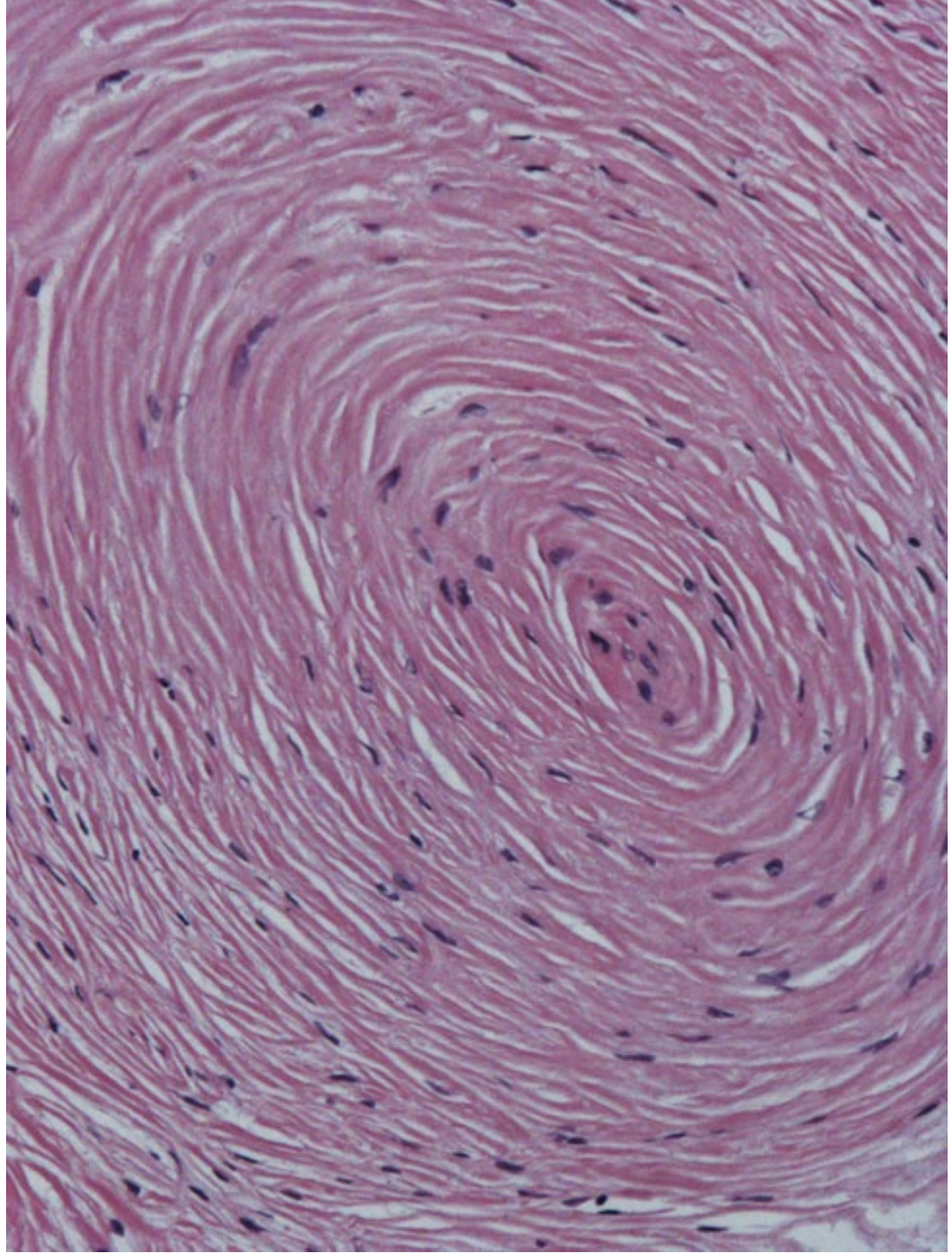
Klinické údaje

- 34 – ročný muž
- Fibróm kapilícia

Makroskopický nález

- Excízia kože na povrchu bez makroskopicky viditeľných zmien.
- Na reze v derme prítomný sivobelavý nodulárny útvar tuhoelastickej konzistencie priemeru 7mm.





