CELLULAR FIBROUS DERMATOFIBROMA OF THE SOLE

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CELLULAR FIBROUS DERMATOFIBROMA OF THE SOLE

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Dermatofibroma (syn. histiocytoma) is a common tumor of dermal connective tissue. In most cases classical clinical presentation, dermoscopy, and medical history allow a straightforward clinical diagnosis. Surgical removal is rarely necessary. There are, however, distinct variants of dermatofibroma which need histopathology and immunohistology to confirm diagnosis [1,2].

One of these variants is cellular fibrous dermatofibroma (CFD) presenting less than 5% of cutaneous dermatofibromas in histopathological files. This tumor is highly cellular, composed of sheets of eosinophilic, storiform and fascicularly arranged spindle cells, sclerotic collagen fibers and a mixed infiltrate of macrophages and lymphocytes at the tumor periphery. Its typical localization is within the

dermis, although it can frequently infiltrate subcutaneous tissue. It has a propensity for localization on the extremities without any clear gender predominance. Relapses of CFD are common [3-5]. CFD needs a careful differentiation from soft tissue malignancies such as dermatofibrosarcoma protuberans, leiomyosarcoma, clear cell sarcoma and malignant dermatofibroma, and from amelanotic melanoma [6].

Material and methods. Case Report

A 61-year-old female presented with a slow-growing, firm, slightly painful tumor on the right sole. On examination we found a subcutaneous tumor with hyperkeratosis and lip formation to the neighboring skin (Fig. 1). Laboratory findings: The routine lab was unremarkable except of a slight increase of C-reactive protein (6.8 mg/l; normal range <5 mg/l).

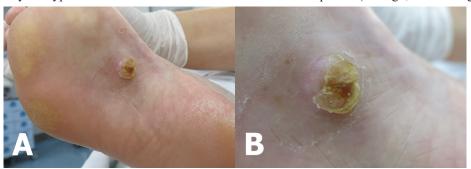


Fig. 1. Hyperkeratotic nodule on the sole

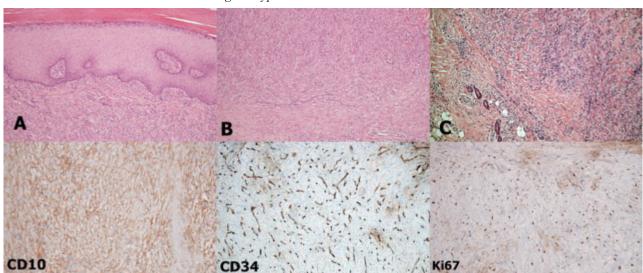


Fig. 2. Histology and immunohistology of CFD. Upper row: Hematoxylin-eosin stains (x10).

(a) Superficial aspect with hyperkeratosis and akanthosis of the epidermis.

(b) Mid-dermis with storiform growth pattern of spindle shaped tumor cells. (c) Deep dermal part.

Lower row: Immunoperoxidase stains (x10). From left to right: CD10 expression,

CD34 expression, Ki67 stain for proliferating cells

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Histopathology: A hyperplastic orthokeratotic epidermis covering a nodular encapsulated tumor composed of spindle and some epithelioid cells in a storiform growth pattern, reaching in depth to the plantar aponeurosis (Fig. 2 A-C). Tumor cells show several mitoses with a slight increase of the mitotic activity at the tumor margins (Fig. 2, Ki67). Atypical mitoses are absent. Entrapping of collagen fibers is evident. There is a rich vascularization as demonstrated by CD34-staining of capillary endothelium. Tumor cells express CD10, focally smooth muscle antigen and desmin, however negative for S100 protein and CD34.

The tumor was removed surgically by slow Mohs surgery within 2 cm negative margin and resection of the muscle fascia. The defect was closed by full thickness skin graft (Fig. 3).



Fig. 3. Full-thickness skin graft of the plantar defect (5 days post-surgery)

Results and their discussion. CFD is a rare variant of dermatofibroma with clinical and histopathological peculiarities, and a variable prognosis [3,4]. CFD is composed of eosinophilic spindle cells without prominent cellular atypia. Mitoses can be found. The tumor expresses vimentin, epithelial membrane antigen, factor XIIIa, focally smooth muscle actin and desmin. Minor expression of CD34, mostly in the periphery, has also been reported. CFD is negative for S100 protein, cytokeratin, and H-caldesmon [3,5,7-9]. The tumor tends to relapse locally after R1-resection but rare cases with metastases have also been observed [10]. The time between diagnosis and metastasis varied in one study between 0 to 180 months, with lung, lymph nodes and soft tissue as the most common involved sites [11]. Histopathology of the primary tumor is not indicative for the risk of metastasis.

