Abstract. – Superficial leiomyosarcoma is an exceeding uncommon malignant tumor, which can be located either cutaneous or subcutaneous tissues. It may occur anywhere in the body, but there is a predilection for the thighs. The distinction between cutaneous or subcutaneous, increased mass size (>5 cm) and depth are considered to have worst prognosis in terms of recurrence and metastasis.

We report a rare case of an 81-year-old woman, presented with a 20-cm leiom yosarcoma of the shoulder. The patient reported that the mass was completely asymptomatic and its growth was slow. A CT study showed a lesion closely opposed to the head of the humerus and revealed no a clear relationship with the surrounding muscles. There was no regional lymphadenopathy or the evidence of metastatic disease.

After tumor resection, a large skin and soft-tissue defect was remained and the repair was performed by myocutaneous rotational free flap of latissimus dorsi.

The conclusion of the histological analysis was: subcutaneous leiom ysarcoma pT2 – grade 1+ (NCI system).

The delayed diagnosis and the surgical treatment and giant size of soft tissue leiom yosarcoma may adversely affect the final result.

Key Words:
Leiom ysarcoma, Skin sarcoma, Subcutaneous leiom yosarcoma, Myocutaneous rotational free flap

Introduction

Subcutaneous soft tissue leiom yosarcomas have a rare incidence accounting for 2.3-5.3% of all malignant soft tissue tumors. This tumor, developed most commonly between the ages 40 and 60 years, appears to be more common in males at a ratio of 2:1 or 3:1. The etiology is unknown; predisposing factors include leiom yomas, trauma, and exposure to radiation. It may occur anywhere in the body, but there is a predilection for the extremities, especially the thighs.

Among several different grading systems for soft-tissue sarcomas, the Fédération Nationale des Centres de Lutte Contre le Cancer grade was the significant prognostic factor for patients with soft tissue sarcomas, including leiom yosarcomas. The standard sarcoma staging system is the tumor-node-metastasis of the American Joint Committee on Cancer.

Leiom yosarcomas are derived from the smooth muscle cells of blood vessels, predominantly in the visceral locations such as the uterus, gastrointestinal tract, mesentery, urogenital system, and retroperitoneal space. Conventionally, superficial leiom yosarcomas may be subdivided into two types by their primary tissue of origin: cutaneous and subcutaneous. The distinction is made on the basis of different histological features.

Surgical excision is the only successful therapeutic procedure. In fact, leiom yosarcoma has been reported to be radioresistant, and chemotherapy was unsuccessful. Surgical treatment for cutaneous leiom yosarcoma, consisting of complete excision with a narrow margin, has been recommended, while a 2- to 5-cm excisional margin has been suggested for subcutaneous or soft-tissue leiom yosarcoma, with a depth including the subcutaneous tissue and deep fascia.

Tsutsumida et al. have treated patients with superficial leiom yosarcoma with a minimum 2-cm margin. The patients with low-grade and early stage disease have not developed any local recurrences or distant metastasis for more than 20 years. However, for high-grade and advanced-stage cases the patients had a poor prognosis.
Case Report

An 81-year-old woman presented with a 20-cm leiomyosarcoma of the shoulder (Figure 1). Seven years before the patient had been operated for a mitral valve replacement. The patient reported that the mass was completely asymptomatic and its growth was slow. A CT study showed a lesion closely apposed to the head of the humerus and revealed no a clear relationship with the surrounding muscles (Figure 2). There was no regional lymphadenopathy or the evidence of metastatic disease.

Under general anaesthesia an en block wide excision of the mass was performed (Figure 3) with a partially removal of the surrounding muscles (pectoralis major and deltoid). After the excision, a large skin and soft-tissue defect was remained and the repair was performed by myocutaneous rotational free flap of latissimus dorsi completely detached from the dorsal and humeral insertions (Figure 5).

Histology revealed, the presence of multiple elongated cells of mesenchymal origin surrounded by myofibrils and many polygonal epithelioid cells with evident pleiomorphism, elevated mitotic index (about 30 mitosis/10 HPF) (Figure 4) and high proliferative index (about 50%) estimated by Ki67. The surgical margins were free of disease. The immunohistochemical staining was positive for smooth muscular actin and desmine and negative for CD34 and S-100 (Figure 6). The conclusion of the histological analysis was: subcutaneous leiomyosarcoma pT2 – grade 1+ (NCI system).

Discussion

Clinically, leiomyosarcoma must be distinguished from basal cell carcinomas, squamous cell carcinomas, epidermoid carcinomas, dermatofibromas, lipomas, neurofibromas, and benign papillomas. The case we reported has an atypical age of onset and atypical manifestation with huge (20 x 25 cm) subcutaneous tumor. However, the location on the shoulder is unusual.

Histological features of leiomyosarcoma alone may occasionally lead to confusion with other cutaneous spindle cell tumors. The histological differential diagnosis includes: fibrosarcomas, neurofibrosarcoma, malignant histiocytoma, neurilemmoma, atypical fibroxanthoma, and dermatofibrosarcoma. Immunohistochem-
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Figure 4. Histopathological examinations revealed spindle cells with dull shaped nuclei in a fibrous stroma and some mitotic figures (original magnification 40 ×).

Figure 5. Post-operative aspect of the patient. Adequate coverage of defects of the shoulder obtained with myocutaneous rotational free flap of latissimus dorsi.

Figure 6. Immunohistochemical staining revealed some mitosis.

ease was the grading and the stage of the tumor and an inadequate wide excision.

Advanced therapies, such immunotherapy and gene therapy, are being developed. Although regional lymphonodes were detected in about 25% of Stout and Hill’s patients 2, it is enough to perform a therapeutic lymphonodes dissection, because there is an insufficient evidence for an elective lymphonodes dissection 13. Adjuvant radiotherapies or chemotherapies were used to treat some patients. However, these therapies may have been unnecessary for the low-grade and early-stage cases, while the efficacy of these therapies is still conflicting for the high-grade leiomyosarcoma.

Inadequate excision or local excision without adequate margin leads to recurrence and increases the risk for the metastatic and possibly fatal disease 3.

In a series reported by Fields and Helwig 3, local recurrence developed with 47% of subcutaneous leiomyosarcomas. Recurrence usually occurred within 1.5 years after initial surgery 3. Metastases have rarely been described in dermal leiomyosarcomas, in contrast to the 33% incidence of metastases in subcutaneous leiomyosarcomas. Usually the lungs, liver and bones are affected.

Awareness of the particularly misleading features of this tumor, especially in elderly patients, is important, as delayed diagnosis is correlated with larger size and invasiveness into contiguous tissues, which influence the surgical procedure of radical resection with 2-5-cm of free margins of disease. Surgery is the most and only way to treat superficial and subcutaneous leiomyosarcomas and a wide local excision is necessary to avoid local and metastatic recurrence.
References