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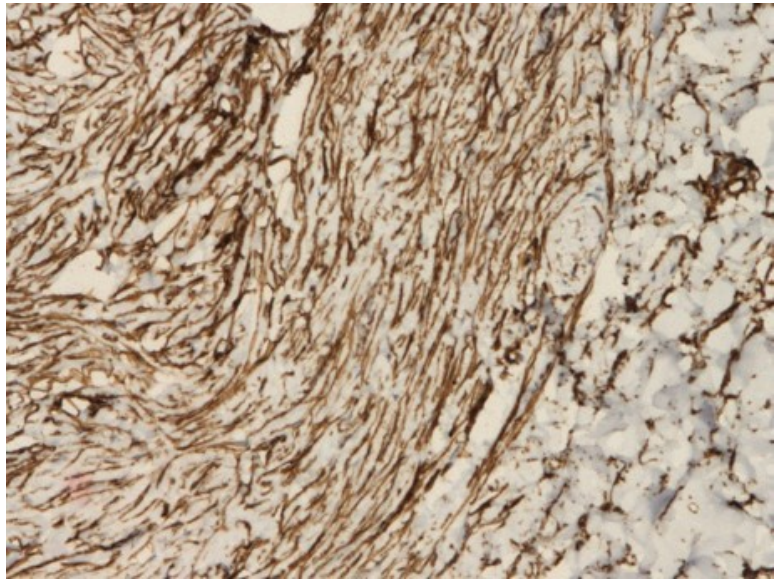
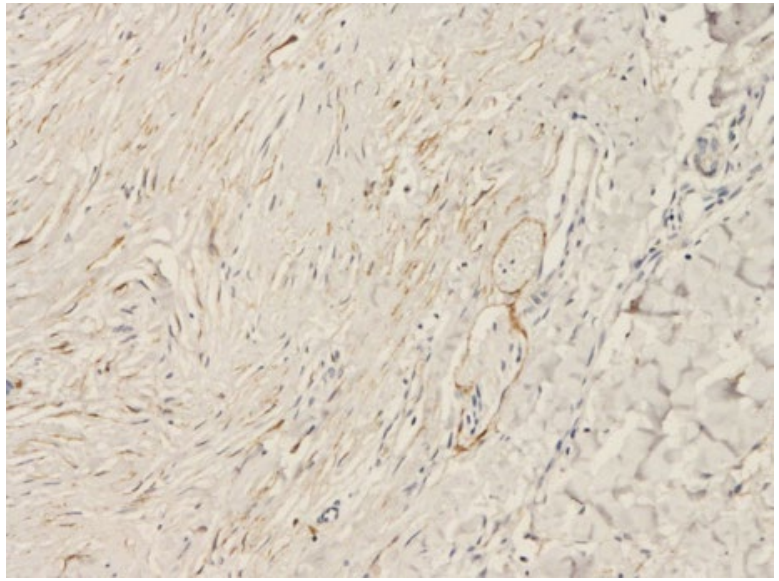
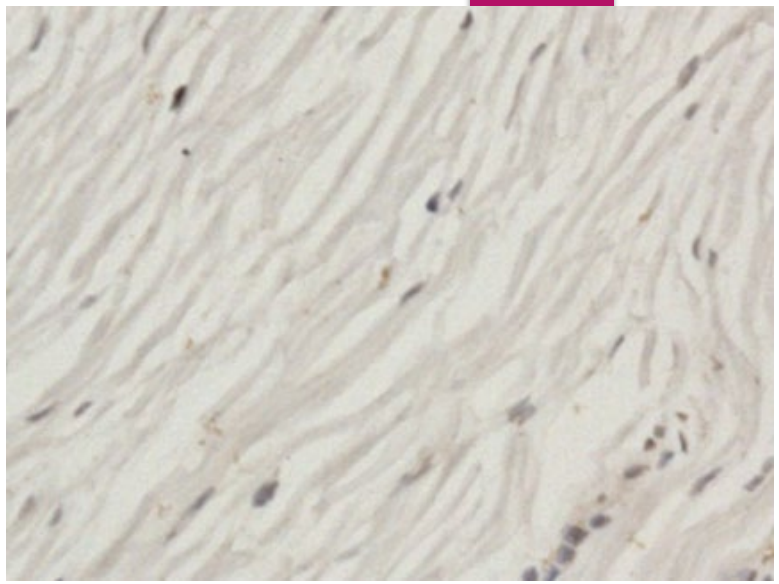


