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CASE REPORT

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# 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

## 1 | INTRODUCTION

Sclerotic fibroma (storiform collagenoma, SF) is a rare benign skin tumor. A solitary tumor, as well as multifocal lesions, are found sporadically,[1,2] or associated with Cowden syndrome,[3,4] for which multiple hamartomas, increased risk of developing some malignant tumors and loss of phosphatase and tensin homolog are exemplary.[5] According to the WHO classification of skin tumors from 2018,[6] SF clinically presents as asymptomatic, slowly growing white to skin-colored papule or nodule. Microscopically, SF is a circumscribed lesion composed of hyalinized bands of collagen with low cellularity with distinctive irregularly whorled or storiform pattern. It is uncommon to find a structure composed of concentric lamellae in fibromas.[7] We describe a case of this unique morphologic variant of SF resembling an enlarged Vater-Pacini corpuscle in the scalp of a 33-year-old male.

## 2 | CASE REPORT

A 33-year-old-male with no significant clinical history presented with an asymptomatic solitary subcutaneous nodule on the scalp. The nodule was of elastic consistency; the adjacent skin was without any pathological changes. The patient had no history of Cowden syndrome. An excisional biopsy was performed.

Grossly, there was a white nodular thickening of the dermis measuring 7 mm on the cut surface, with no visible changes on the skin surface. The sample was fixed in formalin, routinely processed in paraffin and examined histologically.

Microscopically, a circumscribed, noncapsulated lesion composed of hypocellular, eosinophilic concentric lamellae was found in the dermis, which resembled a large Vater-Pacini corpuscle, with no special structure in the center (Figure 1). This pattern transformed into irregular whorled and storiform pattern at periphery. The adjacent epidermis was slightly attenuated with hyperkeratosis.

The tumor stained red with van Gieson staining and was weakly PAS-positive. Silver impregnation showed fine reticulin fibers and only sparse fine elastic fibers. The Sirius red stain showed in the tumor green birefringence with polarized light (type III collagen) in contrast to the yellow color (mixed type I and III collagens)[8] of the surrounding dermis (Figure 2). Immunohistochemistry demonstrated CD34, vimentin and weak, but regular epithelial membrane antigen (EMA) positivity of the slim, elongated fibroblasts (Figure 3). The tumor was negative for glucose transporter 1 (GLUT-1), S100 protein, cytokeratin cocktail AE1/3,

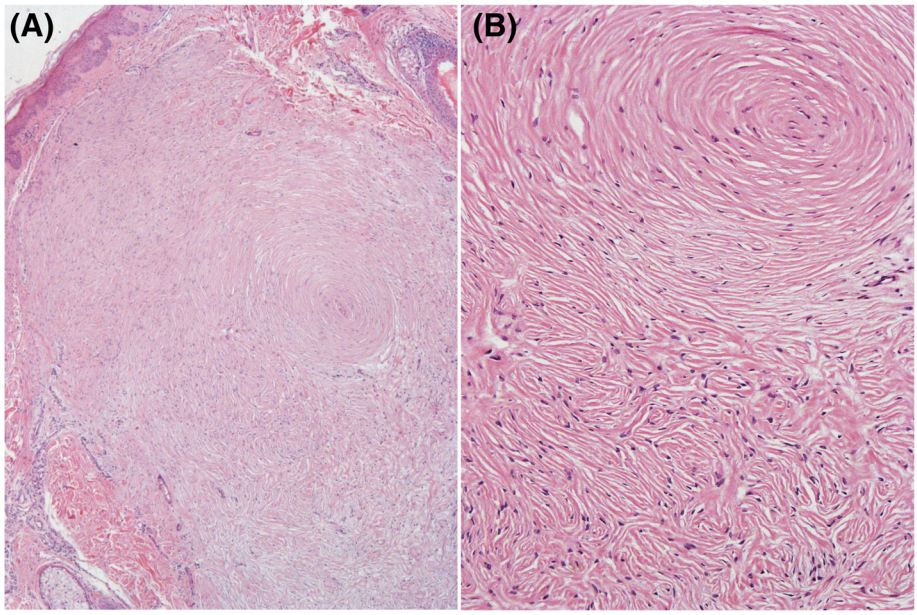


**FIGURE 1** The sclerotic fibroma with concentric arrangement of fibrous lamellae suggestive of Pacinian collagenoma, which continuously transfers into tissue with the pattern of sclerotic (storiform) fibroma. Hematoxylin and eosin; A, 25× and B, 100×

