

Intramuscular spindle cell lipoma of the thigh: A report of a rare tumor in an extremely rare localization

Spindle cell lipoma of the thigh

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Abstract

Spindle cell lipoma (SCL) is a benign adipocytic tumor that typically occurs on the neck/upper back and is located in the dermis or subcutis. This case report highlights the intramuscular SCLs clinical presentation, imaging, histomorphologic, and immunohistochemical features. Intramuscular SCL is a very rare condition and, because of its unusual features, it can be misdiagnosed as liposarcoma. The exclusion of malignancy is the primary concern because liposarcomas need more definitive treatment with a wide excision in the surrounding normal tissue. Because of the heterogeneity of the magnetic resonance imaging features of SCL, the definition and cure for this lesion remain challenging.

Keywords

Intramuscular lipoma; Thigh; Spindle cell

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Introduction

Spindle cell lipoma (SCL) is a rare benign tumor (1.5% of all adipocytic neoplasms), that was first described by Enzinger in 1975 as a specific type of lipoma [1]. In most cases, neoplasm occurred in the dermis or subcutis of the neck and upper back. Other anatomical sites like the face, wrist, hand, upper arm, and thigh were rare, and intramuscular localization of this tumor is very unusual [2]. Histologically, SCL's are composed of an admixture of adipocytes, greatly uniform spindle cells, having a single elongated nucleus, and narrow bipolar cytoplasmic processes [1, 2]. For the diagnostic marker, CD34 positivity has been used by immunohistochemistry [2]. Despite these pathological characteristics, a definite diagnosis is sometimes difficult because this tumor shares similar morphology with another fatty, spindle cell, or myxoid benign or malignant lesions [3].

In this report, we present an unusual intramuscular SCL located in the left gluteus maximus muscle that was treated with a wide excision.

Case Report

A 50-year-old man with a history of a painless left thigh mass admitted to the Department of Orthopedics and Traumatology. His medical history revealed that the mass emerged about three-years ago and has expanded over the last year. On physical examination, non-tender immobile soft tissue mass was observed. The ultrasonography imaging revealed a round, smoothly restricted, lobulated contoured and heterogeneous hypoechoic solid tumor, almost 18x9 cm in diameter. Magnetic resonance imaging (MRI) demonstrated a multilobulated and septated solid mass in gluteus maximus muscle, approximately 16x14x9 cm in diameter and low intensity on T1 sequences and high intensity on T2 sequences (Figure 1).

A tru-cut biopsy was performed. Histologically, the tumor consisted of a small number of randomly oriented spindle-shaped cells that were dispersed in the myxoid ground. The spindle cells were uniform and atypical mitotic figures, nuclear atypia or necrosis were not determined. Immunohistochemically, spindle cells were negative for smooth muscle actin (SMA), Desmin, H-caldesmosom and muscle-specific actin (MSA).

Detailed immunohistochemical examination or fluorescent in situ hybridization (FISH) analysis could not be performed because the tissue was exhausted after serial section and immunohistochemical staining. Tru-cut biopsy reported spindle cell mesenchymal neoplasm without myogenic differentiation, atypical mitotic figures, nuclear atypia or necrosis, most likely benign, low-grade mesenchymal neoplasm. However, it was considered that the biopsy material may not reflect the entire lesion, since the lipomatous component observed on MRI was not observed in the biopsy material. Wide excision of the lesion is recommended to provide a definite diagnosis.



Figure 1. The MRI sequences demonstrate a multilobulated and septated solid soft tissue mass arising from the gluteus maximus muscle. Axial T1 (a) and T2 (b).

