Medallion-like dermal dendrocytoma

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ABSTRACT

Medallion-like dermal dendrocytoma is a benign cutaneous neoplasm that mimics dermatofibrosarcoma protuberans histologically. The distinction between these two entities is critical to prevent unnecessary wide excisions. Herein we describe an acquired MLDD in a 55-year-old female.

Key words: CD34, dendrocytoma, dermal, medallion-like

INTRODUCTION

Medallion-like dermal dendrocytoma (MLDD) is a benign cutaneous neoplasm that mimics dermatofibrosarcoma protuberans (DFSP) histologically. Early descriptions of the neoplasm suggested that it presented as a benign congenital hamartoma, but subsequent reports confirm that it may first appear in adulthood. The lesion usually presents as indurated or atrophic medallion-like plaques that sometimes have a wrinkled surface. Histopathologically MLDD demonstrates a band-like dermal proliferation of spindle cells which strongly express CD34, resembling the plaque portion of DFSP. The distinction between these two entities is critical to prevent unnecessary wide excisions. Herein we describe an acquired MLDD in a 55-year-old female.

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CASE REPORT

A 55-year-old female presented with multiple asymptomatic eruptive lesions over the trunk and extremities of several years duration. Lesions presented as linear brownish and slightly atrophic plaques located parallel to the skin tension lines. The lesions were enlarging (from 3 mm × 3 mm at initial presentation to 3 mm × 10 mm at the time of diagnosis) [Figures 1 and 2]. The patient had no other underlying medical conditions.

Biopsies of two lesions on the trunk and arm demonstrated benign neuromas, but a biopsy from a firmer lesion on the right flank revealed a band-like proliferation of spindle cells. Adnexal structures were preserved within the neoplasm and scattered mast cells were present. Thickened collagen fibers were seen at the periphery of the proliferation and were entrapped by spindle cells, forming onion-like structures [Figures 3-6]. Immunohistochemical stains showed strong CD34 positivity [Figure 7], while S-100, CD68, MelanA and factor XIIIa were negative.

DISCUSSION

MLDD is a benign CD34-positive dermal spindle cell proliferation, important because it may mimic DFSP. It was initially described

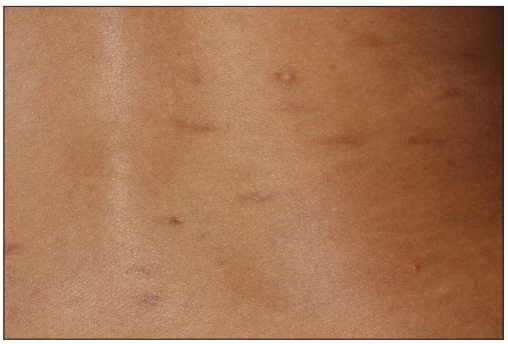

Figure 1: Elongated brownish multiple lesions located along the skin tension lines

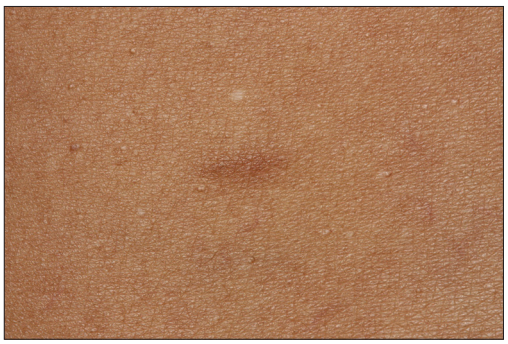

Figure 2: Linear indurated plaque

by Rodríguez-Jurado *et al*. in 2004. These cases presented clinically as solitary, round to elongated, red or brown plaques over the trunk or extremities, measuring 4-15 cm, covered with pliable, atrophic or wrinkled skin.[1] All of the initial cases were present since birth and authors concluded that they represent congenital hamartomas. Histopathologically, the lesions presented as spindle cell proliferations in the reticular dermis positive for CD34 and factor XIIIa.[1] Several congenital CD34-positive tumors, presumably of dendrocytic origin, were reported earlier under different terms with minor variations in their clinical and histopathological picture.[2,3] The first report described a patient with a congenital atrophic plaque on the back with white short hair.[2] Histopathology showed spindle cell proliferation positive for CD34 and factor XIIIa and hair follicles in the papillary dermis.[2] The second report described multiple, congenital, slightly atrophic papules over the face and lower extremities.[3] Histopathologically, these lesions revealed granular cell proliferation positive for CD34 and Factor XIIIa.[3] Acquired cases of MLDD developing in adults were subsequently reported by Kutzner *et al*.[4] Several reports mention that some cases of MLDD are indurated with non-atrophic surface.[4,5] An indurated nodule was described in one of the MLDD cases.[5] A small case series revealed that spindle cells do not always express Factor XIIIa, suggesting that not all examples have a common pathogenesis.[4,6] The descriptive term "medallion-like dermal dendrocyte hamartoma" was proposed by Kutzner *et al*., and both names are currently in use.[4]