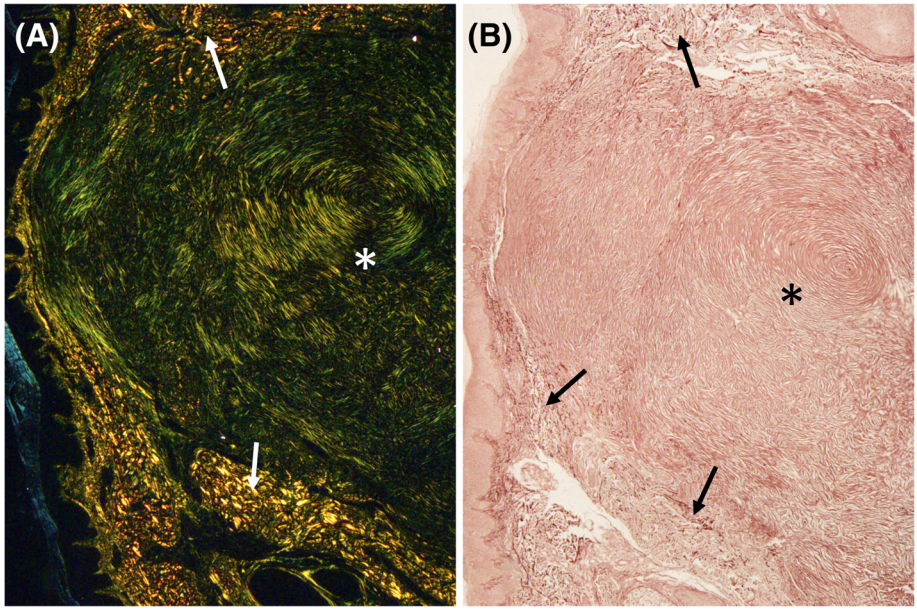


**FIGURE 2** A, Pacinian collagenoma with concentrically arranged lamellae giving green birefringence with polarized light (asterisk) in contrast with the yellowish-red collagen of the surrounding dermis (arrow). B, The collagenoma contained scant thin elastic fibers (asterisk) comparing with elastic fibers of the normal dermis (arrow). A, Sirius red, polarized light, 25×. B, Orcein, 25×

CD68, and NSE (neuron specific enolase). The diagnosis of a rare form of benign skin fibroma, the Pacinian collagenoma of the scalp, was established.

## 3 | DISCUSSION

A concentric lamellar growth pattern is very rare in fibromas. In 1999, Pillay et al[7] described a case of an SF composed of concentric lamellae with onion-skin-like appearance for the first time. Since these structures resembled Vater-Pacini corpuscles, this tumor was named "Pacinian collagenoma." Authors of this contribution, as well as Patterson[9] considered it to be a histological variant of SF.

Our case is histomorphologically similar to the previously reported one. We found a tumor in dermis composed of hypocellular, eosinophilic concentric lamellae with spindle-shaped CD34-positive cells. This growth pattern transformed into whorled and storiform pattern at periphery of the lesion, which is a typical feature of SF. Both cases occurred in young adults; the original case was described in a 36-year-old female,[7] and in our case the patient was a 33-year-old male.

