CASE REPORT

Sclerotic Fibroma of the skin. Case report

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ABSTRACT

Sclerotic fibroma of the skin is a soft tissue tumor, possibly associated to Cowden syndrome if presented as a multiple lesion. However, it can also manifest as a solitary tumor without any association. It occurs in male and female adults, typically manifesting at the face and limbs. The case of a patient with a single palmar lesion is presented without association to Cowden syndrome.

INTRODUCTION

Sclerotic fibroma of the skin, or storiform collagenoma, as denominated by several authors, is a soft-tissue, benign, infrequent tumor, presented as a pink, white, skin-colored papule or well-circumscribed solid nodule. It is slow-growing, and typically occurs in male and female young adults. It is usually located at the face and limbs. Nonetheless, there are cases reported in the scalp, trunk, oral mucosa and nail bed.[1–3]

CLINICAL CASE

A 45-year-old male patient presented with an asymptomatic lesion on the right palm with one-year evolution. Physical examination revealed skin-colored, hard, well-delimited papule with keratotic surface and a diameter measuring approximately 3 mm. (Fig 1) Dermoscopy showed circumscribed lesion, characterized by the presence of a white background separated by grayish areas without visible vessels. (Fig 2)

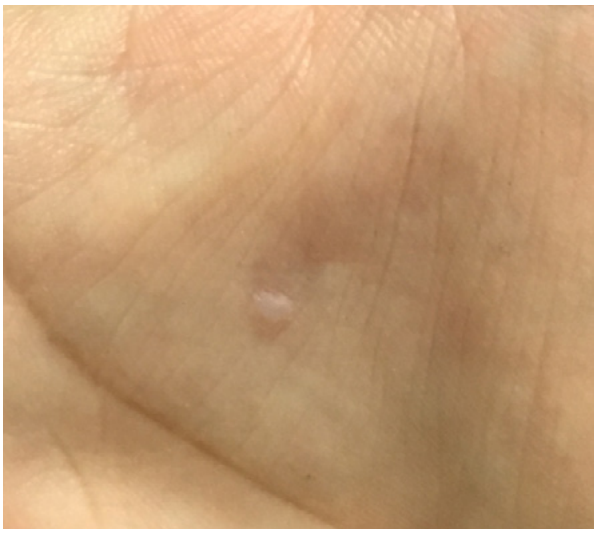

Fig 1. Skin-colored papule located at the right palm.

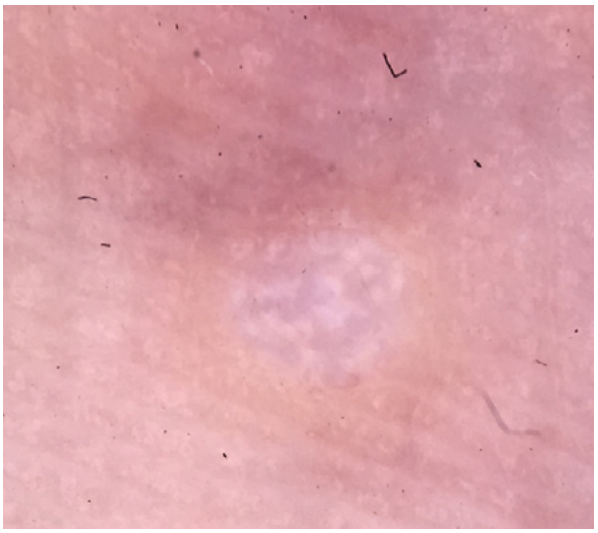

Fig 2. Circumscribed lesion with white background separated by grayish areas.

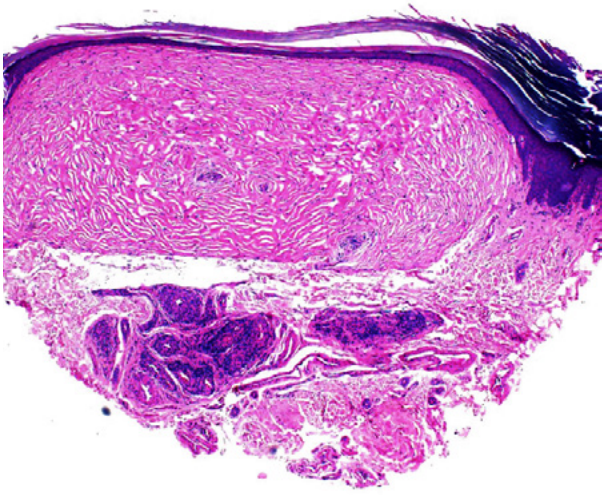

Fig 3. Focally expanded dermis by mesenchymal, hypocellular, hyaline, well-delimited and symmetric proliferation.