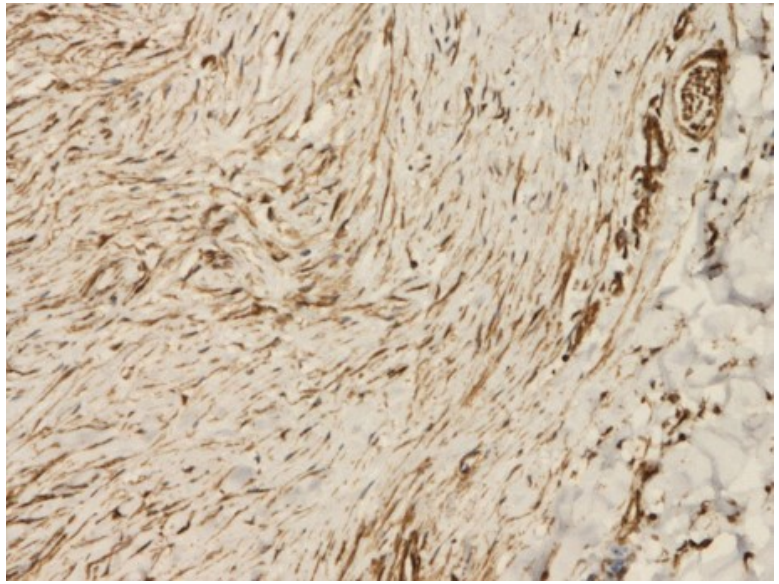
vimentin



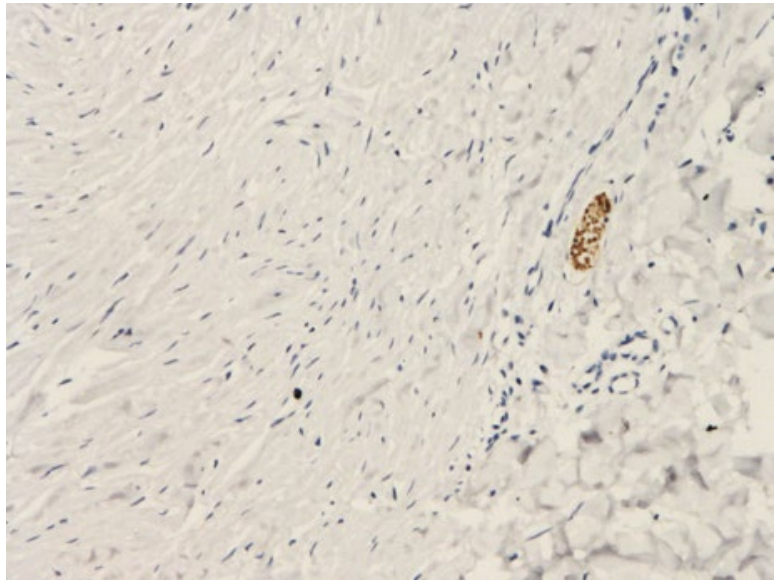
EMA



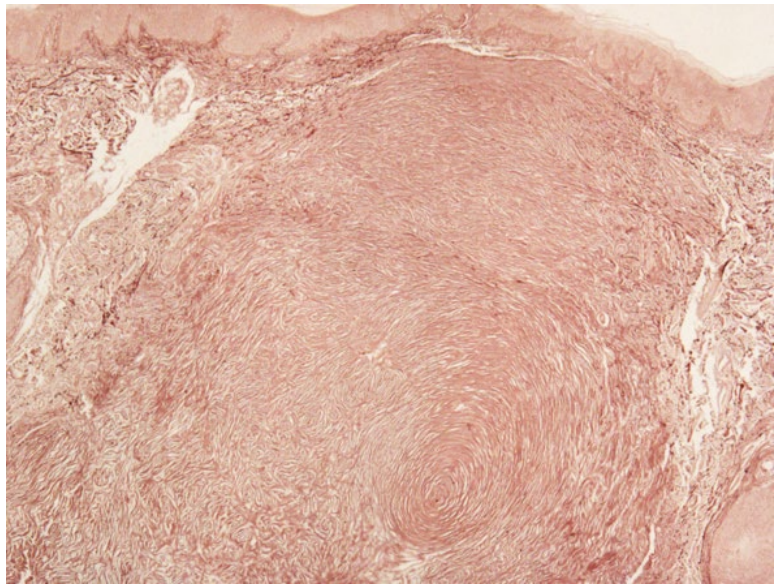
GLUT-1



CD34



S100



Orcein

IHC	výsledok
Vimentín	+++
CD34	++
EMA	+
S100	-
NSE	-
CD68	-
GLUT-1	-
AE1/3	-

Pacinian collagenoma

Morfologický variant sklerotizujúceho fibrómu (storiformného kolagenómu)

British Journal of Dermatology 1999; **141:** 119–122.