Differential diagnoses of some other lesions need to be considered. Similar histological features appear in perineurioma, especially the fibrous and sclerotic variant, which typically contains concentric lamellar structures. However, some authors describe that sclerotic perineurioma typically shows epithelioid features which are not

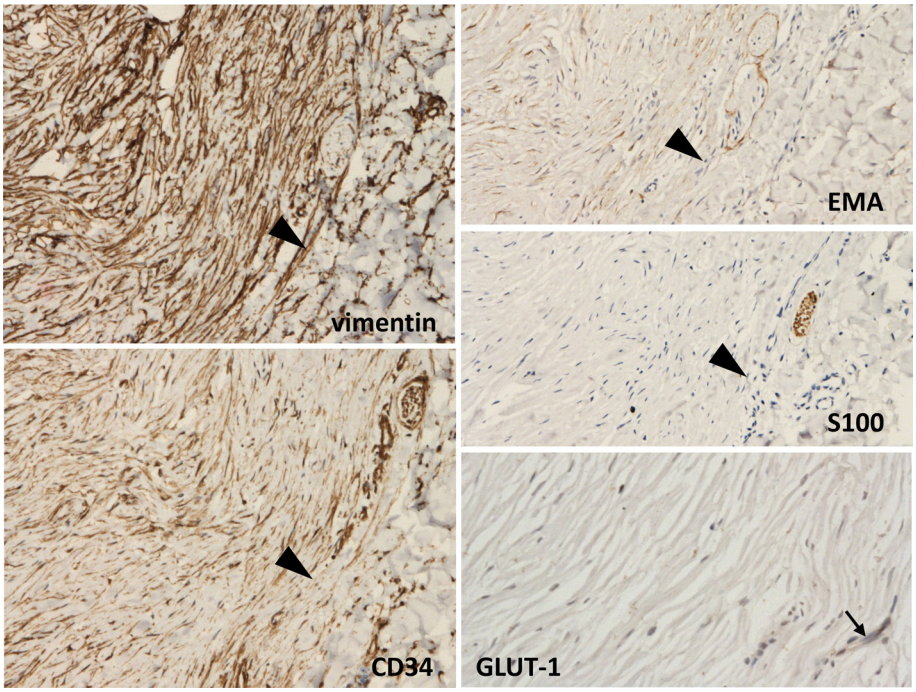


**FIGURE 3** Pacinian collagenoma with diffuse, strong positivity for vimentin and CD34, weak epithelial membrane antigen (EMA) positivity, negative for S100 (arrow points at positivity in nerve tissue; arrowhead indicates the tumor border) and glucose transporter 1 (GLUT-1) negativity in the tumor, with weak positivity in vascular endothelium (arrow). Avidin-biotin-peroxidase, DAB. 100×, GLUT-1200×

present in our case.[9-12] Absence of thick elastic fibers is typical for SF.[1-4,6,9] It has been reported that there is ongoing synthesis of collagen type I in SF.[13,14] However, synthesis of collagen type III seems to be another characteristic feature of this tumor, according to our finding.

According to Pillay et al, immunohistochemically perineurioma was almost identical to SF except for the EMA positivity, which could be hence used to differentiate between these two tumors.[7] Such assumption has not been confirmed by others since one half of the studied SF showed at least focal EMA positivity.[15] Even though expression of CD34 is common in perineuriomas,[16] it is not so in sclerosing perineuriomas, which were uniformly negative for CD34.[12,17,18] However, CD34 expression has been described in some cases of extra-acral sclerosing perineuriomas.[19,20] For differential diagnostics, it is useful to examine GLUT-1 expression, which is considered to be a sensitive marker for perineurial cells and is often positive in perineuriomas.[21,22]

Thus, the morphology, immunophenotype profile of CD34, and EMA positivity and GLUT-1 negativity of the presented case comply with the diagnosis of Pacinian collagenoma as a distinct type of SF.

Pacinian neurofibroma typically contains Vater-Pacini-like structures.[23] Other possible differential diagnosis includes dermatofibroma, in which changes similar to SF have been described, but with CD34 negativity.[24] Lesions histologically resembling SF have also been described in a fibroadenoma of axillary accessory breast tissue,[25] in sclerotic lipomas,[26] and in certain inflammatory conditions, such as chronic folliculitis,[27] but none of these lesions contained concentric lamellar structures.

In summary, we present a unique variant of SF composed of concentrically arranged collagen bundles, which resembles an enlarged Vater-Pacini corpuscle. The occurrence of concentric structures in fibromas is an uncommon trait and may represent a pitfall for establishing the diagnosis of Pacinian collagenoma. The clinical significance, as well as the genesis of these structures, is unknown because of their rare occurrence.

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