

## A Case of Superficial Collagenous Fibroma and Review of the Literature

Selma Emre,<sup>1\*</sup> MD, Gülşen Akoğlu,<sup>1</sup> MD, Gonca Özgün,<sup>2</sup> MD, Ahmet Metin,<sup>1</sup> MD, Sibel Orhun Yavuz,<sup>3</sup> MD

Address: <sup>1</sup>Department of Dermatology, Ankara Atatürk Education and Research Hospital, <sup>2</sup>Department of Pathology, Uludağ University, Faculty of Medicine, <sup>3</sup>Department of Pathology, Ankara Atatürk Education and Research Hospital, Turkey.

E-mail: dr\_semre@yahoo.com

\* Corresponding Author: Selma Emre, MD, Department of Dermatology, Ankara Atatürk Education and Research Hospital, Turkey.

Published:

J Turk Acad Dermatol 2014; 8 (3): 1483c4

This article is available from: <http://www.jtad.org/2014/3/jtad1483c4.pdf>

**Key Words:** Benign soft tissue tumor, collagenous fibroma, desmoplastic fibroblastoma

### Abstract

**Observation:** Collagenous fibroma (desmoplastic fibroblastoma) is a rare and benign soft tissue tumor, affecting mainly men in their 5th through 7th decades. These lesions are well-circumscribed, slow-growing, firm, and painless masses of 1-15 cm diameter with oval, fusiform or disc shape. The tumor consists of hypocellular proliferation of the spindle to stellate-shaped mesenchymal cells, embedded in a hyalinized collagenous stroma. Up till now, approximately 120 cases have been reported in the literature. In the English dermatologic literature, only 5 cases have been discussed before. In this report, a 65 year old male patient with superficial collagenous fibroma presenting with an unusual clinical appearance over the left breast has been described. The clinicopathological features are also discussed in comparing with other cases reported in the literature.

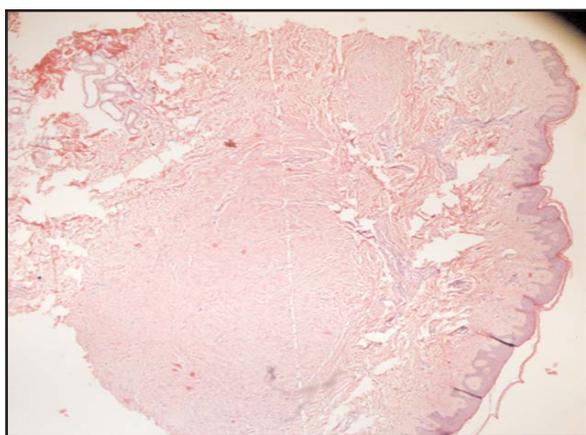
### Introduction

Collagenous fibroma (desmoplastic fibroblastoma) (CF) is a rare and benign soft tissue tumor. It is firstly defined by *Evans* with 7 cases as desmoplastic fibroblastoma in 1995 [1]. These lesions are well-circumscribed, slow-growing, firm, and painless masses of 1-15 cm diameter with oval, fusiform or disc shape [2, 3]. While it can be encountered in both genders, it is more commonly seen in men in 5 to 7 decades. Typically, it affects the subcutaneous tissue and skeletal muscles [4]. CF may be located anywhere in the body, however, most common sites are known to be arms, shoulders, neck and upper part of back, feet and ankles, legs, hands, abdomen and waist, respectively [5]. The tumor consists of hypocellular proliferation of the spindle to stellate-shaped mesenchymal cells,

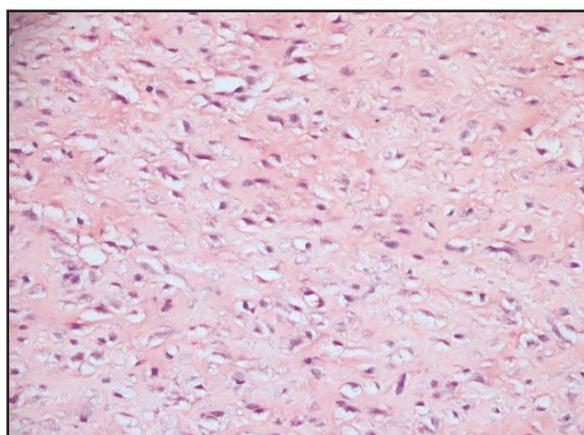
embedded in a hyalinized collagenous stroma. In 1996, *Nielsen et al* renamed this entity as collagenous fibroma depending on the ultrastructural studies which showed the