Pacinian collagenoma

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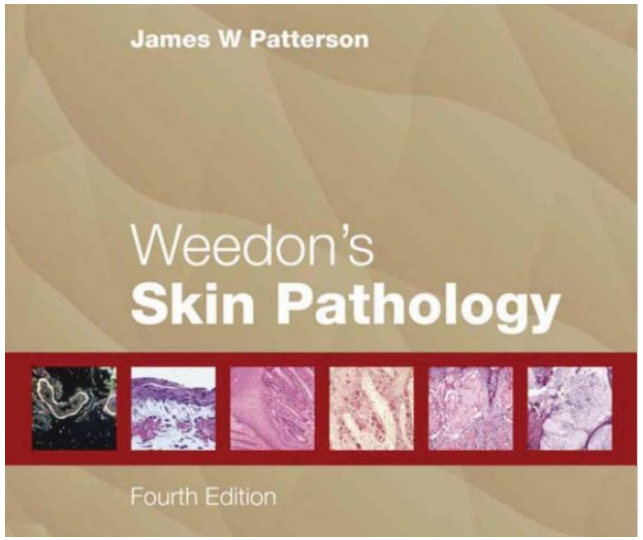
Accepted for publication 18 February 1999

Summary An unusual histological variant of collagenoma is described. A 36-year-old woman presented with a lump in the left hypothenar eminence. Histological examination revealed a well-delineated lesion composed of paucicellular collagen fibres arranged in concentric lamellations giving rise to an onion skin appearance. The overlying epidermis was thin and the lateral borders were demarcated by an epidermal collarette. Inflammation and xanthoma cells were absent and occasional capillaries were present. The lesion was positive for collagen stains, reticulin and CD34. This lesion represents an uncommon histological form of collagenoma or fibroma. It can be distinguished from histological look-alikes on the basis of the characteristic morphology and immunophenotype.

Key words: collagenoma, fibroma, fibrous tumour, pacinian



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CASE REPORT

Pacinian collagenoma: A distinct form of sclerotic fibroma

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Abstract

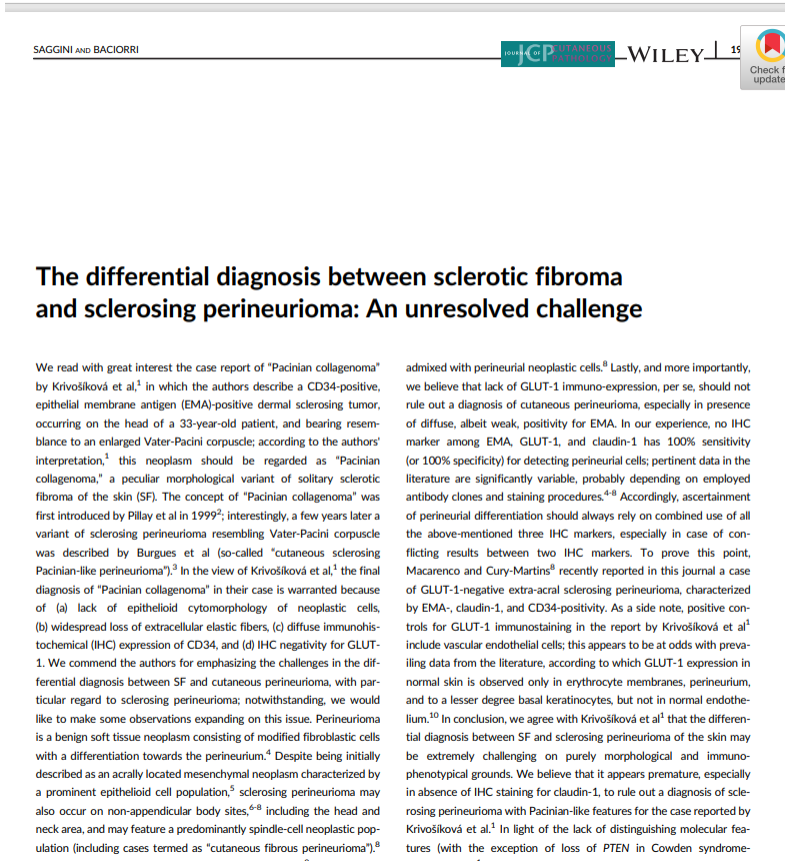
Sclerotic fibroma (storiform collagenoma) is a rare benign skin tumor. A solitary tumor, as well as multifocal lesions, are found either sporadically, or associated with Cowden syndrome. The tumor usually presents as clinically asymptomatic, slowly growing papule or nodule on the skin of the head, neck, and upper extremities. Microscopically the lesion is sharply demarcated, composed of hyalinized bands of collagen with low cellularity and a distinctive irregularly whorled or storiform pattern. We describe a case of a unique variant of this tumor in the scalp of a 33-year-old male. The tumor was microscopically composed of concentrically arranged collagen bundles with prevailing type III collagen, which resembled an enlarged Vater-Pacini corpuscle, with low density of CD34-positive and glucose transporter 1-negative spindle shaped cells. The specific microscopic appearance is suggestive of the term "Pacinian collagenoma" for this unique benign tumor.