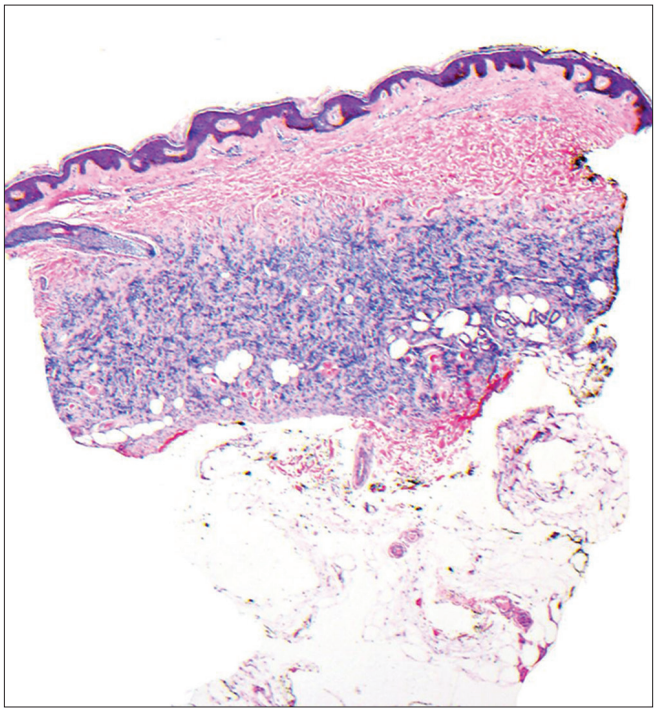

Figure 3: Band-like dermal infiltrate of spindle cells in the reticular dermis, extending in to subcutis, and sparing the papillary dermis. H and E, ×100

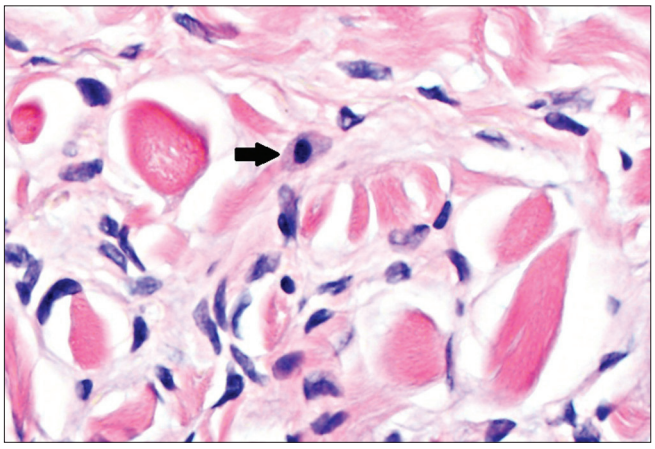

Figure 5: Thickened collagen fibers at the periphery and mast cells (arrow). H and E, ×600, cropped image

MLDD should be differentiated from other spindle cell neoplasms such as dermatomyofibroma, diffuse neurofibroma, digital fibromyxoma, dermatofibroma, and spindle-cell melanocytic neoplasms. The most important differential diagnosis, though,

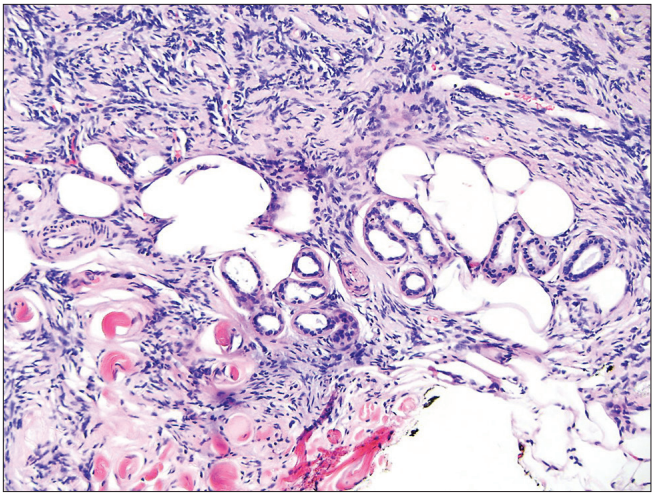

Figure 4: Thickened entrapped collagen fibers, and uninvolved adnexal structures. H and E, ×400