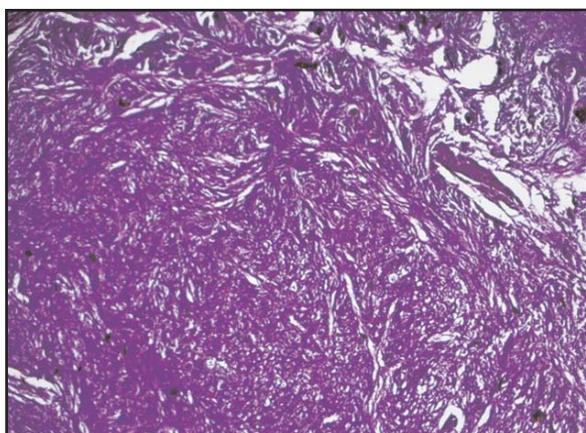
**Figure 1.** Linear papulonodular lesion over the upper-medial part of the left breast.



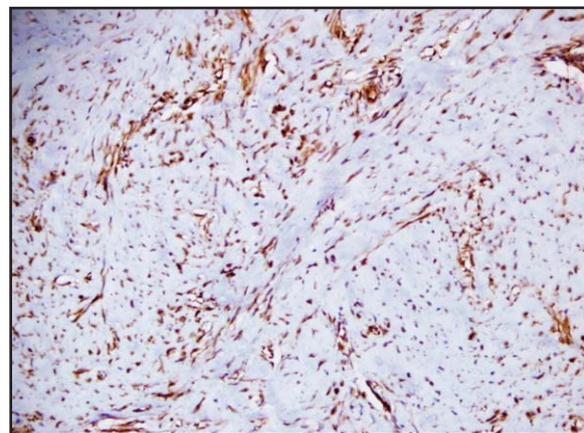
**Figure 2.** Well-circumscribed nodular lesion seen in the dermis (H&E; x40).



**Figure 3.** Spindle and stellate shaped cells within the eosinophilic stroma (H&E; x400).



**Figure 4.** Trichrome staining; diffuse and intense positivity (x200).



**Figure 5.** Positivity with vimentin staining (x200)

tumor cells to be mature fibroblasts and myofibroblasts [6]. Up till now, approximately 120 cases have been reported in the literature. In the English dermatologic literature, only 5 cases have been discussed before. In this report a case of superficial CF on an unusual localization has been described and compared with the clinicopathological features of other cases previously published.

### Case Report

A 65-year-old male patient presented to our outpatient clinic with a fusiform swelling that has been slowly growing for 2 years. Dermatological examination revealed a firm, pinkish, well-circumscribed, linear papulonodular lesion with a size of 1x5 cm over the upper-medial part of the left breast (Figure 1). There was no history of trauma or surgery to the site of the lesion before it was noticed. He had no subjective complaint such as pruritus or pain. He had been operated because

of an inverted papilloma in the bladder and had adrenalectomy due to an adrenal cortical adenoma. Family history was not remarkable. Routine laboratory analyses (complete blood count, liver and kidney function tests, and complete urinalysis) were all within normal limits. Histopathological examination of the skin demonstrated a well-circumscribed, collagen-rich and nodular lesion in the dermis (Figure 2). The lesion consisted of spindle and stellate-shaped cells within the eosinophilic stroma. The cells exhibited an occasional perinuclear transparency and wedge-shaped appearance (Figure 3). Trichrome staining showed a diffuse and intense positivity (Figure 4). Immunohistochemical staining with SMA (smooth muscle actin) and vimentin were positive (Figure 5). S-100, CD34, panCK (cyto-keratin), and CD68 were all negative. Based on these clinical and histopathological findings, the patient was diagnosed as CF. The magnetic resonance imaging of thorax did not show any muscle or bone invasion. After informed consent, CF lesion was treated with

total surgical excision. No recurrence or new lesion was observed after a one year follow-up.