KEYWORDS
CD34, GLUT-1, Pacinian collagenoma, sclerotic fibroma, type III collagen

Diferenciálna diagnóza

- Perineurióm (sklerotizujúci a fibrotizujúci variant)
- Paciniánsky neurofibróm
- Dermatofibróm

  - Potenciálny význam nálezu – asociácia sklerotizujúceho fribómu s Cowdenovým syndrómom.



The differential diagnosis between sclerotic fibroma and sclerosing perineurioma: An unresolved challenge

We read with great interest the case report of "Pacinian collagenoma" by Krivošíková et al,[1] in which the authors describe a CD34-positive, epithelial membrane antigen (EMA)-positive dermal sclerosing tumor, occurring on the head of a 33-year-old patient, and bearing resemblance to an enlarged Vater-Pacini corpuscle; according to the authors' interpretation,[1] this neoplasm should be regarded as "Pacinian collagenoma," a peculiar morphological variant of solitary sclerotic fibroma of the skin (SF). The concept of "Pacinian collagenoma" was first introduced by Pillay et al in 1999[2]; interestingly, a few years later a variant of sclerosing perineurioma resembling Vater-Pacini corpuscle was described by Burgues et al (so-called "cutaneous sclerosing Pacinian-like perineurioma").[3] In the view of Krivošíková et al,[1] the final diagnosis of "Pacinian collagenoma" in their case is warranted because of (a) lack of epithelioid cytomorphology of neoplastic cells, (b) widespread loss of extracellular elastic fibers, (c) diffuse immunohistochemical (IHC) expression of CD34, and (d) IHC negativity for GLUT-1. We commend the authors for emphasizing the challenges in the differential diagnosis between SF and cutaneous perineurioma, with particular regard to sclerosing perineurioma; notwithstanding, we would like to make some observations expanding on this issue. Perineurioma is a benign soft tissue neoplasm consisting of modified fibroblastic cells with a differentiation towards the perineurium.[4] Despite being initially described as an acrally located mesenchymal neoplasm characterized by a prominent epithelioid cell population,[5] sclerosing perineurioma may also occur on non-appendicular body sites,[6-8] including the head and neck area, and may feature a predominantly spindle-cell neoplastic population (including cases termed as "cutaneous fibrous perineurioma"),[8] admixed with perineurial neoplastic cells.[8] Lastly, and more importantly, we believe that lack of GLUT-1 immuno-expression, per se, should not rule out a diagnosis of cutaneous perineurioma, especially in presence of diffuse, albeit weak, positivity for EMA. In our experience, no IHC marker among EMA, GLUT-1, and claudin-1 has 100% sensitivity (or 100% specificity) for detecting perineurial cells; pertinent data in the literature are significantly variable, probably depending on employed antibody clones and staining procedures.[4-8] Accordingly, ascertainment of perineurial differentiation should always rely on combined use of all the above-mentioned three IHC markers, especially in case of conflicting results between two IHC markers. To prove this point, Macarenco and Cury-Martins[8] recently reported in this journal a case of GLUT-1-negative extra-acral sclerosing perineurioma, characterized by EMA-, claudin-1, and CD34-positivity. As a side note, positive controls for GLUT-1 immunostaining in the report by Krivošíková et al[1] include vascular endothelial cells; this appears to be at odds with prevailing data from the literature, according to which GLUT-1 expression in normal skin is observed only in erythrocyte membranes, perineurium, and to a lesser degree basal keratinocytes, but not in normal endothelium.[10] In conclusion, we agree with Krivošíková et al[1] that the differential diagnosis between SF and sclerosing perineurioma of the skin may be extremely challenging on purely morphological and immunophenotypical grounds. We believe that it appears premature, especially in absence of IHC staining for claudin-1, to rule out a diagnosis of sclerosing perineurioma with Pacinian-like features for the case reported by Krivošíková et al.[1] In light of the lack of distinguishing molecular features (with the exception of loss of *PTEN* in Cowden syndrome-

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