Spindle Cell Liposarcoma

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Introduction

Spindle cell liposarcoma (SCL) is an uncommon variant of well-differentiated liposarcoma (or atypical lipomatous tumor, adipocytic liposarcoma, lipoma-like liposarcoma). About 20 cases have been reported since the first description in 1994.

Clinical Features

It presents as a painless, slowly enlarging mass, becoming symptomatic when impinging upon surrounding structures; its indolent course can result in a misdiagnosis of cyst or benign soft tissue neoplasm, especially lipoma. It shows predilection for subcutaneous soft tissue of the extremities and orbit. The extraorbital facial locations, neck, vulva, trunk, and palm have also been reported. It tends to occur in adults (range 11–83).

Pathology

It is composed of a spindle cell bland neural-like proliferation arranged in fascicles and whorls set in a fibrous and/or myxoid stroma (Figs. 40.1 and 40.2) and associated with atypical adipocytes (often including lipoblasts) showing variation in size and shape with scattered enlarged and hyperchromatic nuclei (Figs. 40.3 and 40.4). Although grossly liposarcoma is encapsulated, it extends by infiltration. Spindle cells usually exhibit CD34 and adipocytes show S100 protein immunoreactivity.

Incisional biopsy is not indicated in large adipose tumors as malignant degeneration is usually at the center of the mass, and malignant features can be missed, leading to inappropriate treatment.

Genetically, well-differentiated liposarcomas are characterized by the presence of a supernumerary ring or giant chromosomes containing amplified material from chromosome 12q14–q15, which includes the MDM2 and CDK4 genes. However, SCL tends to lack the amplification of MDM2 and/or CDK4, differently from

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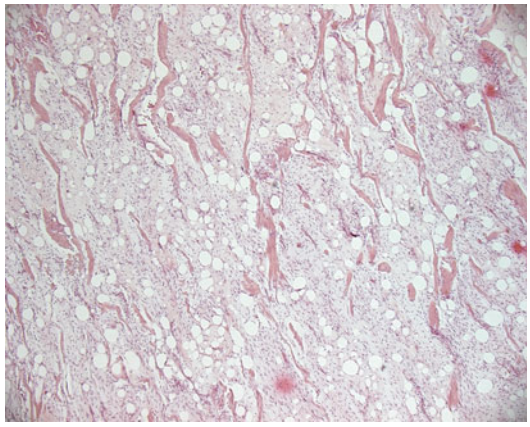

Fig. 40.1 Spindle cell liposarcoma. The neoplasm is composed of a spindle cell bland neural-like proliferation arranged in fascicles and whorls set in a fibromyxoid stroma

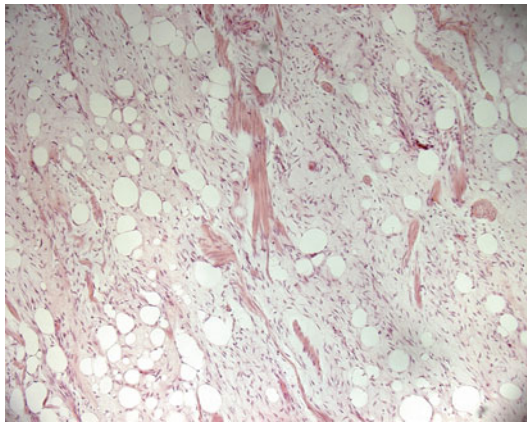

Fig. 40.2 A spindle cell bland neural-like proliferation

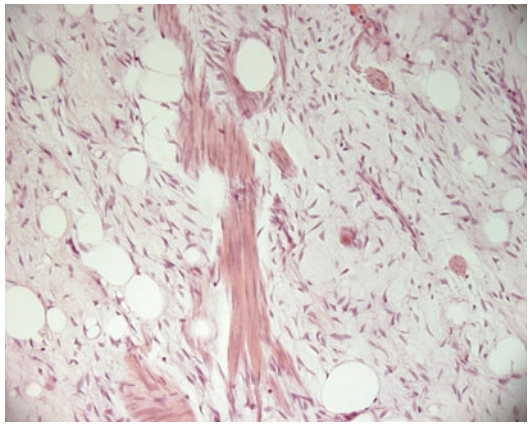

Fig. 40.3 Spindle cells with blunt atypia

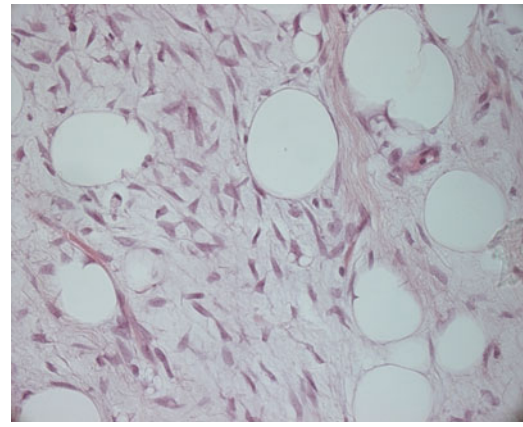

Fig. 40.4 Spindle cells associated with atypical adipocytes and lipoblasts

the other subtypes of well-differentiated liposarcoma. Recently, a partial or complete monosomy of chromosome 7 and a deletion of the Rb-1 gene in the long arm of chromosome 13 without mutations of chromosome 12q region have been identified as other molecular cytogenetic characterization.

Differential Diagnosis

The main differential diagnoses are diffuse spindle cell lipoma (composed of bland, sometimes palisading, CD34-positive spindle cells, admixed with eosinophilic refractile collagen bundles and presenting the Rb-1 deletion) and neurofibroma (characterized by a less cellular S-100-positive spindle cell proliferation with wavy nuclei). Other differential diagnoses are dermatofibrosarcoma protuberans, low-grade malignant peripheral nerve sheath tumor, low-grade sarcoma, low-grade myxofibrosarcoma, dedifferentiated liposarcoma, and the other variants of well-differentiated liposarcoma, such as sclerosing liposarcoma. Low-grade dedifferentiated liposarcoma contains dedifferentiated areas that are generally non-lipogenic, whereas SCL contains well-differentiated atypical adipocytes or lipoblasts, although

sometimes they may dedifferentiate forming non-lipogenic areas.

Prognosis

Mortality seems to be low. The World Health Organization classifies SCL among intermediate (locally aggressive) adipocyte tumors, since local recurrences are quite frequent (around one fourth of patients) and distant metastasis are rare. Prognosis is influenced by adequacy of surgical excision, whereas tumor size and duration of the disease did not appear to be correlated well with prognosis.

Treatment

Surgical removal remains the treatment of choice. The benefit of radiation and chemotherapy remains unproven.

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