CASE REPORT

RJME
Romanian Journal of Morphology & Embryology
http://www.rjme.ro/

Dermal plexiform spindle cell lipoma

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Abstract

Spindle cell lipoma located in the dermis is uncommon. The plexiform variant of this tumor is rare. In fact, only six cases of this variant have been described previously. We report herein a case of dermal plexiform spindle cell lipoma with prominent myxoid matrix. A 47-year-old male patient presented with a solitary, 2.2 cm-cutaneous mass in the right buttock region that had slowly increased in size for over one year. The dermal lesion was characterized by a mixture of mature adipocytes, spindle shaped cells and inconspicuous ropey collagen bundles in a mucinous background. This lesion showed a fascicular and plexiform pattern with adipocytes irregularly arranged, predominant in the depth of the lesion. Immunohistochemically, the spindle cells were positive for CD34, factor XIIIa, and vimentin, and negative for retinoblastoma protein, claudin-1, GLUT-1, epithelial membrane antigen, neurofilament protein, and Sox-10. S100 protein stained a thinned cytoplasmic rim of mature adipocytes and labeled about 25% of spindle cells in the most superficial areas. A review of the seven cases published, including the present report, revealed that there were five females and two males. Most cases located in the thigh-groin-buttock area. The age of the patients ranged from 32 to 58 years with a mean of 45.7 years. Clinical diagnosis suggested a lipomatous or neural tumor in six cases. The main differential diagnosis includes dermal intraneural plexiform neurofibroma and purely intradermal monophasic plexiform spindle cell nevus.

Keywords: spindle cell lipoma, plexiform spindle cell lipoma, plexiform neurofibroma, plexiform spindle cell nevus, skin, dermis.

Introduction

Spindle cell lipoma is a relatively uncommon benign, distinctive lipocytic tumor, constituting approximately 1.5% of all adipocytic neoplasms [1]. Spindle cell lipoma variants can cause diagnostic difficulties due to variation in their constituent elements [2], peculiar location [3], or by virtue of the patterns of growth [4] that differ from the usual spindle cell lipoma.

Spindle cell lipomas have uncommonly been reported to arise in the dermis and may be misdiagnosed in this location. They constitute a distinct subset [4, 5]. Purely cutaneous lesions differ from usual spindle cell lipomas, in that they are unencapsulated, with poorly defined, infiltrative margins; they show female predilection, wider anatomical distribution, and their size is usually less than 2.5 cm [5]. These dermal tumors may show prominent myxoid changes [5] and rarely a plexiform way of growth. In fact, only six cases of the plexiform variant of dermal spindle cell lipoma have been reported [4].

In this report, we describe a case of dermal spindle cell lipoma characterized by a mucinous stroma and a fascicular and plexiform pattern of growth. This type of tumor is under-illustrated and not well recognized among pathologists and dermatopathologists. We justify the publication of this case because of its rarity, the only reference dates from 1995, and because the morphological and immunohistochemical peculiarities can be misinterpreted as those of a tumor of neural or melanocytic origin.

Case presentation

A 47-year-old man presented with a raised skin nodule of about 2 cm in diameter at his right buttock that had slowly increased in size for more than one year. The patient had no history of any other disease. Clinical examination revealed a nondescript, painless, soft nodule with ill-defined margins, non-adhesive to deep layers, in the skin of the buttock. Clinically, it was thought a cutaneous cystic lesion. The patient was in good overall health without any other cutaneous nodules or significant spots.

Surgical excision of the nodular mass was performed under local anesthesia. The excised specimen measured 2.2×1.1×1.1 cm. The cut surface showed a vaguely nodular, white-yellow ill-defined cutaneous lesion.

Histopathological examination revealed no epidermal lesions.

