Case report

Exceptionally large, atypically located spindle cell lipoma

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SUMMARY

Spindle cell lipomas are a rare subtype of lipoma typically occurring in the shawl region of middle-aged men with a characteristic histological appearance consisting of mature adipocytes, ropey collagen and spindle cells. Those of extraordinary size or atypical anatomic location require a proper immunohistological workup to rule out more ominous malignancy. The authors report the rare presentation of a 54-year-old man with a large mass of the left palm causing discomfort and paresthesia for 6 months. Radiographs showed a radiolucent mass without mineralisation or bony involvement. MRI demonstrated a non-contrast-enhancing homogeneous lipomatous mass encompassing flexor tendons of the palm. Elective excisional biopsy and immunohistological analysis were consistent with spindle cell lipoma measuring 11 cm × 7 cm × 4 cm. This case is the largest spindle cell lipoma of the hand reported in the literature and appears to be the largest described in the upper extremity.

BACKGROUND

Spindle cell lipomas (SCLs) are a rare subtype of lipoma. They classically appear as a subcutaneous, slow-growing and painless mass in the shawl region (shoulders, posterior neck and upper back) of men 50–70 years old at an average size of 4–5 cm.^{1,2} In individual cases, SCLs have been reported in atypical locations including the face, thigh, upper arm, forearm, wrist, thumb and very rarely the hand.^{3–7} Because their histology is similar to malignant lipomatous tumours, accurate diagnosis of SCL is important, especially when they appear as larger tumours in atypical locations. We present a case report of a large SCL occurring in the left palm of a

Figure 2 Left hand axial MRI T1 and T2 fat-suppressed images showing lobulated homogeneous palmar mass with a high signal on T1 correlating to a low signal on fat-suppressed imaging.

54-year-old man. This case is the largest SCL of the hand reported in the literature.

CASE PRESENTATION

A 54 year-old Caucasian man presented to our hand clinic with a 6-month history of a slow-growing left palmar mass. The mass was causing moderate discomfort and consistent paresthesia in the palm, ring, long, index fingers and thumb. He also complained of dropping objects. He denied recent weight loss or loss of appetite. He was a retired veteran with a medical history that was unremarkable except for an extensive history of tobacco use and chronic obstructive pulmonary disease. The

Figure 1 Left hand radiographs, anteroposterior, oblique and PA views of a skeletally mature individual showing a radiolucent soft tissue mass of the palm with no mineralisation or bony involvement.

Figure 3 Left hand coronal MRI T1 and T2 fat-suppressed images demonstrating a few low signal bands on T1 and a few high signal bands on T2 sequences. No peritumoural oedema.

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Figure 4 Left hand axial and coronal MRI postcontrast series cuts without enhancement.

physical examination was remarkable for a roughly 4×4 cm firm, mobile mass of the palm extending into the thenar space. There were no open wounds or erythema. He had nearly full active and passive range of motion of all fingers and thumb. He had 4 out of 5 strength with hand grip and thumb opposition. Sensory examination of the left hand was remarkable for diminished sensation in the palm and volar aspect of the first four digits.

INVESTIGATIONS

Radiographs of the hand showed a radiolucent soft tissue mass of the palm with no mineralisation or bony involvement (figure 1). There were no fractures, dislocation or bone lesions. MRI of the left hand with and without contrast demonstrated an approximately 7×4 cm mass in the palm of the left hand encompassing the flexor tendons of the palm and extending into the thenar space. The mass was lobular with homogeneous, high signal on T1-weighted imaging that correlated with low signal on T2-weight fat-suppressed imaging (figure 2). There were a few low signal bands on T1-weighted images. The mass had a

Figure 5 Left hand in the operating theatre. Volar approach was used with Bruner style incision centred over the mass. A common palmar branch of the median nerve tented over the mass. Extension of the mass into the hypothenar/thenar spaces.

Figure 6 Multilobular lipomatous yellow and yellow-grey encapsulated mass.

homogeneous, low signal on T2-weighted imaging with a few high signal bands (figure 3). There was no enhancement of the mass on postcontrast imaging (figure 4). There was no peritumoral oedema. The radiographs and MRI findings were consistent with a benign, lipomatous soft tissue mass of the left hand.

DIFFERENTIAL DIAGNOSIS

The extended differential diagnosis of SCL includes atypical lipomatous tumours (ALT), liposarcoma, angiomyxolipoma, dendritic fibromyxolipoma, solitary fibrous tumour, mammary-type myofibroblastoma and, within the hand, giant cell tumour of a tendon sheath. Corresponding T1 and T2-weighted MRI sequences assist in placing this tumour in the lipomatous family, and immunohistological analysis is key to determining the correct definitive diagnosis. SCLs demonstrate strong, diffuse CD34 expression and vimentin-positive staining within the spindle cell cytoplasm. The absence of lipoblasts and lack

Figure 7 Final specimen approximately 11 cm × 7 cm × 4 cm.

