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# Giant Cutaneous Leiomyosarcoma Originating From the Abdominal Wall: A Case Report

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Patient:		Male, 44	
Final Diagnosis:		Cutaneous Leiomyosarcoma	
Symptoms:		Abdominal mass	
Medication:			
Specialty:		Surgery	
-			
Objective:		Rare disease	
Background:		Leiomyosarcoma, a rare type of tumor, accounts for 5–10% of all soft tissue tumors.	
Case Report:		A 44-year-old male patient was admitted to the emergency service of our medical faculty with the complaints	
Conclusions:		The pathology result was leiomyosarcoma. Leiomyosarcoma of the skin is rare and our case is the largest such lesion reported in the literature.	
MeSH Keywords:		Gastrointestinal Neoplasms • Leiomyosarcoma • Photosensitivity Disorders	
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# Background

Leiomyosarcoma, which is a rare type of tumor, accounts for 5-10% of all soft tissue tumors [1,2]. Cutaneous leiomyosarcoma is a less common subgroup and, although it can be seen in any age groups, the prevalence is highest between the 5<sup>th</sup> and 7<sup>th</sup> decades. It is more frequent in extensor surfaces that have higher hair content on the extremities [3]. It arises from the arrector pili muscle of hair follicles. In the initial diagnosis, cutaneous leiomyosarcoma is a tumor usually smaller than 2 cm [4]. The behavior of these tumors is more consistent with that of deeper tumors than intradermal tumors. We report an interesting case of a leiomyosarcoma with an uncommon location and size.

# **Case Report**

A 44-year-old male patient was admitted to the emergency service of our medical faculty with the complaints of fatigue and abdominal mass. In his physical examination, a giant mass, exhibiting ulcers and umbilications, as well as superficial bleeding regions, was present on the anterior wall of the abdomen, with the approximate dimensions of 45×35×15 cm (Figure 1). Medical history of the patient revealed that the mass first appeared as black dots around the umbilicus 3 years before, subsequently growing to the current dimensions. It was learned that the patient, who had had 3 surgeries for nephrolithiasis 33 years ago and had developed psychosis, did not seek medical attention because he held the belief that his evil spirits passed outside through the mass. He had lost 40 kg of body weight during the last 6 months and his general condition had declined. At admission, his blood pressure was 110/60 mmHg, heart rate was 104/min, and body temperature was 36.7°C. He had deep anemia and thrombocytosis (Hb: 5.4 g/dL, Plt: 704 000/mm<sup>3</sup>). Carcinoembryonic antigen (CEA),



Figure 1. The cutaneous lesion, which included ulcers and foci of bleeding.



Figure 2. Computerized tomography image of the mass.



Figure 3. Intraoperative image that shows adhesions to anterior rectus fascia.

alpha-fetoprotein (AFP), and carbohydrate-associated antigen (CA) 19-9 levels were normal. In computerized tomography, there was a heterogeneous contrasting mass lesion with lobulated contours and necrotic areas, with dimensions of  $30 \times 25 \times 15$  cm. It started from the epigastric level and extended to the pelvis, being located at the median fatty tissues of the skin and sub-skin. This mass was significantly compressing the posterior structures of the abdominal wall, and causing skin ulcers (Figure 2).

After preoperative preparation, the patient underwent surgery on October 23<sup>rd</sup>, 2007. The mass was surrounded by a pseudocapsule and had partial adhesions to the anterior rectus fascia (Figure 3). It was excised with additional removal of the anterior rectus muscle sheath, with dimensions of 10×10 cm. The defect at the fascia was repaired with polyprolene mesh as a patch (Figure 4). The defect at the abdominal wall, sized 30×30 cm, was closed with an anterolateral pediculated thigh flap (Figure 5).



Figure 4. Anterior rectus sheath repair with a polyprolene patch.



Figure 5. After reconstruction with an anterolateral pediculated thigh flap.

#### Histopathological findings

Macroscopic evaluation revealed a lesion with the dimensions of  $45 \times 35 \times 15$  and an elliptical skin surface of  $45 \times 35$  cm, which had a nodular appearance. There was a hard, gray-white-colored, lobulated mass with relatively regular borders, and dimensions of  $26 \times 24 \times 15$  cm. Fibrillary deposits were visible in the cross-section of the material.

In histopathological evaluation, the tumor formation was characterized by a number of atypical cells extending in various directions as clusters that were fusiform and spindle-like, and had obtuse ends, with hyperchromatic nuclei and pale eosinophilic cytoplasms (Figure 6). There were widespread necrosis spots in the tumor, and 6 mitosis regions in a single area of ×10 magnification.

Although immunohistochemical studies revealed immunoreactivity with vimentin and smooth muscle actin, there was no immunoreactivity with CD 117 or S100 in tumor cells (Figure 7).



Figure 6. Fusiform tumor cells that were extending in various directions (H&O ×200).



Figure 7. Smooth muscle actin immunoreactivity in tumor cells (DAB ×400).

The diagnosis of cutaneous leiomyosarcoma was made based on morphological and immunohistochemical findings.

At 3-year follow-up there was no recurrence or metastasis.

## **Discussion**

There are various types of gastrointestinal cancers [5–7]. Soft tissue sarcomas are rare neoplasms that constitute approximately 1% of all adulthood cancers [5]. The locations of these tumors are: 43% in the extremities, 19% in the organs, 15% in the retroperitoneal space, 10% in the body/chest, and 13% in other areas [8]. Leiomyosarcomas constitute 5–10% of these tumors [1,2]. When 6486 patients diagnosed with soft tissue sarcomas in Memorial Sloan-Kettering Cancer Center between July 1<sup>st</sup> 1982 and June 30<sup>th</sup> 2005 were reviewed, it was observed that 15% of them (978 cases) were leiomyosarcomas [8].

Leiomyosarcomas are classified into 4 main groups. The first group includes leiomyosarcoma of the retroperitoneal space and abdominal cavity and is the most common subgroup. The second group is soft tissue leiomyosarcoma, which has a better prognosis. The third group is cutaneous leiomyosarcomas; because its localization is superficial, the prognosis is better than in the other groups. The fourth subgroup includes vascular leiomyosarcomas [9]; these tumors, which occur more frequently in adulthood, are more prevalent in females than males, whereas cutaneous leiomyosarcomas are more common in males than in females [3].

It was previously suggested that cutaneous leiomyosarcomas originate from the dermis or subcutaneous tissue, but it was recently precluded that these lesions develop from the dermis, later extending this to subcutaneous tissues [9]. Even its etiology is not clearly defined; precancerous lesions, physical trauma, and radiation exposure are reported as possible causative factors [3]. Cutaneous tumors are generally smaller than 2 cm at the time of early diagnosis [3,9]. Generally, they include cutaneous findings like color changes, umbilications,

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and ulcerations. As the tumor develops, it extends to subcutaneous tissues, compresses surrounding tissues, and may form a pseudocapsule [9].

Leiomyosarcomas have only limited response to radiotherapy or chemotherapy [3]. Immunotherapy and gene therapy are treatment models still at developmental stages. The only treatment option with proven success is surgery [10,11].

In cutaneous leiomyosarcomas, a 42% local recurrence rate was reported by Fields and Helwig [3] and 14% by Bernstein and Roenigk [12] and Jensen et al. [4].

## Conclusions

Leiomyosarcoma of the skin is a rare condition and our case is the largest such lesion in the literature. The patient's psychosis, associated with a superstitious belief, allowed the mass to reach an enormous size. After extensive resection, the prognosis of the patient was good at 3-year follow-up.

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