Fibroma Esclerótico Solitário

R. Bouceiro-Mendes[1], M. Mendonça-Sanches[1], S. Antunes-Duarte[1], L. Soares-de-Almeida[1,2]
[1]Centro Hospitalar Universitário Lisboa Norte, Hospital de Santa Maria, Serviço de Dermatologia
[2]Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa

RESUMO – O fibroma esclerótico (FE), também conhecido por colagenoma estoriforme, é um tumor cutâneo benigno e raro, que normalmente se manifesta como uma pápula ou nódulo assintomático e de crescimento lento. É mais comum na cabeça, região cervical e membros superiores, mas tem sido descrito também noutras localizações, como no tronco, mucosa oral e leito ungueal. O FE pode surgir como uma lesão solitária ou múltiplas, podendo ocorrer de forma esporádica ou em associação à síndrome de Cowden. Apresentamos um caso de FE solitário não associado à doença de Cowden com características clínicas e histológicas típicas.
PALAVRAS-CHAVE – Esclerose; Fibroma; Neoplasias da Pele.

Solitary Sclerotic Fibroma

ABSTRACT – *Sclerotic fibroma (SF) or storiform collagenoma is a rare and benign skin tumor. Clinically, it usually presents as an asymptomatic, slowly growing papule or nodule on the skin of the head, neck and upper extremities. Other locations have been described including the trunk, oral mucosa and nail bed. SF can present either as a solitary tumor or as multifocal cutaneous lesions and it may occur sporadically or in association with Cowden syndrome. Herein, we report a case of a solitary SF with typical clinical and histologic features, not associated with Cowden disease.*
KEYWORDS – *Fibroma; Sclerosis; Skin Neoplasms.*

INTRODUCTION

Sclerotic fibroma (SF), also known as storiform collagenoma is a rare, benign skin tumor that usually presents as a clinically asymptomatic, slowly growing, round to ovoid shaped, skin-colored papule or nodule.[1-3] It is more common on the skin of the head, neck and upper extremities but other locations have been described including the trunk, oral mucosa and nail bed.[2,4] It occurs both in males and females and affects mainly young adults and middle-aged individuals. SF manifests either as a solitary sporadic tumor or as multiple lesions within the cutaneous manifestation of Cowden disease, an autosomal dominant genodermatosis.[2-4] Herein, we present a case of a solitary SF with typical clinical and histologic characteristics, displaying hypocellular collagen bundles with clefts in a patient with no evidence of Cowden disease.

CASE REPORT

A 66-year-old Caucasian male patient was seen in our outpatient clinic due to an asymptomatic skin nodule on his left shoulder. The lesion had developed slowly over a period of more than twenty years. The patient denied previous local trauma, surgical interventions or any other similar skin lesions elsewhere. Physical examination revealed an asymptomatic, well-delimited, flesh-colored, 2 cm nodule, with a hard consistency, situated on the patients' left shoulder (Fig. 1). There were no other significant skin lesions.

The nodule was surgically excised in total. Histological examination revealed a well-circumscribed hypocellular dermal lesion consisting of hyalinized collagen bundles in a storiform pattern, separated by clefts. There was no architectural or cytologic atypia of the tumor cells, which were covered by an atrophic epidermis (Fig. 2A-B). Immunostaining was positive for CD34 (Fig. 2C). These findings were consistent with the diagnosis of SF. There was an uneventful postoperative recovery with no signs of relapse at 12-months' follow-up.

DISCUSSION

SF was first reported in 1972 by Weary *et al*. in a biopsy of a lesion from the tongue of a patient with Cowden syndrome.[1,5] When multiple lesions are present, SF serves as a

Correspondência: Rita Bouceiro Mendes
Hospital de Santa Maria
Serviço de Dermatologia, Piso 5
Av. Prof. Egas Moniz, 1649-035 Lisboa, Portugal
E-mail: rita.bouceiro.mendes@gmail.com
DOI: https://dx.doi.org/10.29021/spdv.78.2.1189

Recebido/*Received* 2020/04/15 | **Aceite/*Accepted*** 2020/05/14 | **Publicado/*Published*** 2020/06/30