The localization on the sole as described in this report is quite uncommon [10]. A deep penetrating benign fibrous dermatofibroma is a possible differential diagnosis. Sarcomas such as dermatofibrosarcoma protuberans, leiomyosarcoma, epithelioid sarcoma, and malignant fibrous dermatofibroma and amelanotic melanoma should be ruled out by histopathologycal and immunohistochemistry evaluation [12,13]. An overlap expression of CD34 and factor

XIIIa has been reported in both CFD and dermatofibrosarcoma protuberans [14]. In addition, CD163 (receptor of hemoglobin oxygen scavenger) and tyrosine kinase ALK (anaplastic lymphoma kinase) have been described as more specific markers for CFD [15-17].

CFD is a rare connective tissue tumor with a propensity to relapse and spread. Therefore, three-dimensional histopathological analysis is necessary to ensure a complete excision. CFD on the sole is a very rare observation.

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SUMMARY

CELLULAR FIBROUS DERMATOFIBROMA OF THE SOLE

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Cellular fibrous dermatofibroma is a rare variant of dermatofibroma/ histiocytoma. We present a 61 years old female with a slow-growing, firm tumor on the sole of her right foot. The tumor was removed by slow Mohs surgery within 2 cm negative margin. Histopathologic investigation revealed a nodular encapsulated tumor composed of spindle and some epithelioid cells in a storiform growth pattern. Minimal mitotic activity was reported, however without evidence of atypical mitoses. Tumor cells expressed CD10, focally smooth muscle antigen and desmin, but remained negative for S100 protein and CD34. The diagnosis of cellular fibrous dermatofibroma was confirmed. The defect was closed by full thickness skin graft.

Keywords: Benign connective tissue tumors, dermatofibroma, histocytoma, histology, surgery.

РЕЗЮМЕ

КЛИНИЧЕСКИЙ СЛУЧАЙ КЛЕТОЧНОЙ ВОЛОКНИСТОЙ ФИБРОМЫ КОЖНОГО ПОКРОВА ПОДОШВЫ

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Клеточная волокнистая дерматофиброма - редко встречающаяся форма фибромы/гистиоцитомы кожи.

Автором представлен клинический случай медленно растущей, твердой опухоли кожи правой подошвы у 61-летней женщины.

Опухоль была удалена методом замедленной операции Мохса в пределах 2 см здоровой ткани. Гистологическое исследование выявило узловидную осумкованную опухоль, состоящую из шпиндельных и эпителиоидных клеток, организованных в колесоподобной конфигурации. Отмечалось наличие минимальной митотической активности, однако без признаков атипичного митоза. Опухолевые клетки выделяют CD10, гладкомышечный антиген и десмин, но отрицательны на наличие белков S100 и CD34.

Подтвердился диагноз клеточной фиброзной дерматофибромы. Хирургический дефект был закрыт трансплантатом полной толщины кожи.

რეზიუმე

ფეხის გულის კანის უჯრედულ-ბოჭკოვანი ფიბრომის კლინიკური შემთხვევა

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აკადემიური სასწავლო პოსპიტალი დრეზდენფრიდრიხშტატი, ¹დერმატოლოგიის და ალერგოლოგიის დეპარტამენტი; ²პათოლოგიის ინსტიტუტი "გიორგ შმორლი"; ³ანესტეზიოლოგიის და ინტენსიური თერაპიის,გადაუდებელი დახმარების და ტკივილის მკურნალობის დეპარტამენტი; დრეზდენი, გერმანია

უჯრედულ-ბოჭკოვანი დერმატოფიბრომა კანის ფიბრომის/ჰისტიოციტომის ი'შვათი ფორმაა. ავტორის მიერ წარმოდგენილია 61 წლის ქალის მარჯვენა ფეხის გულის კანის ნელა მზარდი, მკვრივი სიმსივნის კლინიკური შემთხვევა. სიმსივნე მოკვეთილ იქნა მოხსის შენელებული ოპერაციის მეთოდით ჯანმრთელი ქსოვილის 2 სმ-ის ფარგლებში. პისტოლოგიური კვლევით დადგინდა კვანძოვანი, შემოფარგლულგარსიანი სიმსივნე, შემდგარი ბორბლისებრ კონფიგურაციად ორგანიზებული თითისტარისებრი და ეპითელოიდური უჯრედებისაგან. აღინიშნებოდა მინიმალური მიტოზური აქტივობა, ატიპური მიტოზის ნიშნების გარეშე. სიმსივნური უჯრედები გამოყოფენ CD10, გლუვკუნთოვან ანტიგენს და დესმინს,მაგრამ ნეგატიურნი არიან S100 და CD34 ცილების არსებობის მიმართ.

დადასტურდა უჯრედული ფიბროზული დერმატოფიბრომის დიაგნოზი. ქირურგიული დეფექტი დაიფარა სრული სისქის კანის ტრანსპლანტანტით.

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