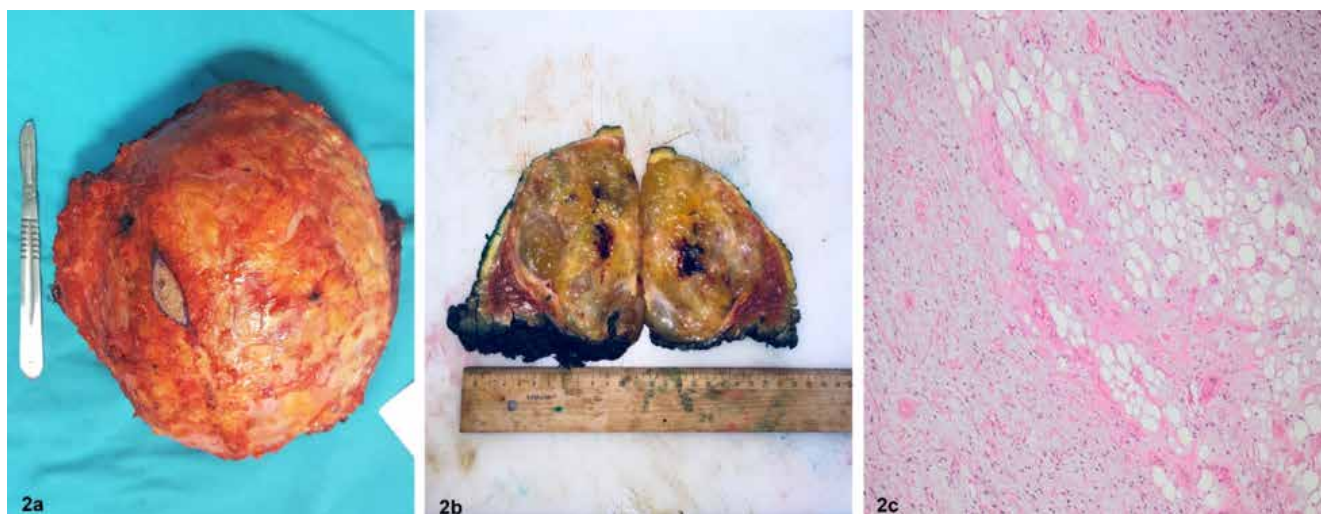


Figure 2. Excised biopsy tract and the tumor as one piece with adequate surgical margins (a). The cut surface of the tumor was yellow-transparent with yellowish fatty and glistening in appearance (b). Microscopic examination of the tumor revealing mature adipocytes interspersed between wavy collagen bundles and spindle cells (H&E, $\times 100$) (c).

