Circumscribed storiform collagenoma, an unusual tumor

Francisco Javier Lira-Valero[1] MD, Eunice Rebeca Carrillo-Cisneros[1] MD, Nancy Pulido-Díaz[1] MD, Marissa de Jesús Quintal-Ramírez[2] MD, Liliana Godínez-Aldrete[1] MD

Affiliations: [1]Department of Dermatology, Centro Médico Nacional La Raza, Instituto Mexicano del Seguro Social, México City, México., [2]Department of Pathology, Centro Médico Nacional La Raza, Instituto Mexicano del Seguro Social, México City, México

Corresponding Author: Francisco Javier Lira-Valero, Seris y Zaachila s/n, Colonia La Raza, Delegación Azcapotzalco, CP 02990, México City, México, Tel: 52-8712600968, Email: fjaver.valero@gmail.com

Abstract

Circumscribed storiform collagenoma is a rare benign tumor. It appears as an isolated skin lesion or as part of the clinical spectrum of Cowden syndrome. The pathogenesis is still controversial. Although its clinical expression is heterogeneous, it has a characteristic histological pattern. We describe a case of a solitary circumscribed storiform collagenoma not associated with Cowden syndrome.

Keywords: circumscribed storiform collagenoma, solitary sclerotic fibroma, Cowden syndrome

Introduction

Circumscribed storiform collagenoma (CSC), also known as solitary sclerotic fibroma, is a rare benign tumor; there are less than 100 case reports in the world literature [1,2]. This condition was recognized in 1972 by Weary et al. in the tongue biopsy specimen of a patient with Cowden syndrome (CS), [2]. This syndrome is characterized by the development of multiple hamartomas in different body segments, mainly in the skin and gastrointestinal tract; its prevalence has been estimated as 1:200,000 individuals [3]. We describe a case of a CSC not associated with Cowden disease.

Case Synopsis

A 56-year-old woman with a medical history of long-standing psoriasis treated with subcutaneous methotrexate 25mg/week reported an asymptomatic skin lesion located on the fourth finger of the left hand. The skin lesion had been present for seven years, but she never requested medical assessment. The physical examination revealed a firm, skin-colored, dome-shaped nodule that was 7mm in diameter (**Figure 1**). On dermoscopic evaluation there was a structureless white background surrounded by a delicate erythematous halo. Short arborizing vessels and a focal area with thick yellowish scale were also noted (**Figure 2**). The patient underwent excisional biopsy of the nodule. Histopathological examination showed a well-circumscribed, hypocellular dermal nodule, composed of hyalinized collagen bundles arranged in a whorled pattern separated by clefts, consistent with a CSC (**Figure 3**). There was no recurrence of the skin lesion in the first year.

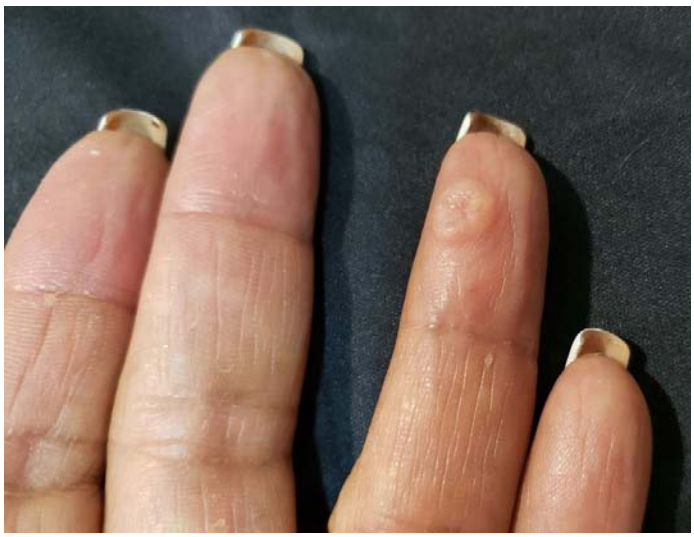

Figure 1. *A firm, skin color, dome-shaped nodule that was 7 mm in diameter located on the fourth finger of the left hand.*

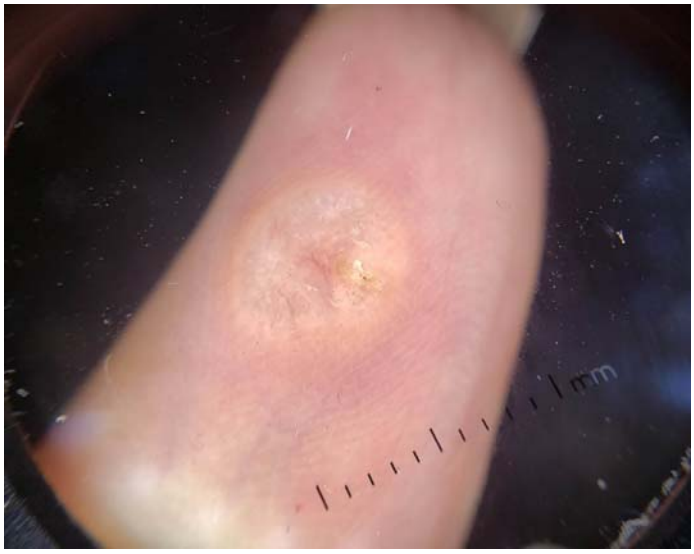

Figure 2. *Dermoscopy. Structureless white background surrounded by a delicate erythematous halo, short arborizing vessels and a focal area with thick yellowish scale.*

