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Case Report

Subcutaneous spindle cell/ pleomorphic lipoma: A case report of a peculiar tumor entity

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Abstract Spindle cell/ pleomorphic lipoma (SCL/PL) is a rare histologic subtype of lipoma. It occurs in a characteristic clinical settings, arising mainly in middle-aged and older men in subcutaneous tissue of the posterior neck, upper back, and shoulders (shawl region). In women, it much more comonly develops outside the typical shawl location. The author describes a 66-year-old man with a subcutaneous tumor mass in the left back side of the neck. It consisted of bland spindle cell population situated in a fibromyxoid stroma containing thick collagen bundles. Scattered pleomorphic and multinucleated floret-like giant cells were also found. The neoplastic cells showed CD34-positive/S100-negative immunophenotype. A diagnosis of a SCL/PL was established. Subcutaneous SCL/PL is an unfamiliar tumor entity to most dermatologists. Despite the worrisome name it is a clearly benign lesion with no tendency to recur after surgical treatment. The present paper is excellent for demonstrating its unique clinical and histomorphological characteristics.

Key words
Lipoma, spindle cells, pleomorphic cells, CD34, S100.

Introduction

Lipomas are the most frequent soft tissue tumor.[1] By far the most common form is a conventional lipoma, but there also exist rarer histological variants, which are diagnostically more difficult.[2,3] Spindle cell lipoma is a morphological and molecularly distinct subtype of lipoma. It was first described in 1975 by Franz M. Enzinger and Dean A. Harvey,[4] who revised a large number of consultation cases of lipomas that differed considerably in their microscopic appearance from ordinary lipoma. They mainly consisted of an intricate mixture of adipocytes and bland spindle cells aligned in parallel bundles which were situated within a mucinous matrix traversed by collagen fibers.[4] Because of their worrisome histomorphology, they were frequently misdiagnosed as liposarcoma. Yet there was no evidence that these tumors behaved in an aggressive manner or had a tendency to recur locally.[4] They originally called this entity a spindle cell lipoma (SCL). Less than a decade later, Barry M. Shmookler and Franz M. Enzinger[5] reported another variant of lipoma characterized by hallmark floret-like multinucleated giant cells, introducing the term pleomorphic lipoma (PL). Despite pleomorphic picture, which also not infrequently led to a misdiagnosis of liposarcoma, follow-up data established a benign clinical behavior.[5] Since their initial identification, both tumors were thought to be closely linked based on similar clinical, morphologic, and genetic features and nowadays, they are considered[2,6] to be the morphological spectrum of the same neoplasm. SCL/PL occurs in a characteristic clinical settings, arising mainly in middle-aged and older men in the subcutaneous tissue of the posterior neck, upper back, and shoulders (shawl region).[2,3,6] In biopsy practice, when a pathologist encounters a tumor with the typical histomorphology in an older male patient at a

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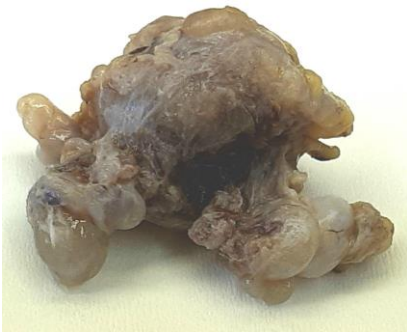

Figure 1 Gross appearance of tumor mass.

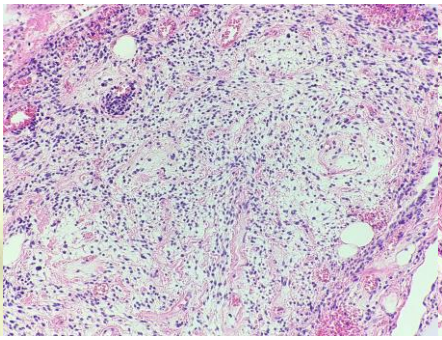

Figure 2 Population of uniform spindle cells in a background of fibromyxoid stroma. (H&E, 10x)

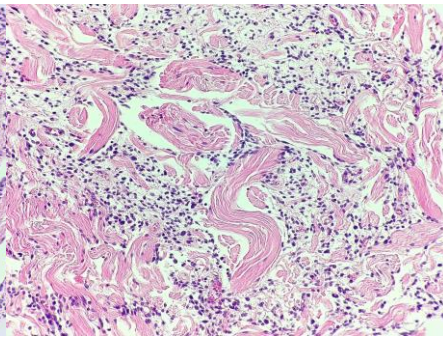

Figure 3 Characteristic ropy collagen bundles among spindle cell population. (H&E, 20x)

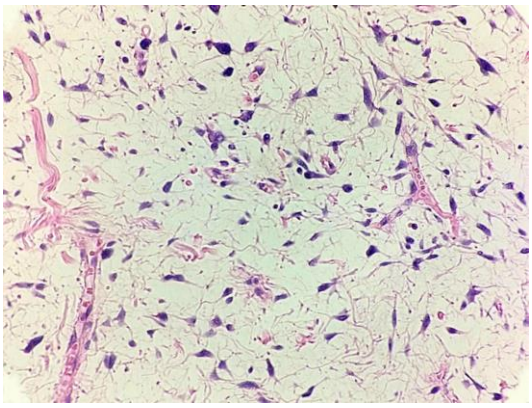

Figure 4 Extensive myxoid stroma containing neoplastic spindle cells with bipolar cytoplasmic processes. (H&E, 40x)

classic site, diagnosis is straightforward. However, a tumor with similar morphologic picture arising in other locations, particularly in women, often triggers concern for other diagnosis. In this article, a rare case of SCL/PL of soft tissue is reported.