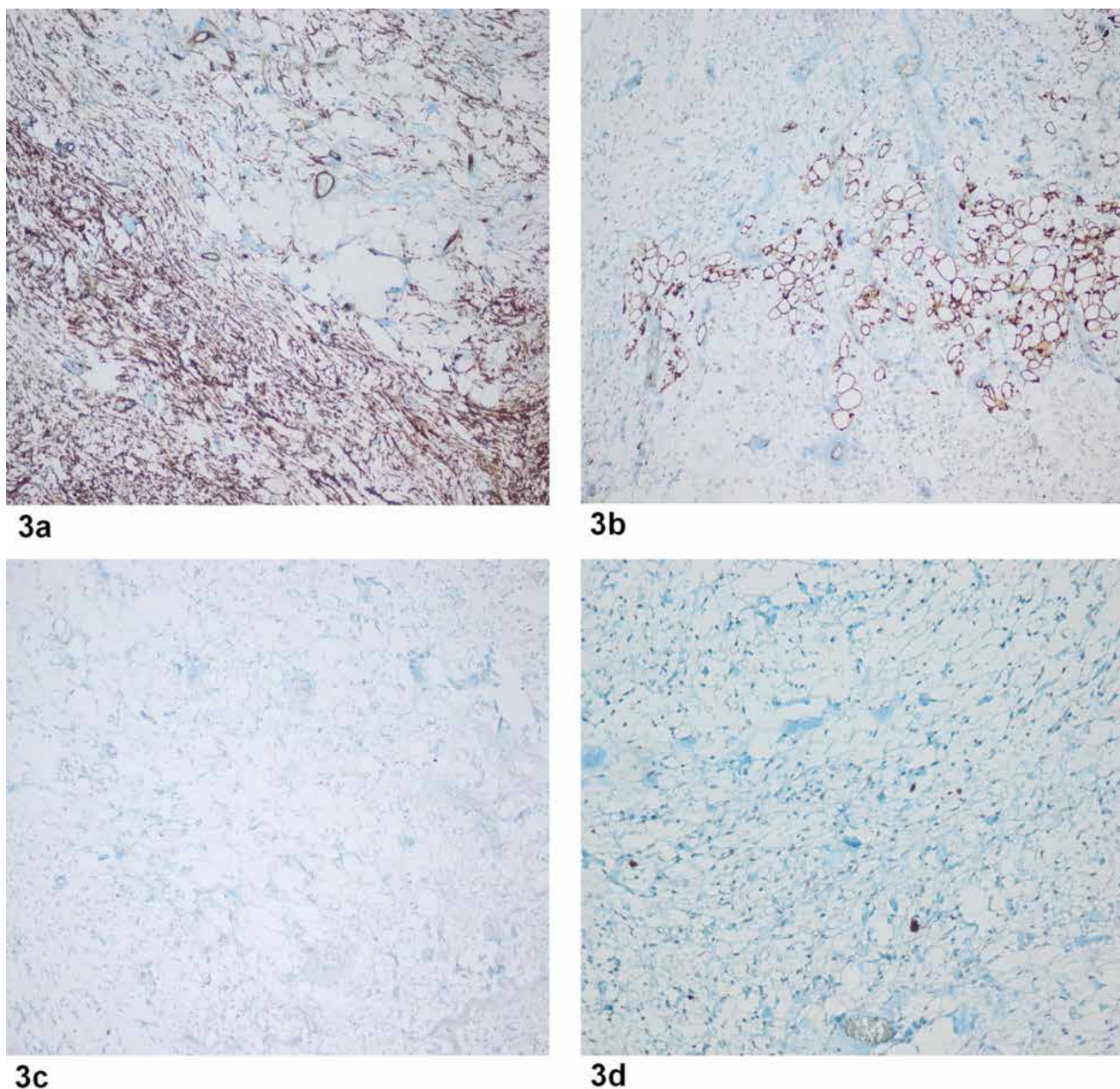


Figure 3. Immunohistochemically, the spindle cells were diffusely and strongly positive for CD34 (a), mature adipocytes were positive for S-100 (b). There was no immunoreactivity for p16 (c). Ki-67 proliferation index was less than 1% (d).

Due to the discrepancy between MRI and biopsy findings, pulmonary and abdominal computerized tomography screenings were performed to rule out metastases, and no mass was detected in any area other than the patient's thigh. Then wide excision was performed with a biopsy tract (Figure 2a). In the course of operation, the entire mass was restricted beneath the fascia and placed within the gluteus maximus muscle. The tumor was excised marginally through the sciatic nerve and the nerve was intact.

Macroscopically, the discoid-shaped mass had a soft consistency and was about 17x10x8 cm in diameter. Cut sections revealed that the tumor was well delineated from the surrounding tissues by a thin capsule and had a heterogeneous yellow-transparent with yellowish fatty and extensive glistening areas on the cut surface (Figure 2b). There was no recognizable gross hemorrhage or necrosis. On the microscopic examination, the tumor was located intramuscularly and separated from the muscle and fat tissue by a thin fibrous capsule. The tumor

consisted of a few eosinophilic collagen fibers and extensive myxoid areas that intermixed with spindle cells and a few mature adipocytes (Figure 2c). The spindle cells were uniform, had a single elongated nucleus and narrow bipolar eosinophilic cytoplasmic processes.

Atypical mitotic figures, nuclear atypia, necrosis, or lipoblasts were not identified. Immunohistochemically, the spindle cells were diffusely and strongly positive for CD34 despite that negative with Mouse double minute 2 (MDM2) and p16. Ki-67 proliferation index was less than 1% (Figure 3a-d). The histopathologic and immunohistochemical features of the tumor suggested a spindle cell lipoma. However, due to the fact that tumor was located in a rare anatomical localization and radiological imaging appearance was suspected for malignancy, we also performed the cytogenetic analysis to exclude atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDL) and myxoid liposarcoma.

FISH analysis was performed using ZytoLight SPEC MDM2/

CEN 12 Dual Color Probe (ZytoVision, Bremerhaven, Germany) to exclude ALT/WDL. Two hundred cells were scored for the MDM2/CEN12 probe set. The cells were captured on the AKAS computer system with a digital camera (Argenit-AKAS Imaging, TURKEY). The MDM2:D12Z3 ratio was 1, average MDM2 signals per cell were 2, 3 and average D12Z3 signals per cell was 2, 3 so MDM2 amplification was not detected. Subsequently, the specimen was examined with ZytoLight SPEC DDIT3 Dual Color, Break Apart Probe for excluding myxoid liposarcoma. (ZytoVision, Bremerhaven, Germany) and rearrangement of the DDIT3 gene region was not identified.