## Discussion

Since CF may be mistaken for many benign and malign soft tissue tumors, the pathologists generally diagnose this rare neoplasm by histopathological examination. Therefore, approximately 90% of CF cases were found to be reported primarily by pathologists when the literature was looked through. Although CF is a benign neoplasm, the localization of the tumor may have a mass effect and invade the adjacent tissues. About 10% of all cases were reported by ophthalmologists, otolaryngologists, orthopedists, or surgeons who excised these tumors which were symptomatic in their localization, or by radiologists who diagnosed interesting radiological findings. In the present case, the lesion was an asymptomatic mass that has been growing for 2 years. Depending on the typical histopathological findings of CF, the diagnosis was established.

Histopathologically, CFs are well-circumscribed and noncapsulated tumors. Stellate or spindle shaped fibroblast-like cells forming the nodules are dispersed within the collagenous or fibromyxoid matrix. Mitotic activity is either absent or very limited. Immunohistochemical methods are beneficial in verification of the diagnosis. Vimentin stains diffusely positive. Muscle-specific actin and SMA display varying degrees of positivity. Desmin, CD34, and S-100 protein are negative [5, 7]. In the present case, results of typical stains were found for CF. Keloid, dermatofibroma and leiomyoma were the main differential diagnoses. Negative stain for CD34 excluded dermatofibroma. Actin and desmin stains were negative in contrast to leiomyoma. The histopathological findings demonstrated relatively well-circumscribed, collagen-rich and nodular pseudo-capsulated lesion. The cells were dispersed into the fibrohyalinized stroma. These findings were not similar with keloid tissue, which has an ill-defined, infiltrative appearance with fibrotic areas consisting of rough, bright, and thick collagen fibers in the center. Histopathological findings together with no recurrence after surgery supported that the lesion was not a keloid.

*Miettinen* and *Fetsch* reported the largest series of CF with 63 cases, which had lesions mainly

on the upper extremities [5]. The other affected body regions were lower extremities, head and neck region, and trunk, respectively [8, 9].

Most of the CF lesions are localized in deep soft tissue, infiltrating the underlying muscle and/or bone, or in articular or intraabdominal spaces. The muscle invasion was detected in about 80% of these cases. CF lesions were usually defined as painless, long standing, slowly growing movable masses. Most of them were excised because of mass effect as an eyelid edema due to CF of the lacrimal gland [10], lower abdominal mass or invasion of the lesion into the adjacent tissues such as vertebral bone [11], the neck causing neurological symptoms [12], and hip [13]. Trunk appears to be the least involved body region with CFs. These tumors were mostly reported to be deep seated. They usually infiltrate the underlying muscle or bone [2, 3, 4, 5, 11, 14, 15, 16, 17]. Their localizations were scapular [3], infrascapular region [2], abdominal wall [5], vertebra [11], lower intraabdomen [2, 14], trapezius muscle [15], supraclavicular fossa [16], pectoralis minor muscle [4], and back [17]. Moreover, there are some cases of uncommon CF locations, in the oral mucosa, tongue and orbital area [18, 19, 20, 21, 22]. The last reported unusual CF case was characterized as an ulceration on toe that mimics a diabetic foot ulcer [23]. The present case represents the CF lesion over the left breast, which has not been described before.

The superficial form involving dermis and/or subcutis has been found to be well documented only in 5 recent cases in the English literature [2, 7, 24]. Among them, 2 are located on the trunk as in our case.

Depending on localization, clinical and histopathological features of the tumor, many benign and malign soft tissue tumors and reactive fibroblastic lesions should be considered in the differential diagnosis of CF. Desmoid tumor, low grade fibromyxoid sarcoma, neurofibroma, calcifying fibrous pseudofibrous tumor, solitary fibrous tumor, elastofibroma, sclerotic fibroma of the skin, fibromatosis, perineuroma, nodular fasciitis, Gardner fibroma, and aponeuritic fibroma, should be kept in mind [25, 26]. These lesions may be usually differentiated by histopathological examination.

