

Paravertebral leiomyosarcoma mimicking a chronic ongoing inflammatory process

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ABSTRACT

Leiomyosarcomas usually present with symptoms associated with the tumor site or as painless soft tissue masses. We report the case of a young woman with spiking fever and elevated acute reaction proteins for months, in the context of a paravertebral high grade leiomyosarcoma.

Case report

The patient, a 20-year-old female student, was admitted to our hospital in June 2001 with a three-month history of spiking fever. From November 1999 anemia, thrombocytosis, high ESR, CRP and diffuse hypergammaglobulinemia were evident. Extensive investigations for infectious and malignancies in previous hospitalizations were unrevealing.

The patient's past medical history included heterozygote α -thalassemia and left heminephrectomy and thyroidectomy at the ages of 2 and 16 years, respectively. She had no history of weight loss, anorexia, pain, diarrhea, surgery, travel, exposure to ill persons or animals, or use of tobacco, alcohol and medications.

On physical examination she was pale and had a fever of 38.7°C. Cardiac, pulmonary and abdomen examination revealed no abnormal findings except for mild splenomegaly. An elastic, soft tissue mass in the left paravertebral area (C7-T2) was observed. This mass had been first noted in January 2000 and was clinically characterized as a lipoma. In June 2000 a magnetic resonance imaging scan (MRI) was suggestive of hemangioma.

Laboratory examinations revealed: Hb 6.7 g/dl, WBC 6100/mm³, platelets 652000/mm³, CRP 23 mg/l (cut off: 0.5 mg/l), ESR 145 mm/h, SGOT 87 U/l, SGPT 112 U/l, GT 135 U/l, ALP 465 U/l, PT 16 sec, Fe 10 µg/dl, ferritin 156 µg/dl, and urine tests with no pathologic findings. Serum protein electrophoresis showed hypergammaglobulinemia with no monoclonal bands. Immunological tests were negative. Blood and urine cultures and serological markers for herpes viruses, HIV, HBV, HCV, parvovirus, CMV, EBV were negative.

Chest x-ray was normal. A skin tuberculin test was negative. Bone marrow aspiration demonstrated mild dyserythropoiesis; no cytogenetic abnormalities were detected. Transesophageal heart ultrasonography, gastroscopy and colonoscopy were negative. Duodenum biopsy showed mild infiltration of plasma cells. Liver biopsy revealed non-specific findings. CT of the abdomen and digital angiography of the mesenteric, hepatic and renal arteries showed no abnormalities except for a hemangioma of the right liver lobe.

Based on the clinical symptoms and laboratory findings, Still's disease seemed to be the most probable diagnosis and treatment with methylprednisolone 48 mg daily *per os* was started. Two months later she reported no fever. CRP and ESR were slightly decreased. In September 2001 she presented with high fever (39.5°C) and chills. On physical examination the mass in the left paravertebral area had increased in size, but no pain, swelling or erythema was noted. MRI was repeated and documented a well defined, heterogeneously enhancing mass (C7-T4) with multiple septations and cystic formations, suggestive of fibrous histiocytoma (Fig. 1). Complete surgical resection was performed.

The histologic analysis showed macroscopically a neoplastic tumor (dimensions 8x 7.2 x 3.5 cm) partially covered by fascia, focally ruptured and in contact with the skeletal muscle. Microscopic examination and immunohistochemical studies (strong positive cells for desmin, synaptophysin and focally for EMA) revealed a high grade undifferentiated leiomyosarcoma, with epithelioid and pleomorphic features and areas of necrosis. After tumor excision the fever disappeared and the inflammatory markers fell to normal. The treatment plan adopted was: multi-agent chemotherapy followed by re-excision of the tumor bed, local radiotherapy and an additional course of chemotherapy. The patient completed pre-operative chemotherapy with etoposide IV, ifosfamide IV and doxorubicin IV with good response. The margins of the lesion were negative for tumor. Radiation therapy with Iridium

192 at a total dose of 4500 cGy was given without complications. Two months after the completion of the six cycles of chemotherapy, the patient remains in excellent condition based on both clinical and laboratory parameters.

