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Spindle cell liposarcoma of the face: case report and literature review

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Summary

Liposarcoma is a common soft tissue sarcoma accounting for approximately 20% of all mesenchymal tumours across all ages. Recently, collaborative research in the specialties of pathology and genetics has led to the delineation of several tumour variants with different behaviours and prognoses, one of which includes the very rare spindle cell liposarcoma (SCL) subtype. We present the first case of an SCL arising in the subcutaneous tissue of the forehead of a 78-year-old man. In light of the rarity of this tumour, we describe the tumour and its clinical and pathological characteristics and undertake a literature review to clarify the surgical management and prognosis of SCL, and increase awareness to avoid misdiagnosis of a benign soft tissue neoplasm.

According to the World Health Organization classification scheme in use since 1994, liposarcomas can be classified into three groups: (i) well-differentiated (including sclerosing, adipocytic, inflammatory, spindle cell and dedifferentiated variants) subtypes, (ii) myxoid/round cell liposarcoma and (iii) pleomorphic liposarcoma.1,2 Well-differentiated liposarcoma represents almost half of all liposarcomas with a tendency to develop both in the retroperitoneum and in the limbs. The spindle cell variant was first described in 1994 as a subtype of well-differentiated liposarcoma characterized by subcutaneous tissue involvement with an anatomical distribution comparable with the other subtypes.1

Case report

We report a case of a white 78-year-old man (nonsmoker), who was referred to our unit because of a left frontoparietal lesion following a fall 3 months earlier. Clinical examination revealed a well-circumscribed, painless, firm lesion with undamaged overlying skin and no regional lymphadenopathy. Ultrasound showed an ovoid mass (6 × 2 cm) with solid, nonhomogeneous content and with clear cleavage from the underlying tissues; Doppler imaging detected small vessels surrounded by a previous organizing haematoma. Computed tomographic scan confirmed an ovoid mass (7 × 3 cm) just above the squamous temporal bone with nonhomogeneous enhancement with intravenous contrast (Fig. 1). The clinical examination and radiological reports suggested that the lesion was probably a vascular anomaly.

The mass was enucleated through an overlying incision and a well-defined cleavage plane was observed. Macroscopically, the lesion presented as two compact yellow masses, with irregular surface and borders (Fig. 2). Histologically, the tumour had a multinodular appearance, and it consisted predominantly of a proliferation of spindle cells arranged in fascicles or in whorls, with scarce intervening matrix, that focally exhibited a myxoid appearance (Fig. 3). On these bases a histopathological diagnosis of spindle cell liposarcoma (SCL) was made. Distant and local metastatic disease was excluded. Radiotherapy followed in a standard protocol with fractioned doses of 50 Gy 5 days a week for 1 month. After 21 months the patient is free from disease.

Discussion

Nineteen previous cases of SCL were identified. The mean age was 37±1 years (range 11–83) (age was unavailable in seven cases). This rare subtype showed predilection for upper limbs (seven cases),1–5 lower limbs (five cases)6,7 and orbit (three cases),8,9 one of which was a metastasis.9 Neck,10 vulva,7 chest7 and lung7 accounted for only one case each. To our...
knowledge SCL described in this paper appears to be the first case involving the face arising in a site other than the orbit. Mortality seems to be low: three patients died of the disease within 4 years from primary diagnosis, two patients died of unrelated illness and one patient is still alive with unresectable metastasis. In four cases, the SCL recurred locally, whereas in two further cases distant metastasis developed. Tumour size and duration of the disease did not appear to be correlated well with prognosis.

The benefit of radiation and chemotherapy remains unproven from the collected data. In 13 cases treatment was not available, in two cases radiotherapy was associated with surgical resection and in one case palliative radiotherapy and chemotherapy were considered for a metastatic SCL to the orbit. It seems to be a minimally aggressive tumour either locally (it recurred in four cases) or distantly (two cases were distant metastases). Although three patients died of the disease, follow-up with a mean period of 54.8 months revealed a good survival rate after surgical resection as five patients were still alive with no evidence of disease. Two patients died of unrelated illness and the remaining cases were without follow-up data.

Among liposarcomas the spindle cell variant is one of the rarest as fewer than 20 cases are reported in the current medical literature. Although histological, clinical and molecular features are still poorly understood, its cytogenetics are well established, consisting of the presence of supernumerary ring or giant chromosomes containing amplified material from chromosome 12q14–q15 which includes the MDM2 and CDK4 genes. Recently a loss of genomic material from chromosome 7 without supernumerary chromosomes containing 12q amplified sequences or duplication of the 12q region has been demonstrated to be another molecular cytogenetic character-
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What's already known about this topic?

- A century and a half after Virchow originally described liposarcoma, several subtypes have been classified.
- We still have relatively poor knowledge of this disease and several unknowns remain in relation to prognosis and treatment despite the advent of cytogenetic and molecular investigations, and increased understanding and classification of lipomatous neoplasms.

What does this study add?

- Our review of the literature and illustrative case report provide an update as to the current state of play in relation to the spindle cell variant, investigations, prognosis and treatment.
- In this paper are collected all the spindle cell liposarcomas (SCLs) available from a detailed literature review.
- The case report of SCL arising in the soft tissue of the face is the first described in the medical literature, to our knowledge.

References