The dermis showed fascicles constituted by dispersed tumor cells enmeshed in abundant myxoid matrix (Figure 1). These fascicles consisted of loosely arranged, delicate spindle-shaped cells with a small amount of cytoplasm and fusiform nuclei (Figure 2A); scattered single or in small clusters, mature adipocytes irregularly arranged along the lesion (Figure 2B); inconspicuous collagen bundles (Figure 3); small vessels; and mast cells interspersed in the myxoid stroma. The adipocytes were more abundant in the depth of the lesion. The spindle cells were haphazardly arranged (Figure 3). The fascicles were separated by normal dermal collagen. The lesion was centered in the dermis and extended to the neighboring hypodermis.

Immunohistochemistry reaction was performed using tissue controls positive for the antibodies utilized. Internal controls were also scrutinized. For the retinoblastoma protein (RBBP-6), human testis sections were used as controls. Immunohistochemically, the spindle cells were diffusely positive for CD34 (Figure 4) and vimentin, widely positive for factor XIIIa (Figure 5A), and negative for

retinoblastoma protein, claudin-1, GLUT-1, epithelial membrane antigen, neurofilament protein, and Sox-10. S100 protein stained a thinned cytoplasmic rim of mature adipocytes and labeled about 25% of spindle cells in the most superficial areas (Figure 5B). Scattered CD117+ mast cells were present in the fascicles.

Antibodies used in the immunohistochemical study are detailed in Table 1.

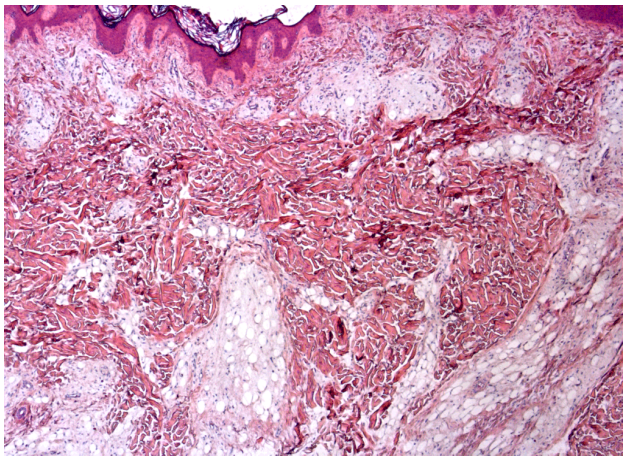

Figure 1 – *Panoramic view of the plexiform spindle cell lipoma. Multiple dermal myxoid fascicles show irregular distribution of mature adipocytes [Hematoxylin–Eosin (HE) staining, ×25].*

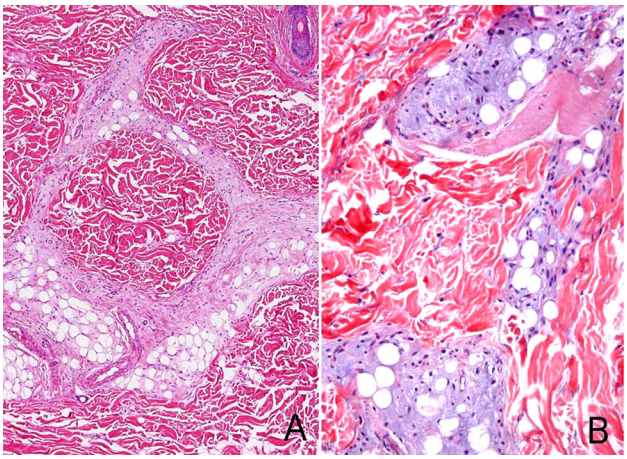

Figure 2 – *Plexiform spindle cell lipoma: (A) Dermal myxoid fascicles show scattered single mature adipocytes and scant spindle cells (HE staining, ×100); (B) Fascicles showing prominent myxoid matrix (HE staining, ×100).*