Discussion

Leiomyosarcomas are aggressive soft tissue tumors that account for 5% to 10% of all soft tissue sarcomas. They can be divided into groups based on their location: retroperitoneal, deep soft tissue other than abdominal site, cutaneous or subcutaneous, and vascular. The histologic features of leiomyosarcoma show a wide variety of patterns and the histopathological diagnosis is difficult even with the assistance of immunocytochemistry (1). Prognosis has been correlated with the tumor size and site, its histologic pattern, and the resection quality (2).

The clinical manifestations of leiomyosarcomas may vary according to the tumor site. Pain and tenderness can be found in the leiomyosarcomas of the joints, extremities, and trunk wall. While leiomyosarcoma most commonly presents as a painless mass, it occasionally emerges in a misleading manner resulting in an unfavorable delay or error in diagnosis.

We describe here a patient with high grade paravertebral leiomyosarcoma mimicking a chronic ongoing inflammatory process. This patient presented with chronic anemia, high levels of acute reaction proteins and high grade, spiking fever in the later stages. To our knowledge, no constitutional symptoms or laboratory findings characteristic of an inflammatory process have been associated with leiomyosarcomas of the external soft tissue in the litera-

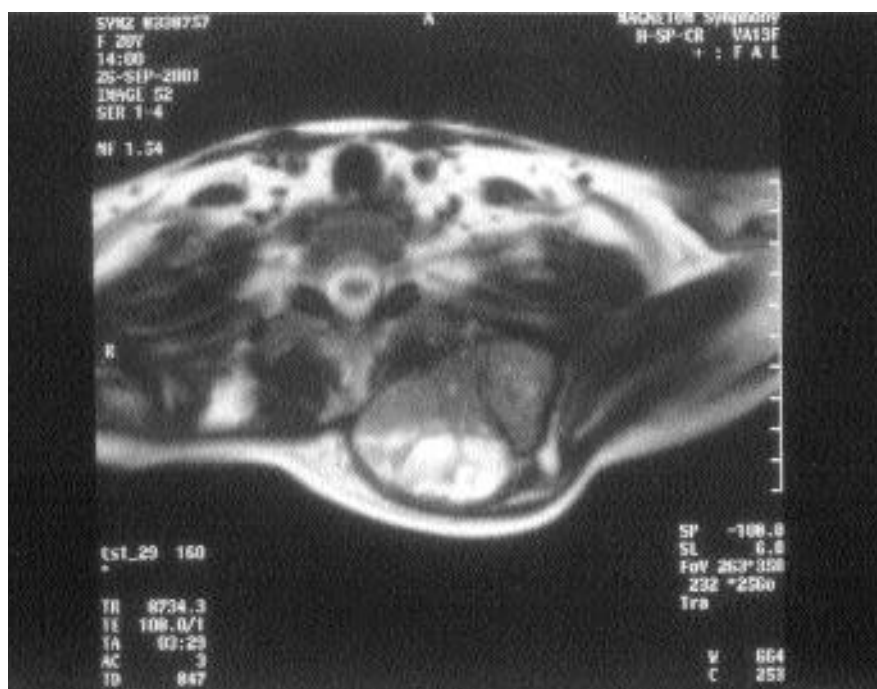


Fig. 1. MRI shows a well-defined mass with multiple septations and cystic formations suggestive of fibrous histiocytoma in this patient with leiomyosarcoma.

ture. Acute or prolonged fever has been reported only in sporadic cases of leiomyosarcoma affecting the intra-abdominal organs (3).

This case illustrates that an inflammatory reaction can be added to the spectrum of the clinical manifestations of leiomyosarcomas. The pathogenetic mechanism of this process is unclear. Cytokine expression by sarcoma cells may be one hypothesis (4).

MRI has been reported in the literature as the most valuable method for the detection and staging of leiomyosarcomas. Leiomyosarcomas, unless they are aggressive tumors, may have a surprisingly benign imaging appearance (5). The differential diagnosis from other benign or malignant soft tissue masses is often difficult. Histologic examina-

tion of the lesion is essential for an accurate diagnosis.

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