Case Discussion

Multiple CSCs have been considered as another specific cutaneous marker of CS, as clinically useful as facial trichilemmomas [4]. In some case reports, a solitary CSC was the only clue before establishing a definitive diagnosis of CS [5,6]. Rapini et al. reported eleven patients with solitary lesions without clinical evidence of this genodermatosis [7]. Circumscribed storiform collagenoma has also been described in patients with Lhermitte-Duclos disease, Bannayan-Ruvalcaba-Riley syndrome, and Rubinstein-Taybi syndrome [8].

The pathogenesis of CSC is still controversial. Its association with Cowden syndrome, the tendency to recurrence, the expression of cell proliferation markers such as PCNA (proliferating cell nuclear antigen) and Ki-67, and the evidence of synthesis of type I collagen support the concept that it is a fibrous tumor with active growth [4-6,9-11]. In contrast, the presence of focal areas with CSC-like changes in other neoplastic and inflammatory lesions (dermatofibroma, neurofibroma, angiofibroma, fibroma of the tendon sheath, pleomorphic fibroma, melanocytic nevi, sclerotic lipoma, giant cell collagenomas, giant cell angiohistiocytoma, erythema elevatum diutinum, or folliculitis) supports the theory that CSC is an involutional stage of a preexisting lesion or a distinctive pattern of degenerated connective tissue [1,2,12,13].

Circumscribed storiform collagenoma occurs in middle-aged individuals with a slight predominance in women [2,5]. It appears as a firm, pink, whitish or skin color papule or nodule. It is usually an asymptomatic skin lesion with a slow growth pattern [2,8]. It is more frequent in the face, neck, and limbs, but it can also appear on the trunk, scalp, oral mucosa and even unusual sites as nail bed or nasolacrimal duct [1,2,5,12-14].

Histopathologically, storiform collagenoma is a well-circumscribed but unencapsulated, hypocellular dermal proliferation, composed of hyalinized collagen bundles arranged in a whorled or plywood-like pattern, separated by clefts [6,12]. Immunohistochemical stains are positive for vimentin, focally positive for CD34, and factor XIIIa

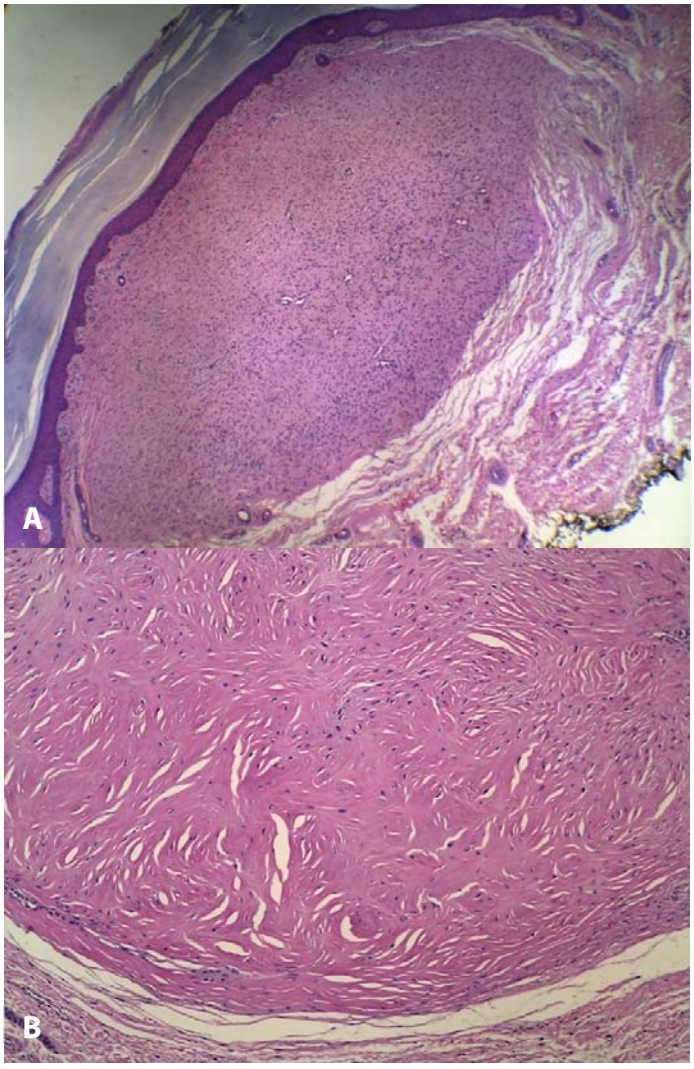

Figure 3. *A well-circumscribed, hypocellular dermal nodule, composed of hyalinized collagen bundles arranged in a whorled pattern separated by clefts. H&E,* ***A)*** *10×,* ***B)*** *40×.*

[2,6]. Electron microscopy reveals collagen fibrils of 40-50nm in diameter interspersed with electrodense parallel linear structures; these collagen fibrils also show cross striations that appear approximately every 70nm [15].