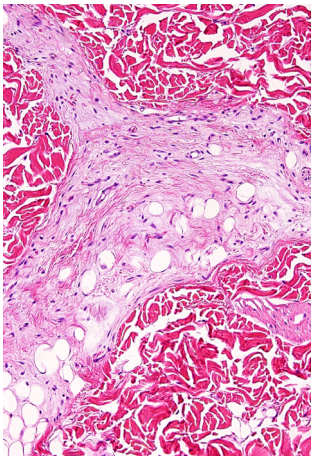

Figure 3 – *Detail of a fascicle showing mature adipocytes arranged individually, spindle cells, and scant collagen fibers in a myxoid matrix. Spindle cells do not show parallel arrangement (HE staining, ×200).*

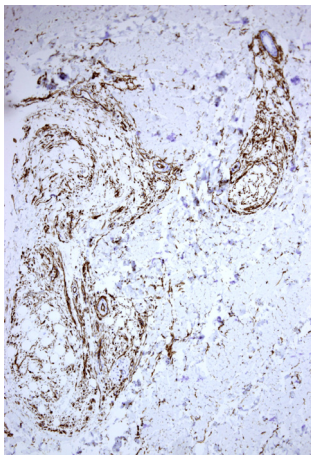

Figure 4 – *Immunohistochemistry of the dermal plexiform spindle cell lipoma. Diffuse strong positivity in spindle cells for CD34 can be observed within the fascicles (×100).*

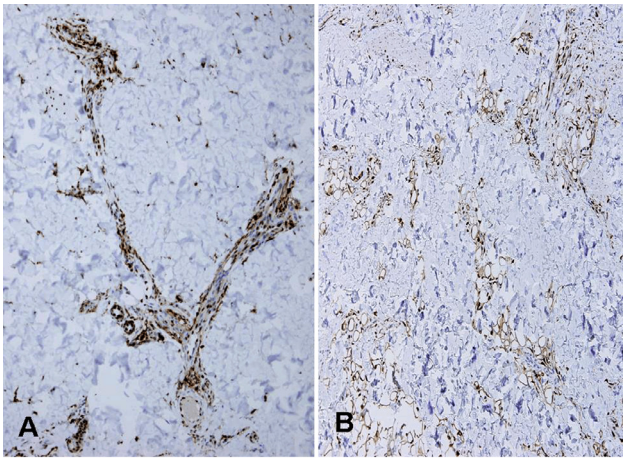

Figure 5 – *Immunohistochemistry of the dermal plexiform spindle cell lipoma: (A) Fascicles displaying positive staining for factor XIIIa (×100); (B) Dermal fascicles showing reactivity, in adipocytes and some spindle cells, for S100 protein (×100).*

Table 1 – *Antibodies used in this study*

Antibody	Source	Clone	Dilution	Retrieval solution pH (Dako)
CD34	Dako	QBEnd 10	FLEX RTU	High
Vimentin	Dako	V9	FLEX RTU	High
Claudin-1	Abcam	Polyclonal	1:200	Low
GLUT-1	GeneTex	Polyclonal	1:50	Low
EMA	Dako	E29/EP1	FLEX RTU	High
Sox-10	Biocare Medical	BC34	1:100	High
S100 protein	Dako	Polyclonal	FLEX RTU	High
CD117	Dako	Polyclonal	1:200	High
RBBP-6	Abcam	Polyclonal	1:100	High
Neurofilament protein	Dako	2F11	FLEX RTU	High
Factor XIIIa	Thermo Scientific	AC-1A1	Prediluted	High

EMA: Epithelial membrane antigen; RBBP-6: Retinoblastoma binding protein-6; Dako, Glostrup, Demark; Abcam, Cambridge, UK; GeneTex, Irvine, CA, USA; Biocare Medical, Concord, CA, USA; Thermo Scientific, Waltham, MA, USA; RTU: Ready-to-use.

Discussion

The lesion we are reporting exhibited features that fulfilled histopathological and immunohistochemical criteria for the diagnosis of dermal plexiform spindle cell lipoma. Two peculiar features in our lesion were

remarkable such as: (*i*) it arose in the dermis and (*ii*) it showed a plexiform pattern of growth.