In the recent studies, cytogenetic analysis has been performed to understand the pathogenesis of CF [5]. Upon these findings, the breakpoint at chromosome 11q12 was considered to be pathogenetic in CF. Researchers suggested that chromosomal rearrangements involving 11q12 may be a useful diagnostic adjunct in distinguishing CF from desmoid tumors and nodular fasciitis [27, 28]. Desmoid tumors usually have extra copies of chromosome 8 or 20 or both of them, or loss of the 5q21~q22 region [29]. Although only 3 cases of nodular fasciitis have been cytogenetically analyzed, 2 of them showed rearrangements with a 3q21 breakpoint [30].

The cytogenetic analyses of CF both help to distinguish CF from other important lesions and also answer the questions on the speculations whether CF is a neoplasm or a reactive condition. Specific break points in the DNA [29], no inciting events and no defined specific cause of a reactive fibrous proliferation [1, 4] support the suggestions that CF is a benign neoplasm rather than a reactive process.

The treatment of CF is basically total excision. Therefore, the correct diagnosis of CF avoids unnecessary wider excisions and many other therapies such as chemotherapy and radiotherapy. *Miettinen* and *Fetsch* reported no recurrence after excision of the tumor in 39 patients who were followed up for a period of 11 years [5]. There is no data indicating what would happen if the tumor is remained untreated. In this report, the CF lesion was totally excised. After a follow-up period of 1 year, no new lesion or recurrence was observed.

In conclusion, CF is a rare tumor which has a limited available data about its typical clinical appearance and behavior of the disease. This case is a remarkable one due to having a clinical appearance. We believe that CF should be kept in mind in the differential diagnosis of hypertrophic lesions.

## References

- Evans HL. Desmoplastic fibroma: A report of seven cases. *Am J Surg Pathol* 1995; 19: 1077-1081. PMID: 7661281
- Huang HY, Sung MT, Eng HL, et al. Superficial collagenous fibroma: immunohistochemical, ultrastructural, and flow cytometric study of three cases, including one pemphigus vulgaris patient with a dermal mass. *APMIS*. 2002; 110: 283-289. PMID: 12076263
- Merriman DJ, Deavers MT, Czerniak BA, Lin PP. Massive desmoplastic fibroblastoma with scapular invasion. *Orthopedics* 2010; 33. doi: 10.3928/01477447-20100625-23. PMID: 20704102
- Dagli M, Eryilmaz E, Acar A, Kulacoglu S, Akmansu H. Collagenous fibroma (Desmoplastic fibroblastoma). *Yonsei Med J* 2004; 45: 941-943. PMID: 15515209
- Miettinen M, Fetsch JF. Collagenous fibroma (desmoplastic fibroblastoma): A clinicopathologic analysis of 63 cases of a distinctive soft tissue lesion with stellate-shaped fibroblast. *Hum Pathol* 1998; 29: 676-682. PMID: 9670823
- Nielsen GP, O'Connell JX, Dickersin GR, Rosenberg AE. Collagenous fibroma (desmoplastic fibroblastoma): A report of seven cases. *Mod Pathol* 1996; 9: 781-785. PMID: 8832562
- Kim JH, Oh DY, Kim SW, Lee JH, Ahn ST, Rhie JW. Desmoplastic fibroblastoma of the finger tip in an adult. *Arch Plast Surg* 2012; 39: 84-86. PMID: 22783503
- Singh NG, Mannan AA, Kahvic M. Desmoplastic fibroblastoma (collagenous fibroma): report of a case. *Indian J Pathol Microbiol*. 2011; 54: 206-207. PMID: 21393923
- Kamata Y, Anazawa U, Morioka H, et al. Natural evolution of desmoplastic fibroblastoma on magnetic resonance imaging: a case report. *J Med Case Rep* 2011; 5: 139. PMID: 21470431
- Ahn M, Osipov V, Harris GJ. Collagenous fibroma (desmoplastic fibroblastoma) of the lacrimal gland. *Ophthal Plast Reconstr Surg* 2009; 25: 250-252. PMID: 19454950
- Osipov V, Carrera GF. Collagenous fibroma (Desmoplastic fibroblastoma) with vertebral body erosion. *Sarcoma* 2009; 2009: 682687. PMID: 19503798
- Fong F, Odell E, Simo R. Collagenous fibroma (desmoplastic fibroblastoma) of the neck presenting with neurological symptoms. *Head Neck Pathol* 2009; 3: 47-50. PMID: 20596989
- Beggs I, Salter DS, Dorfman HD. Synovial desmoplastic fibroblastoma of hip joint with bone erosion. *Skeletal Radiology* 1999; 28: 402-406. PMID: 10478622
- Shuto R, Kiyosue H, Hori Y, et al. CT and MR imaging of desmoplastic fibroblastoma. *Eur Radiol* 2002; 12: 2474-2476. PMID: 12271387
- Alberghini M, Pasquinelli G, Zanella L, et al. Desmoplastic fibroblastoma: a light and ultrastructural description of two cases. *Ultrastruct Pathol* 2004; 28: 149-157. PMID: 15471428
- Bernal K, Nelson M, Neff JR, et al. Translocation (2;11)(q31;q12) is recurrent in collagenous fibroma (desmoplastic fibroblastoma). *Cancer Genet Cytogenet* 2004; 149: 161-163. PMID: 15036892
- Hasegawa T, Shimoda T, Hirohashi S, et al. Collagenous fibroma (desmoplastic fibroblastoma): report of four cases and review of the literature. *Arch Pathol Lab Med* 1998; 122: 455-460. PMID: 9593348
- Stacy RC, Jakobiec FA, Fay A. Collagenous Fibroma (Desmoplastic Fibroblastoma) of the Orbital Rim. *Ophthal Plast Reconstr Surg* 2013 Feb 26. PMID: 23446303
- Nonaka CF, Carvalho Mde V, de Moraes M, de Medeiros AM, Freitas Rde A. Desmoplastic fibroblastoma (collagenous fibroma) of the tongue. *J Cutan Pathol* 2010; 37: 911-914. PMID: 19909389
- Ahn M, Osipov V, Harris GJ. Collagenous fibroma (desmoplastic fibroblastoma) of the lacrimal gland. *Ophthal Plast Reconstr Surg* 2009; 25: 250-252. PMID: 19454950
- De Sousa SF, Caldeira PC, Grossmann SMC, De Aguiar MCF, Mesquita RA. Desmoplastic fibroblastoma (collagenous fibroma): A case identified in the buccal mucosa. *Head and Neck Pathol* 2011; 5: 175-179. PMID: 21221866
- Bhagalia S, Jain M, Pardhe N, Sireesha SK. Collagenous fibroma (desmoplastic fibroblastoma) of the oral cavity. *J Oral Maxillofac Pathol* 2012; 16: 277-279. PMID: 22923904
- Park JM, Koh JK, Lee MG. Unusual case of desmoplastic fibroblastoma (collagenous fibroma) with ulceration on toe: mimicking diabetic foot ulcer. *J Dermatol* 2012; 39: 420-422. PMID: 22044125
- Junkins-Hopkins JM, Johnson WC. Desmoplastic fibroblastoma. *J Cutan Pathol* 1998; 25: 450-454. PMID: 9826172