Caso Clínico

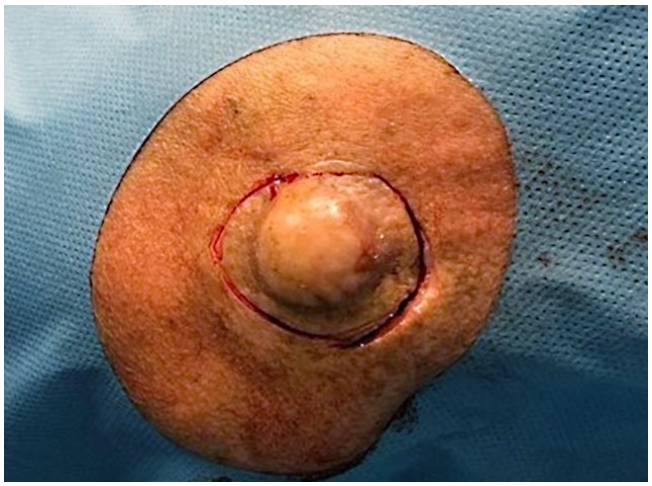

Figure 1 - Sclerotic fibroma, intraoperative picture: an elliptical excision was performed followed by primary wound closure.

clue for the presence of Cowden syndrome.[3] In 1989, Rapini and Golitz reported 11 cases of solitary SF in patients without Cowden disease.[1,2] In fact, when present as a solitary lesion, such as in our case, SF is benign, and treatment is usually not warranted, apart for aesthetic reasons.[1] However, even in such cases, a detailed clinical and physical investigation of the patient is recommended, in order to rule out the diagnosis of Cowden syndrome.

Despite its low incidence it must be considered in the differential diagnosis of other well-circumscribed skin tumors, such as dermatofibroma, sclerotic lipoma, fibrolipoma, giant cell collagenoma, pleomorphic fibroma, benign fibrous histiocytoma, intradermal Spitz nevus and giant cell angiohistiocytoma.[2] Since the clinical signs are non-specific, histology is required to confirm the diagnosis.[6] On histopathology sections, SF is a sharply circumscribed, non-capsulated, but well demarcated dermal nodule comprising hyalinized and dense collagen bundles arranged in a plywood-like or whorled pattern, with striking hypocellularity to almost complete acellularity (with scarce fibroblasts). Prominent clefts are present between the thick collagen bundles.[1-3,5-7] Immunohistochemistry of these lesions reveals positivity for CD34+ and vimentin and negativity for neurofilaments, S-100, neuron-specific enolase (NSE), carcinogenic embryonic antigen (CEA), embryonic membrane antigen (EMA), high molecular weight keratin and cytokeratin.[1,2] It is unclear whether SF represents the later and sclerotic stage of other more cellular neoplasms or a true individualized fibrocystic tumor.[3,4,6,7] The former opinion is based by the findings that dermatofibroma-like tissue is often admixed with SF tissue.[6,8] The opinion that SF is a distinct, sharply defined fibroma with a peculiar fast tendency towards an early sclerotic stage is supported by the presence of ongoing type I collagen synthesis and positive immunostaining for proliferating cell nuclear antigen (PCNA) and Ki-67, that suggest that it is in fact a growing and proliferating neoplasm.[6,7]