There are no dermoscopic criteria for CSC owing to its rarity. Ebadina et al. made the first dermoscopic description and noted a homogeneous white background with erythema and peripheral arborizing vessels [16]. A dermoscopic and histopathological correlation has not been established. Our hypothesis is that the structureless white area corresponds to hyalinized thick bands of collagen, whereas the short arborizing vessels correlate with vascular structures in the upper dermis adjacent to the tumor. The erythema may correspond to a phenomenon of local vascular congestion.

This tumor can be treated with standard surgical resection; occasionally, it may show recurrence [8]. A recurrence time from 2.5 to 7 years after removal has been reported [9,10].

Conclusion

Circumscribed storiform collagenoma is a rare benign tumor with a fairly heterogeneous clinical expression. However, the histopathological features are diagnostic in most cases. Although, it can appear as an isolated skin lesion we must exclude syndromic associations, mostly with Cowden disease.

Potential conflicts of interest

The authors declare no conflicts of interests.

References

1. Prasher P. Solitary sclerotic fibroma as a rare cause of nasolacrimal duct obstruction. *Orbit*. 2008;27:441-3. [PMID: 19085300].
2. Stocchero GF. Storiform collagenoma: Case report. *Einstein (Sao Paulo)*. 2015;13:103-5. [PMID: 25993076].
3. Lopes S, Vide J, Moreira E, Azevedo F. Cowden syndrome: Clinical case and a brief review. *Dermatol Online J*. 2017;23:13030/qt0023k3x0. [PMID: 29469739].
4. Requena L, Gutiérrez J, Sánchez Yus E. Multiple sclerotic fibromas of the skin. A cutaneous marker of Cowden's disease. *J Cutan Pathol*. 1992;19:346-51. [PMID: 1430474].
5. Al-Daraji WI, Ramsay HM, Ali RB. Storiform collagenoma as a clue for Cowden disease or PTEN hamartoma tumour syndrome. *J Clin Pathol*. 2007;60:840-2. [PMID: 17513505].
6. 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. [PMID: 22377798].
7. Rapini RP, Golitz LE. Sclerotic fibromas of the skin. *J Am Acad Dermatol*. 1989;20:266-71. [PMID: 2464630].
8. Zavras N, Mennonna R, Maris S, Vaos G. Circumscribed storiform collagenoma associated with Rubinstein-Taybi syndrome in a young adolescent. *Case Rep Dermatol*. 2016;8:59-63. [PMID: 27065842].
9. Cohen PR, Tschen JA, Abaya-Blas R, Cochran RJ. Recurrent sclerotic fibroma of the skin. *Am J Dermatopathol*. 1999;21:571-4. [PMID: 10608253].
10. McCalmont TH. Sclerotic fibroma: A fossil no longer. *J Cutan Pathol*. 1994;21:82-5. [PMID: 8188939].
11. Shitabata PK, Crouch EC, Fitzgibbon JF, et al. Cutaneous sclerotic fibroma. Immunohistochemical evidence of a fibroblastic neoplasm with ongoing type I collagen synthesis. *Am J Dermatopathol*. 1995;17:339-43. [PMID: 8600796].
12. Abbas O, Ghosn S, Bahhady R, Salman S. Solitary sclerotic fibroma on the scalp of a young girl: Reactive sclerosis pattern? *J Dermatol*. 2010;37:575-7. [PMID: 20536679].
13. Ide F, Mishima K, Saito I. Solitary sclerotic fibroma of the lip. *Br J Dermatol*. 2003;149:433-5. [PMID: 12932266].
14. González-Vela MC, Val-Bernal JF, Sánchez-Santolino S, Saiz-Bustillo R. Solitary sclerotic fibroma of the oral mucosa. *Am J Dermatopathol*. 2004;26:519-20. [PMID: 15618939].
15. Lo WL, Wong CK. Solitary sclerotic fibroma. *J Cutan Pathol*. 1990;17:269-73. [PMID: 2258475].
16. Ebadian M, Citarella L, Collins D, Diaz-Cano S, Pozo-Garcia L. Dermoscopy of a solitary storiform collagenoma. *Dermatol Pract Concept*. 2018;8:120-122. [PMID: 29785328].