Figure 8 (A) Mature adipocytes and spindle cells without atypia and ropey collagen (H&E, 40×). (B) CD34 positivity in spindle cells (immunohistochemistry, 100×). (C) Desmin negativity in spindle cells and adipocytes (immunohistochemistry, 100×). (D) CDK4 negativity in spindle cells and adipocytes (immunohistochemistry, 200×).

of significant nuclear atypia are key to ruling out liposarcoma, and other similar cytological pathologies, such as pleomorphic lipoma.² The absence of MDM2 amplification or CDK4 assists in differentiating SCL from ALT (previously well-differentiated liposarcoma).^{7–9} Additionally, negative desmin staining helps to differentiate SCL from mammary-type myofibroblastoma.

TREATMENT

The patient elected to undergo an excisional biopsy of the left-hand soft tissue mass. In the operating room, a Bruner style volar incision to the palm was centred over of the soft tissue mass (figure 5). The mass was deep to the common palmar branches of the median nerve, and it extended into the hypothenar and thenar spaces. The encapsulated soft tissue mass was bluntly excised from the median nerve and flexor tendons. The mass was lobular with a predominantly yellow, lipomatous appearance and areas of grey-yellow lobulations (figure 6). The final specimen measured 11 cm × 7 cm × 4 cm (figure 7).

OUTCOME AND FOLLOW-UP

Histopathology of the soft tissue mass showed mature, homogeneous adipose tissue with transecting bands of spindle cells and ropey collagen (figure 8A). There was no cellular atypia and a Ki-67 showed no significant cellular proliferation (not shown). The spindle cells showed strong and diffuse expression of CD34 (figure 8B). Desmin and CDK4 were negative (figure 8C, D). In situ hybridisation showed no amplification of MDM2 (not shown). These findings were diagnostic of a SCL.

The patient returned to the hand clinic on postoperative day 12 and his sutures were removed from an incision that was clean, dry and intact. His symptoms were resolving with improved sensation and strength in his hand. He was instructed to follow-up as needed. He was asked to return at 5 months for an exam and confirmed that his symptoms were completely resolved and his incision was completely healed. He has returned to work in construction without any deficits.

DISCUSSION

SCLs were originally described in 1975 by Enzinger and Harvey.¹ SCLs are a rare subtype of benign lipomatous tumours often considered to be on a histological spectrum with pleomorphic lipoma, another rare subtype of lipoma with similar cytogenetic abnormalities. The two subtypes share a known genetic chromosomal abnormality, specifically losses at 16q and 13q chromosomes.^{2,3,9} A recent series examining 18 SCLs and histologically similar tumours demonstrated that all SCLs lacked Rb-1 expression, which is located on chromosome 13q.¹⁰ Pleomorphic liposarcoma can be differentiated from SCL based on the presence of unique giant cells with overlapping, marginally placed nuclei—referred to as ‘floret giant cells’.²

Macroscopically, SCLs are soft, lobular masses with yellow, grey-yellow colouration and a mucoid component, a somewhat ambiguous gross appearance lipomatous tumour family. The growth rate and advanced imaging characteristics of this mass were consistent with a benign lipomatous lesion; however, special consideration must be given to masses with certain characteristics including those >5 cm in size, which are considered to be ‘giant’ lipomas of the hand, and those in atypical anatomic locations or tissue depths which should raise suspicion for possible malignancy.⁴ Mentzel *et al* describe a similar clinical case of a large, palmar lipomatous neoplasm found on histological analysis to be an ALT with spindle cell features.¹¹ To date, very few palmar SCLs and one palmar ALT have been described in the literature and are significantly smaller than the one presented in this case.^{3–5,11} Thus, the cytological and immunohistological analyses become imperative to accurate diagnosis in similar cases. The classic histological appearance of SCLs contains a triad of mature adipocytes, ropey collagen and bland spindle cells—occasionally within a myxoid stroma. Lipoblasts may be present but are very rare, and there are no mitoses with minimal, if any, nuclear atypia compared with liposarcoma or pleomorphic lipoma.^{2,7,9}

The treatment of SCLs is marginal resection. A recent case series and numerous case reports of SCLs with up to a 2-year follow-up are devoid of local recurrence. Postexcision recurrence of subcutaneous lipomas in general is stated to be <1%–5% with adequate marginal resection.^{3,4,8}

Learning points

- ▶ Spindle cell lipomas are a rare subtype of lipoma typically appearing in men during the fifth to seventh decade of life in the upper back or shoulders.
- ▶ Atypical anatomical presentations and extraordinary size should raise concern for more concerning or malignant masses of similar gross and histological appearance, such as atypical lipomatous tumour or liposarcoma. This includes a mass outside of the shawl region and those >5 cm.
- ▶ The classic immunohistological triad for SCL shows positive CD34 and vimentin staining and negative desmin. Negative MDM2 and CDK4 can help confirm the diagnosis of spindle cell lipoma over ALT.
- ▶ Marginal excision is the definitive management and the rate of recurrence is very low with negative margins.

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