Case report

A 66-year-old man presented with a painless subcutaneous tumor mass arising in the left back side of the neck. He claimed that the lesion had been present for a few years without any clinical difficulties. It appeared as a relatively well demarcated, partly mobile resistance in the subcutis covered by intact skin. A presumptive clinical diagnosis was a lipoma. A total surgical extirpation of the lesion was carried out.

The formalin-fixed resected biopsy consisted of an irregularly shaped, lobulated tumor tissue (3x2,5x2,5 mm) containing cystic cavities filled with mucin **(Figure 1)**. It was yellow-brownish in color and elastic in consistency, but some parts were apparently gelatinous on section. The specimen was processed into paraffin-embedded tissue blocks which were stained with hematoxylin-eosin (H&E). Selected sections were then analyzed immunohistochemically with a variety of anti-human antibodies.

The lesion histologically consisted of a non-encapsulated mass of bland spindle cell population and a relatively small amount of mature adipocytes situated in a fibromyxoid stroma **(Figure 2).** The predominant proliferated spindle cells were uniform, with a single elongated nucleus and narrow, bipolar cytoplasmic processes. They were randomly placed or arranged in short parallel fascicles which were interspersed among multiple eosinophilic thick collagen bundles **(Figure 3)**. Some areas were highly myxoid and hypocellular **(Figure 4**). Pseudoangiomatous growth pattern of spindle cell population was also seen, characterized by irregular branching spaces with well formed slit-like connective tissue projections. In addition, an aggregates of scaterred pleomorphic and multinucleated floret-like giant cell were found **(Figure 5).** The latter

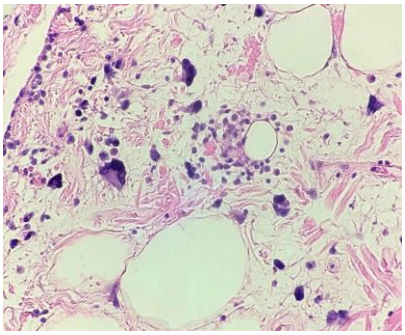

Figure 5 Tumor area with a few pleomorphic and multinucleated floret-like giant cells. (H&E, 40x)

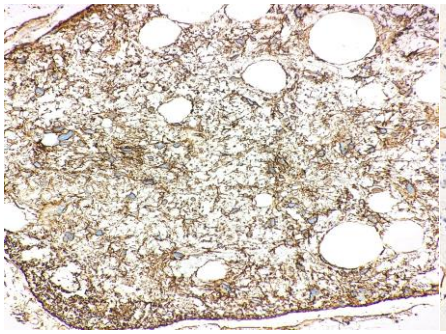

Figure 6 Strong immunoreactivity for CD34 in spindle and pleomorphic cell population. (10x)

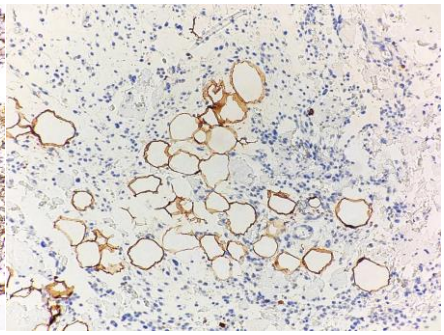

Figure 7 Immunohistochemical negativity for S100 in the spindle cell population, while mature adipocytes are reactive. (20x)

cells displayed a ring of peripherally placed, hyperchromatic nuclei around central eosinophilic cytoplasm (so called flower-petal arrangement). Mitotic figures, necrosis and lipoblasts were absent. The tumor was accompanied by focal stromal lymphocytic infiltration with numerous mast cells (detected by CD117).

By immunohistochemistry, the spindle, pleomorphic, and multinucleated giant cells stained strongly for vimentin, Bcl-2, and CD34 **(Figure 6)**. S100 protein did not mark them, but mature adipocytes showed strong peripheral immunoreactivity **(Figure 7).** Other markers (desmin, polyclonal cytokeratins, STAT-6, CD45/LCA) revealed negative results. Proliferative activity (Ki-67/MIB1) of tumor cells was about 1%. Based on histopathology and immunophenotype, diagnosis of a SCL/PL was established. After sending the final result of the pathologist's report, the author has not had any information about further clinical management of the patient, but this matter was not a goal of this paper.