Spindle cell lipomas are uncommonly reported in the dermis. In fact, dermal location is observed in only 9.8% of spindle cell lipomas [1]. Purely cutaneous tumors have some unique clinicopathologic features. They occur more often in female patients, and they have a wider anatomic distribution than usual spindle cell lipoma, with involvement of the lower trunk and the extremities, including the fingers. Interestingly, most cases are poorly circumscribed and have an infiltrative growth pattern [4, 5]. Taking into account the infiltrative growth, the presence of lipoblasts in some cases [5], and the prominent myxoid changes [5], they can mimic an atypical lipomatous tumor or a myxoid liposarcoma.

The plexiform variant of dermal spindle cell lipoma is rare. In fact, only six cases have been previously reported [4]. A review of the seven cases published, including the present report, revealed that there were five females and two males. Six cases located in the thigh-groin-buttock area and one case in the neck. The age of the patients ranged from 32 to 58 years with a mean of 45.7 years. Clinical diagnosis suggested a lipomatous or neural tumor in six cases.

Although spindle cell adenolipomas may show a plexiform pattern they are categorized separately [6–8]. Thus, they are not included in the present review.

The main differential diagnosis includes dermal plexiform intraneural neurofibroma (PIN) and purely intradermal monophasic plexiform spindle cell melanocytic tumor. PIN may show abundant mucinous endoneural matrix that can obscure the Schwann cell component. These lesions show endoneural dominance and qualify as mucinous PIN [9]. On the other hand, a PIN may contain lipoblast-like signet ring cells reactive for S100 protein [10]. Furthermore, plexiform spindle cell lipoma and PIN display an overlapping immunoprofile including S100 protein (in adipocytes and some spindle cells) [4], CD34 (in spindle cells) [11], and factor XIIIa (in spindle cells) [12, 13] immunoreactivity. Thus, spindle cell lipoma may show S100 protein reactivity in 10% to 20% of spindle cells [4]; neurofibromas may display positivity for factor XIIIa in 30% to 70% of cells [13]; and spindle cell lipomas may show reactivity for factor XIIIa in 30% to 40% of spindle cells [12]. However, in PIN, the positive staining for other common neural markers such as collagen IV, claudin-1, epithelial membrane antigen, GLUT-1, Sox-10, and neurofilament protein argues against the diagnosis of plexiform spindle cell lipoma. Moreover, spindle cell lipomas show deficient nuclear expression of retinoblastoma protein by immunohistochemistry [14]. The basis of this loss stems from evidence that spindle cell lipomas are characterized by deletions of chromosome 13q, often with loss of 16q [15]. The RB1 gene is located within the deleted region of chromosome 13q. Loss of nuclear expression of retinoblastoma protein is a sensitive and specific marker for the family of tumors composed by mammary-type myofibroblastoma, cellular angiofibroma and spindle/pleomorphic lipoma [14]. Differentiation between PIN and plexiform spindle cell lipoma is crucial because the former tumor occurs commonly in patients with type 1 neurofibromatosis and it is significantly prone to undergo malignant change [16]. The plexiform spindle cell nevus may be monophasic or biphasic, associated with an overlying or adjacent conventional nevus. The lesion shows granular melanin pigmentation and diffuse immunoreactivity for MART-1 and HMB-45 [17].

Conclusions

Dermal plexiform spindle cell lipoma represents a very uncommon, distinctive, benign tumor of the skin that mostly occurs in the thigh-groin-buttock area and it is predominant in middle-aged women. The tumor shows a partial overlapping immunoprofile with dermal PIN. Deficient nuclear expression of retinoblastoma protein by immunohistochemistry has been shown to be a sensitive and specific marker for spindle cell lipoma in this particular differential diagnostic setting. A complete panel of neural markers can also be helpful in the differential diagnosis with dermal PIN.

Conflict of interests

The authors declare they have no conflict of interests.

Consent

Written informed consent was obtained from the patient for publication of this case report and all accompanying images.

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Received: March 3, 2016

Accepted: August 7, 2016