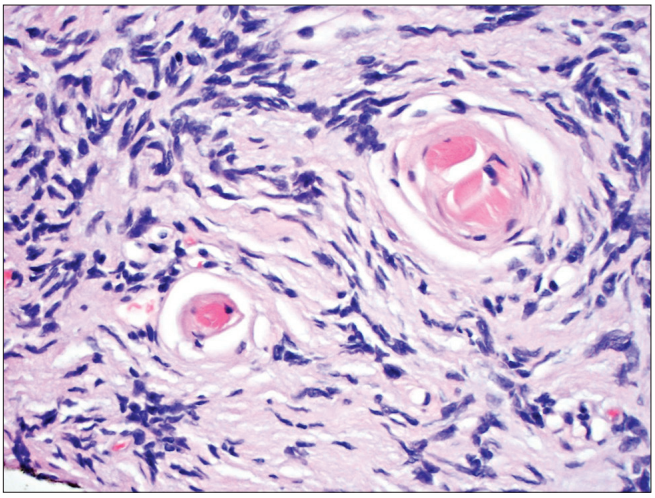

Figure 6: Concentric layers around the entrapped collagen fibers mimicking onion skin. H and E, ×600

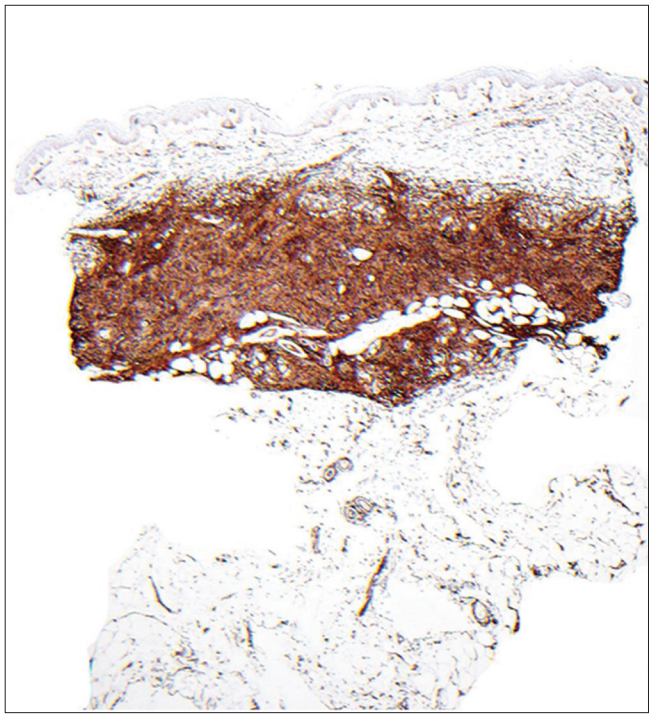

Figure 7: Strong CD 34 positivity in the neoplasm. CD34-stained sections, ×100

is DFSP, in particular its congenital form. This distinction is important to prevent wide re-excisions of the tumor resulting in disfigurement.

Though both MLDD and DFSP show dermal proliferation of spindle cells, MLDD demonstrates a band-like pattern that often does not destroy or displace adnexae, vessels and nerves and does not involve the papillary dermis. Thickened collagen bundles and multiple mast cells may be present.[4] The epidermis and reticular dermis of MLDD are often atrophic. Other described histopathological features of MLDD include widened venules in the upper dermis, vertical orientation of the cells in the upper portions of the proliferation and horizontal orientation in the lower ones, as well as presence of pushed elastic fibers at the bottom of the proliferation.[4] This contrasts with dermatomyofibroma which demonstrates thickened elastic fibers throughout the neoplasm.

Immunohistopathology is of little use in the differentiation of MLDD from DFSP as CD34 is strongly positive in both of these spindle cell neoplasms. MLDD shows variable expression of factor XIIIa and is negative for melanocytic markers (S100, MelanA, HMB45). Our patient's lesion was also negative for CD68. While the size of the lesion and a stable clinical course are suggestive of the diagnosis, molecular techniques can be used to help exclude DFSP in difficult cases.[4]

The eruption of multiple simultaneous skin lesions following the skin lines and association with eruptive neuromas in our patient expands the spectrum of dermal CD34-positive benign neoplasms that represent a group of clinically diverse lesions with unifying histopathological features and a benign course.

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