Clear Cell Dermatofibroma on the Chest Wall: A Case Report and Its Diagnostic Traps

Abstract

Dermatofibromas are common lesions of the skin. Although they occur at any part of the body, they are most commonly observed on the lower legs of middle-aged women. The lesion comprises fibroblast-like cells, histiocytes, collagenous tissue, and blood vessels. Many histological variants have been defined based on the ratio of cell components and their location. These variants of dermatofibroma may cause problems during differential diagnosis between benign and malignant mesenchymal lesions of the skin and may lead pathologists to overdiagnose this lesion. Here, we report a case of clear cell dermatofibroma, which is a rare variant of dermatofibroma, together with its diagnostic traps.

Keywords: *Clear cell dermatofibroma, clear cell lesions, diagnostic traps, differential diagnosis*

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Introduction

Dermatofibroma (DF) is the most common soft tissue lesion of the skin. It accounts for 3% of the total skin biopsies submitted to each pathological laboratory.[1] Although it can occur at any part of the body, it is most commonly observed on the lower legs of middle-aged women.[2] Despite its benign nature and being considered as an ordinary lesion of the skin by dermatologists at the first glance, pathological diagnosis may be challenging, particularly for rare variants and may lead pathologists to overdiagnose this lesion.[2] The current report presents a case of clear cell dermatofibroma (CCDF), which is a rare variant of DF.

Case Report

A 33-year-old male patient with a nodular mass on the chest wall was admitted to the hospital. He had no significant systemic disease or history of surgery at this site. Abdominal ultrasonography was unremarkable and there were no masses on kidneys. With the clinical diagnosis of hemangioma, excisional biopsy was performed.

Grossly, a yellowish white, well-demarcated but not encapsulated, nodular lesion of the largest diameter 1 cm was observed in the dermis without necrosis or hemorrhage. Microscopic examination revealed a partially-demarcated nodular lesion in the dermis and a Grenz zone between the epidermis and lesion [Figure 1a]. There was an extension toward subcutaneous tissue, but its deep margins were well-circumscribed. At higher magnification, it was observed that the lesion comprised haphazardly distributed vacuolated to optically clear cells. These cells were round-oval shaped with well-defined cell borders and large nuclei showing single prominent, eosinophilic nucleoli [Figure 1b]. Some cells were spindle shaped as well. Cells were surrounded by reticulin fibers and a variable degree of fine-to-sclerotic collagen [Figure 1b]. The lesion was richly vascularized. Occasional lymphocytes were interspersed among the clear cells. Rare mitoses were seen and Ki-67 index was below 1%.

A standard three-step streptoavidin–biotin complex method was used for immunohistochemical staining, and all markers were obtained from Novocastra Laboratories (Newcastle, England). The infiltrate was negative for most epithelial and mesenchymal markers, including HMB-45, Melan A, epithelial membrane antigen, pancytokeratin, inhibin, desmin, smooth muscle actin, myoglobin, cytokeratin-8, laminin, calretinin, CD68, CD1a, S-100, renal cell carcinoma (RCC) marker, CD99, CD57, and neurofilament.

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Vimentin and CD10 were diffusely positive [Figure 2], whereas up to 50% of the lesional cells were positive for FXIIIa [Figure 2]. CD34 only highlighted vascular structures.

The histopathologic diagnosis rendered was CCDF. No recurrence was seen 2 years after complete excision.

Discussion

The incidences of histological variants of DF are as follows: common variant, 80%; aneurysmal variant, 5.7%; hemosiderotic variant, 5.7%; epithelioid variant, 2.6%; cellular variant, 2.1%; lipidized variant, 2.1%; atrophic variant, 1.0%; and clear cell variant, 0.5%.[2] CCDF, a rare variant of DF, was described by Zelger *et al*. in 1996.[3] To date, only 14 cases have been reported in the literature, of which four of them were presented as as posters.[4-6]

When the clear cell changes are seen in a lesion of the skin, a high number of benign or malignant lesions should be considered in the differential diagnosis.[7] Based on histopathologic and immunohistochemical features, the main lesions in the differential diagnosis are summarized in Table 1. In the differential diagnosis of epidermal neoplasm, prominent intraepidermal component and/or connection to the epidermis are important indications. In the differential diagnosis of adnexal neoplasms, peripheral palisading of cells and duct-like structures and outer root sheath differentiation are seen; epidermal neoplasms and adnexal neoplasms are positive with epithelial