Discussion

The SCL/PL is a rare and peculiar clinicopathological entity. It accounts for approximately 1.5% of all adipocytic neoplasms.[2] The present case is excellent for demonstrating its unique clinical and morphological characteristics. From a clinical point of view, the most interesting feature of subcutaneous SCL/PL is a different gender prevalence and anatomical distribution between males and females. As noted above, it predominantly occurs in men (the male-to-female ratio is approximately 9:1) and typically arises in the shawl region.[2,6] In the original studies of Enzinger & Harvey[4] and Shmookler & Enzinger[5] this body part comprised about 85% and 78% of all documented SCLs and PLs, respectively. Knowledge of this classical clinical presentation serves as a valuable aid in the final diagnosis. However, this has been shown to be characteristic only for male individuals. In a recent analysis[7] of 384 SCLs the shawl region comprised 71% of cases in men while only 32% of cases in women. The most common affected site in female patients was the upper extremity, followed by the face. In addition, the median age at presentation in women was a decade younger than that seen in men (53 vs. 64 years). The biological mechanisms which determine these differences have been unknown. The contribution of sex-specific hormones could be considered, but the results of a previous study[8] did not suggest that androgen receptor reactivity alone in tumor cells could explain the marked male predominance of SCL.

From a histopathological point of view it is noteable that the morphologic variability observed in this entity is striking. Classic SCL basicaly shows the three structural components: bland spindle cells, ropy-like collagen, and mature adipose tissue.[2,3,6,9-11] However, depending on the proportion of the constituent elements, several histologic subtypes of SCL have been identified.[2.3,6,9,10] Some tumors contain predominantly mature adipocytes with a paucity of spindled or floret-like cells (fat-rich subtype). On the other hand, fat-poor and fat-free subtypes, as the name suggests, have little or almost no mature adipose tissue. The pseudoangiomatous subtype exhibits many villiform projections that form clefts or pseudovascular fissures. Some SCLs harbor an extensively mucinous stroma (myxoid subtype). The structural diversity along with the fact that SCL/PL may be similar to many other soft tissue tumors, often lead to diagnostic pitfalls in biopsy practice. The most important tumor entities that come into consideration in differential diagnosis include atypical spindle cell/ pleomorphic lipomatous tumor, atypical lipomatous tumor/ well-differentiated liposarcoma, cellular angiofibroma, mammary-type myofibro-blastoma, and solitary fibrous tumor. Their detailed description would go far beyond the scope of this article but the substantial clinicopathological features are summarized in **Table 1**. An immunohistochemistry is usually a necessary tool in distinguishing SCL/PL from other soft tissue tumors.

Table 1. Characteristic clinicopathological features of SCL/PL and other soft tissue tumors considered in differential diagnosis. (summarized from ref.[2,3])

Spindle cell/pleomorphic lipoma
- usually occurs in men in the subcutaneous tissue of the shawl region
- the most common location in women is the upper extremity
- variable proportions of bland spindle cells, ropy-like collagen, and mature adipocytes
- scattered pleomorphic and multinucleated floret-like giant cell
- spindle and pleomorphic cells typically CD34 positive and S100 negative
Atypical spindle cell/pleomorphic lipomatous tumor
- predilection for upper and lower extremities
- unencapsulated with ill-defined tumor margins
- variable proportions of spindle cells with mild to moderate atypia, pleomorphic cells, adipocytes and lipoblasts, generally lacks ropy collagen bundles
- variable expression of CD34, S100 and desmin
Atypical lipomatous tumor/well-differentiated liposarcoma
- various locations, but the most common are proximal extremities and trunk
- usually larger and deep-seated (subfascial) tumor
- spindle cells show pronounced atypia, striking variation in the size of adipocytes, various number of lipoblasts, ropy collagen bundles are not typical
- expression of MDM2 and CDK4
Cellular angiofibroma
- most cases occur in vulvovaginal/inguinoscrotal region
- bland spindle cells with prominent medium-sized hyalinized blood vessels
- strong expression of CD34, variable expression of SMA and desmin
Mammary-type myofibroblastoma
- typically inguinal/groin area
- haphazardly intersecting fascicles of bland spindle cells with interspersed bands of hyalinized collagen
- expression of CD34 and desmin
Solitary fibrous tumor
- spindled or ovoid cells arranged around prominent branching (staghorn) and hyalinized vessels, ropy collagen bundles are not feature
- expression of CD34 and STAT6

As a general rule, the spindle, pleomorphic, and multinucleated floret-like giant cells in SCL/PL show a consistent CD34-positive/S100-negative immunophenotype.[2,3,6] Further, they are strongly reactive for vimentin and androgen receptor, sometimes positive for estrogen receptor, constantly negative for desmin and they exhibit loss of RB (retinoblastoma) expression.[2,3,6-9]

The results of immunohistochemistry, however, should always be considered in conjunction with overall histomorphology of tumor.

Conclusion

Subcutaneous SCL/PL is an unfamiliar tumor entity to most dermatologists. Despite the worrisome name it is a clearly benign lesion with no tendency to recur after surgical treatment. The present paper points out this peculiar neoplasm that may sometimes be encountered in dermatopathological practice.

Consent. The examination of the patient was conducted according to the Declaration of Helsinki principles.

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