25. Watanabe H, Ishida Y, Nagashima K, et al. Desmoplastic fibroblastoma (collagenous fibroma). *J Dermatol* 2008; 35: 93-97. PMID: 18271804
26. Weisberg NK, DiCaudo DJ, Meland NB. Collagenous fibroma (desmoplastic fibroblastoma). *J Am Acad Dermatol* 1999; 41: 292-294. PMID: 10426912
27. Nishio J, Akiho S, Iwasaki H, Naito M. Translocation t(2;11) is characteristic of collagenous fibroma (desmoplastic fibroblastoma). *Cancer Genet* 2011; 204: 569-571. PMID: 22137488
28. Maghari A, Ma N, Aisner S, et al. Collagenous fibroma (desmoplastic fibroblastoma) with a new translocation involving 11q12: a case report. *Cancer Genet Cytogenet* 2009; 192: 73-75. PMID: 19596257
29. Bridge JA, Swarts SJ, Buresh C, et al. Trisomies 8 and 20 characterize a subgroup of benign fibrous lesions arising in both soft tissue and bone. *Am J Pathol* 1999; 154: 729-733. PMID: 10079250
30. Evans H, Bridge JA. Nodular fasciitis. In: *Pathology and Genetics of Tumours of Soft Tissue and Bone*. Fletcher CDM, Unni KK, Mertens F, editors. WHO Classification of Tumours. Lyon: IARC Press, 2002, pp. 48e49.