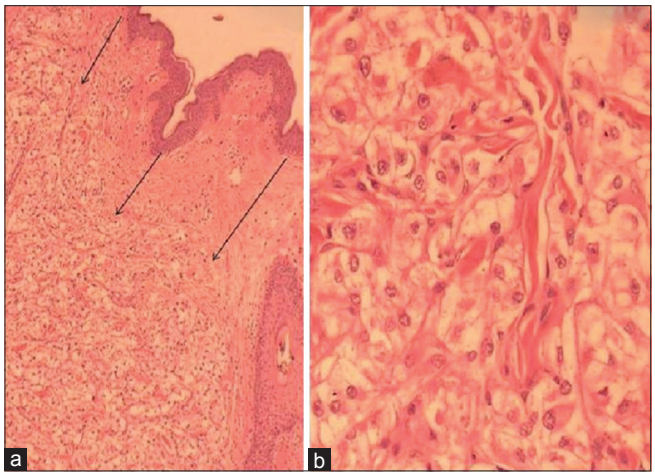

Figure 1: (a) Clear cell lesion situated under the epidermis. Of note, the Grenz zone between the epidermis and lesion (black arrow, H and E, ×20). (b) Optically clear cytoplasm is easily observed in this picture along with haphazardly arranged thick collagen bundles entrapped within the lesion (H and E, ×400)

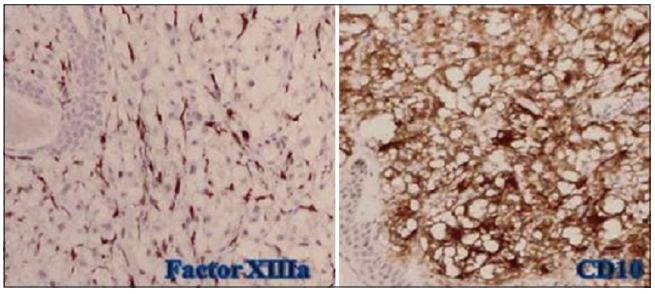

Figure 2: Tumor cells are positive with Factor XIIIa and CD10 (both ×100)

Table 1: Differential diagnosis of clear cell changes in skin lesions

	CCDF	Epithelial neoplasms	Adnexal neoplasms	Mesenchymal/ fibrohistiocytic neoplasms	Melanocytic neoplasms	Metastatic lesions (RCC, CCS, etc.)
Histopathological features						
Clear cell change	+	+	+	+	+	+
Well-demarcated	+	±	±	±	±	-
Prominent intraepidermal component and/or connection to the epidermis	-	+	±	-	-	-
Peripheral palisading of cells	-	±	+	-	-	-
Outer root sheath differentiation	-	-	+	-	-	-
The presence cholesterol clefts and foamy macrophage	-	-	-	+	-	-
Nested pattern	-	-	-	-	+	-
Irregular growth pattern	-	±	±	±	±	+
Necrosis and hemorrhage	-	±	±	±	±	+
Significant cellular atypia and mitosis	-	±	±	±	±	+
Renal mass	-	-	-	-	-	+*
The localization in deeper structures of younger individuals	-	-	-	-	-	+**
Immunohistochemical features						
Epithelial IHC markers (EMA, cytokeratins, CEA etc.)	-	+	+	-	-	+
Mesenchymal and fibrohistiocytic IHC markers (vimentin, desmin, SMA, CD10, FXIIIa, s100, etc.)	+	-	-	+	+	+
Melanocytic IHC markers (HMB45, melan A)	-	-	-	-	+	-

CCDF = Clear cell dermatofibroma; RCC = Renal cell carcinoma; CCS = Clear cell sarcoma; IHC = Immunohistochemical. *It is observed in RCC; **It is observed in clear cell sarcoma

immunohistochemical staining. Myoepithelial lesions and mesenchymal lesions look like CCDF but relevant immunohistochemical staining makes the distinction easy.

Melanocytic lesions generally have a nested appearance and are positive with melanocytic markers (S-100, HMB45). In addition, clear cell sarcoma is found mainly in deeper structures of younger individuals. Morphological analysis reveals fascicles and islands of round spindled clear cells with vesicular nuclei showing the expression of S-100.

RCC is notorious for metastasizing the unusual sites and, although rare, skin metastasis of RCC occur. CD10 positivity, as observed in our case, may be seen in CCDF and represents the major pitfall in the differential diagnosis of clear cell type RCC.[8] However, the growth pattern of metastasis of RCC is irregular, with necrosis, hemorrhages, marked cellular atypia, and mitoses. Unlike RCC metastasis, FXIIIa positivity is the most important distinctive feature of CCDF.

Prolonged sun exposure, traumatic injury, insect bites, and chronic infection have been suggested as causative agents in the pathogenesis of DF.[4] It was claimed that the documented history of trauma was an evidence of its reactive nature.[9] However, these lesions do not regress spontaneously. Therefore, the neoplastic theory cannot be entirely dismissed. In some cases of DF, clonality has been found, without any consistent karyotypic aberrations.[10]

In summary, although the behavior of DF is generally very indolent, CCDF is confused with other entities showing clear cell changes that have malignant course and pathological diagnosis may be challenging. It is important to avoid misinterpretation of this lesion, in particular metastasis of RCC or clear cell sarcoma to the skin.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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