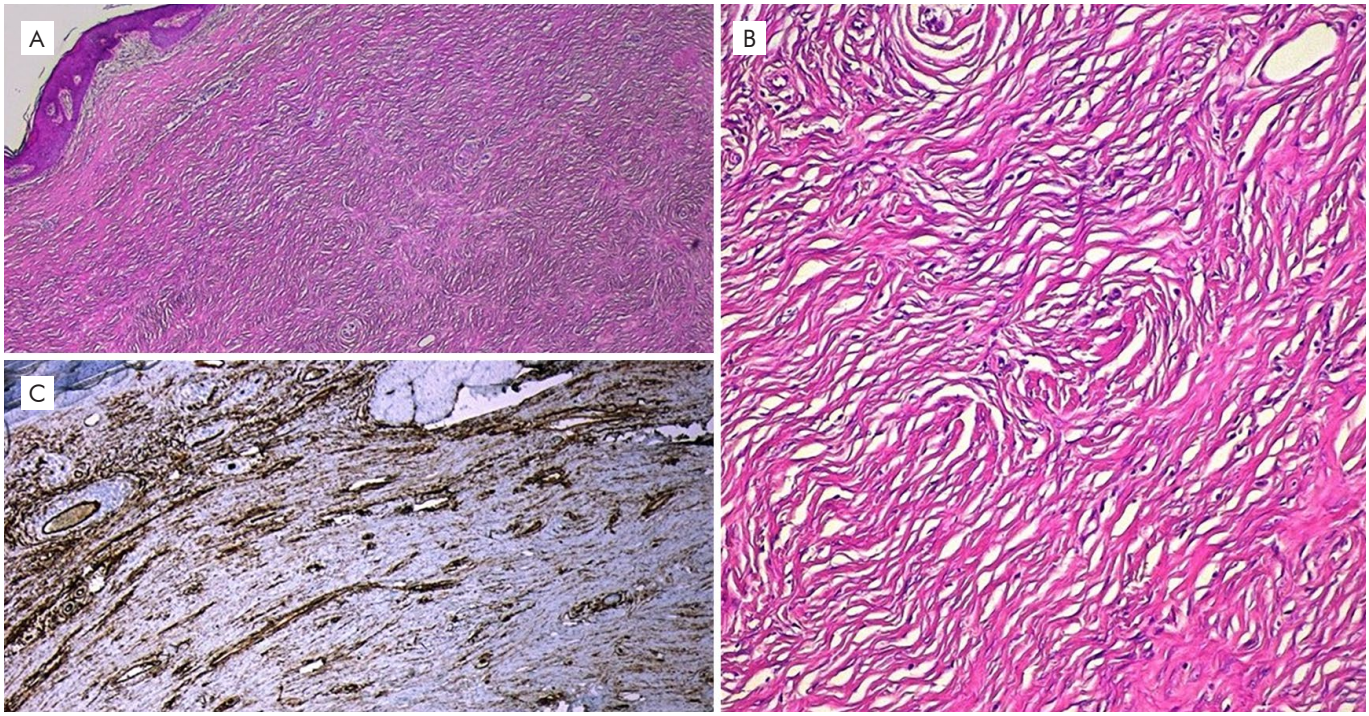

Figure 2 - Sclerotic fibroma, histopathological aspects: **(A)** Well-circumscribed diffuse hypocellular dermal nodule (H&E x25); **(B)** Hyalinized collagen bundles in a storiform pattern, separated by empty clefts (H&E, x100); **(C)** Positive immunostaining for CD34 (CD34, x100).

In sum, SF is a rare skin tumor that has a very characteristic histology, but whose clinical signs are non-specific. As such, SF must be considered in the differential diagnosis of other well-circumscribed skin tumors. When multiple lesions are present, Cowden disease must be excluded. When present as a solitary lesion, as in the presented clinical case, no further investigation or treatment are generally required.

Conflitos de interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
Fontes de financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.
Confidencialidade dos dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
Consentimento: Consentimento do doente para publicação obtido.
Proveniência e revisão por pares: Não comissionado; revisão externa por pares.

Conflicts of interest: *The authors have no conflicts of interest to declare.*
Financing support: *This work has not received any contribution, grant or scholarship.*
Confidentiality of data: *The authors declare that they have followed the protocols of their work center on the publication of data from patients.*
Patient Consent: *Consent for publication was obtained.*
Provenance and peer review: *Not commissioned; externally peer reviewed*

ORCID
R. Bouceiro-Mendes
https://orcid.org/0000-0002-5034-3613
M. Mendonça-Sanches
https://orcid.org/0000-0001-6940-0123
S. Antunes-Duarte
https://orcid.org/0000-0003-3662-0423
L. Soares-de-Almeida
https://orcid.org/0000-0003-4026-6105

REFERENCES

1. Bhambri A, Del Rosso JQ. Solitary sclerotic fibroma. J Clin Aesthet Dermatol. 2009;2:36-8.
2. Stocchero GF. Storiform collagenoma: case report. Einstein. 2015;13:103-5. doi: 10.1590/S1679--45082015RC2907.
3. Pernet C, Durand L, Bessis D, Guillot B, Dereure O. Solitary sclerotic fibroma of the skin: a possible clue for Cowden syndrome. Eur J Dermatol. 2012;22:278-9. doi: 10.1684/ejd.2012.1655.
4. Ebadian M, Citarella L, Collins D, Diaz-Cano S, Pozo--Garcia L. Dermoscopy of a solitary storiform collagenoma. Dermatol Pract Concept. 2018;8:120-2. doi: 10.5826/dpc.0802a09.
5. Nakashima K, Yamada N, Adachi K, Yoshida Y, Yamamoto O. Solitary sclerotic fibroma of the skin: morphological characterization of the 'plywood-like pattern'. J Cut Pathol. 2008;35:74-9. doi: 10.1111/j.1600--0560.2008.01001.x.
6. Tosa M, Ansai S-i, Kuwahara H, Akaishi S, Ogawa R. Two cases of sclerotic fibroma of the skin that mimicked keloids clinically. J Nippon Med Sch. 2018;85:283-6. doi: 10.1272/jnms.JNMS.2018; 85-45.
7. Herrera Sánchez M, Suánez Fernández R, del Cerro Heredero M, Rueda Gómez-Calcerrada M, Vigaray Conde J, Sánchez Yus E. Sclerotic fibroma. Dermatology. 1998;196:429-30.
8. Pujol RM, de Castro F, Schroeter AL, Daniel Su WP. Solitary sclerotic fibroma of the skin: a sclerotic dermatofibroma? Am J Dermatopathol. 1996;18:620-4.