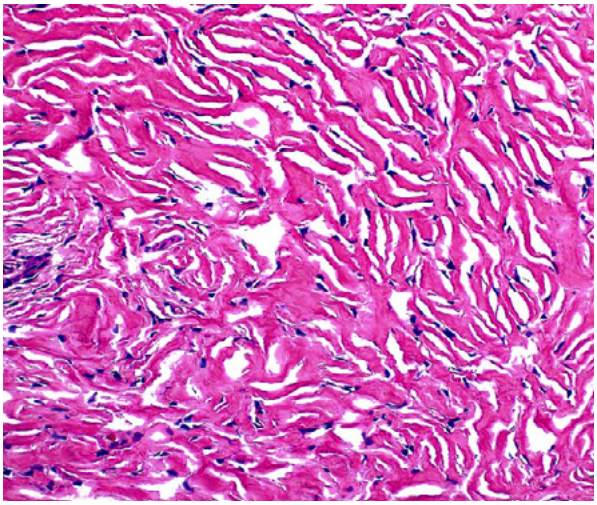

Fig 4. Dense, hyaline collagenous fibers separated by clefts.

Exeresis and histological study of the lesion was performed, revealing focally expanded dermis by well-delimited and symmetrical proliferation (Fig. 3) of dense, hyaline and retractile collagenous fibers in hypocellular stroma (Fig. 4). Histological diagnosis confirmed Sclerotic Fibroma.

DISCUSSION

Sclerotic fibroma of the skin (SF) was described by Weary[4] in 1972, when located in the tongue of a patient with Cowden's disease.[5] In 1989, Rapini and Golitz[6] presented 11 cases without any association to Cowden's disease, which afterwards denominated sclerotic fibroma of the skin.[5,7,8] In 1991, Requena et al.[9] concluded that the presence of multiple sclerotic fibromas constituted a cutaneous sign of Cowden's disease which may contribute to the syndrome's early diagnosis.[5]

SF is a benign, scarcely reported tumor which appears in young adults between the third and fourth life decade, without race or sexual predilection. It does manifest fairly more on women.[7] It can be presented as a solitary or multiple lesion. The latter must be considered as an indicator for Cowden's disease. Some authors suggest it to be the first clinical sign of the aforementioned syndrome. As a consequence, even when presented as a solitary lesion, the patient must be examined.[5,7,10]

Clinically, the lesion manifests as a well-circumscribed, solid, variable sized papule or nodule. In addition, it is usually white, pink or skin-colored. It is a slow-growing (months to years), asymptomatic lesion. SF commonly appears at the head or limbs. However, there are cases reported on the palms, plants, and even oral mucosa.[5,7]

Dermoscopically, a white homogeneous background with arborizing peripheral vessels is defined. Some types of dermatofibroma and amelanotic blue nevus can show similar patterns.[1]

Its etiopathogeny is still unknown. Notwithstanding, there are two theories that oppose its origin. The first theory describes SF as a proliferative fibrous tumor. The second theory defines it as a preexisting lesion's final involution phase, such as in dermatofibromas, angiofibromas, neurofibromas, melanocytic nevi, among others.[5,11,12]

Histopathologically, SF is described as a non-encapsulated, well-circumscribed neoformation, constituted by fascicles of thick, hyalinized and hypocellular collagen bundles, intertwining and forming clefts in between. These thick collagen bundles expose a whorled, whirlwind-like or windmill pattern, also known as storiform.[3,7] There are no adnexal structures and the suprajacent epidermis is usually atrophic.[11,13]

Immunohistochemistry reveals scarce mesenchymal cells, constituting the tumor, testing positive for vimentin and CD34, and testing negative for S-100, carcinoembryonic antigen, epithelial membrane antigen, high molecular weight cytokeratin.[3,5]

CD34 is selectively expressed by hematopoietic stem cells, endothelial cells, fusiform cells amidst adnexal structures and interstitial cells in the reticular dermis. This protein is not only found on sclerotic fibromas, but in other tumors as well: pleomorphic fibroma, breast fibroadenoma, dermatofibrosarcoma protuberans, hemangiopericytoma, glomus tumor, among others.[12]

Some studies support the theory about an active-growing neoformation, based on positive immunohistochemistry results for PCN and KI67, or human type I procollagen.[12]

It was noted that cells found on dense foci of the sclerotic fibroma presented with myofibroblastic differentiation. Therefore, a term (myopericyte) to label the transition between pericytes and smooth muscle cells, which are compared to myofibroblasts, was established. Myofibroblasts evidence features of smooth muscle cells and fibroblasts, which may exhibit different phenotypes, suggesting several functional phases or an origin from different precursors.[12]

Differential diagnosis includes dermatofibroma, pleomorphic fibroma, sclerotic lipoma, fibrolipoma, giant cell collagenoma, benign fibrous histiocytoma, intradermal Spitz nevus and giant cell angiohistiocytoma.[2,5]

Some consider it as a dermatofibroma variant. However, there are differentiating features:

- Clinical appearance.
- Atrophic epidermis.
- Dermal, hyalinized and hypocellular collagen bands which are separated by clefts containing abundant mucopolysaccharides.[8]

A discussion exists about whether sclerotic fibroma, which many authors denominate “storiform” collagenoma, corresponds to hypocellular dermatofibromas. It has been demonstrated that fibroblasts that are present in the lesion actively synthetize type I collagen. Thus, the aforementioned lesion must not be considered as a dermatofibroma.[13]

CONCLUSION

Sclerotic fibroma of the skin is a benign, infrequent tumor which does not usually require treatment. Its origin is discussed to date. There are few cases with dermoscopy reported in the literature. It may or may not be associated to Cowden’s disease. However, it is essential to emphasize that, even when presented as a solitary lesion, there must be a research about probable associations with such syndrome.

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