Eventually, with the histopathologic, immunohistochemically, and cytogenetic findings the lesion was reported as an intramuscular spindle cell lipoma. After surgery, the patient was followed for 36 months and underwent an ultrasound examination every 6 months. We did not detect any signs of lesion recurrence.

Discussion

Spindle cell lipomas are benign adipocytic neoplasms, generally located in the subcutaneous tissue, and usually found in men older than 45 years of age [1]. Typically, the lesion consists of bland spindled cells intermixed with mature adipose tissue [1]. The tumor arises in the posterior neck, upper back, shoulders, face, arms, and lower thigh [1, 2].

Magnetic resonance imaging is effective in detecting localization and invasion of the tumors but not always useful for establishing a specific diagnosis. When the lipogenic component of the lesion is abundant and the non-lipogenic components such as spindle cells, collagen fiber, or myxoid matrix are small, MRI findings of the lesion resemble an ordinary lipoma. On the other hand, if the low-fat or fat-free components of the lesion are abundant, and the distribution of the component is heterogeneous, the lesion can differ from a conventional lipoma on the basis of MRI findings; however, in this situation, MRI findings may resemble liposarcoma. In this case, MRI demonstrated a multilobulated and septated solid mass in the gluteus maximus muscle approximately 16x14x9 cm in diameter, and low intensity images on the T1 sequences and high intensity images on the T2 sequences. As a result of this, mesenchymal tumor or sarcoma was anticipated through MRI scans. The final identification was provided by histological studies of the tumor.

Histologically, spindle cell lipomas are characterized by three basic histologic components: greatly uniform spindle cells; mature adipocytes; and the stroma having ropy-like collagen fibers and myxoid matrix [1]. Despite these pathological characteristics, definite diagnosis is difficult because this tumor shares similar morphology with another fatty, spindle cell, or myxoid benign or malignant lesions. Spindle cell lipomas must be distinguished from malignant lesions like liposarcomas because liposarcomas need to be treated with radical excision. In addition to MRI findings, histopathological features of spindle cell lipoma may be evocative of myxoid liposarcoma and ALT/ WDL, especially the spindle cell subtype. Liposarcomas may include variably sized adipocytes and fibromyxoid stroma containing spindle cells with large, deep-staining nuclei and marked nuclear enlargement or pleomorphism [4].

Myxoid liposarcoma shows a more disorderly cellular picture, consisting of multivacuolated lipoblast and plexiform capillary pattern, and CD34 is negative in myxoid liposarcoma [15, 16]. ALT/ WDL spindle cell subtype-atypical spindle cell lipomatous (ASCL) tumor is occasionally located in the subcutaneous tissue and contains spindle cells in short fascicles set in a fibrous or fibromyxoid stroma. This tumor contains an atypical lipogenic cell, lipoblasts are relatively common. Stroma is collagenous but ropy-like collagen fibers are not seen. The spindle cells in atypical spindle cell lipomatous tumors may express CD34, but its expression is often seen in the focal area [5]. Identification of the neoplastic adipocytic component admixed with spindle cells is key for the differential diagnosis between ASCL with SCL.

Cytogenetic analysis has demonstrated that a growing number of soft tissue tumors carry consistent karyotypic abnormalities. ALT/WDL is characterized by amplification of the 12q12-21 (which includes several genes such as MDM2 and CDK4) and 10p11-14 regions. MDM2 amplification can be evaluated by FISH analysis; it is a highly sensitive and specific ALT/WDL diagnostic tool [6]. Although ASCL is a subtype of ALT/WDL, it does not contain MDM2 amplification [7].

Nearly all myxoid liposarcomas are characterized by a reciprocal translocation between chromosomes 12 and 16: t(12; 16)(q13; p11). This molecular event leads to the fusion of the DDIT3 and FUS gene [8]. These findings also provided an important diagnostic tool. In this case, MDM2 gene amplification or chromosome 12 polysomy was not identified, and the rearrangement of the DDIT3 gene region was not identified. Thus, ALT/WDL and myxoid liposarcoma were also excluded by FISH analysis.

In a summary, although the deep location of soft tissue tumors, large size, and imaging heterogeneity are associated with an increased risk of malignancy, benign spindle cell lipoma should be considered in the differential diagnosis in patients with the intramuscular lesion. Histopathological, immunohistochemical, and cytogenetic findings can assist in the differential diagnosis and guide in the correct management of these patients.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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