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Case Report

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Painful Abdominal Mass with undetected growth in a 46 years old man

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Abstract

Malignant fibrous histiocytoma (MFH) is a sarcoma originating from fibroblast cells. Retroperitoneal soft-tissue sarcomas are rare tumors with an incidence of 0.5 to 1 new cases per 100.000 inhabitants per year. We report a case of MFH in the retroperitoneum in a 46-year-old man who presented a left lower abdominal pain and an abdominal mass with slow growth. CT scan of the abdomen demonstrated a well-circumscribed mass in the left iliopsoas compartment extending to the root of thigh and to left femoral head. The patient underwent an extraperitoneal approach with extension for infra-inguinal exposure and the tumor was resected. Histopathological examination of the specimen measuring 12 centimeters revealed a Malignant Fibrous Histiocytoma. The patient had an uneventful postoperative course and was given adjuvant radiotherapy. He is currently well 5 years after his operation.

Keywords: Abdominal mass, Retroperitoneal soft-tissue sarcomas, Malignant Fibrous Histiocytoma.

History and Physical Examination

A 46-years-old male patient was referred to our department with a two month history of left lower abdominal pain. The pain was located, nonradiating, presented at night, it didn´t increased with the movement, it didn´t tend to be relieved with nonesteroidal anti-inflammatory drugs. Approximately a month before the patient noted the presence of an abdominal mass with slow growth.

On physical examination a well-defined firm mass was palpable in the left flank, adherent to deep planes. There was a negative percussion sing. He had limited the hip flexion and started feeling numbness in the left anterior thigh. There was no antecedent of trauma or constitutional symptoms. The medical history was otherwise unremarkable and there was no family history of neoplasia. A plain radiograph of the pelvis was normal. An ultrasound was performed to localize the lesion, and a computed tomography (CT) and magnetic resonance (MR) to characterize the lesion.

Imaging Interpretation

Ultrasound showed a solid tumor on the left inguinal ring in relation with the iliopsoas muscle. CT scan of the abdomen demonstrated a well-circumscribed mass in the left iliopsoas compartment extending to the root of thigh and to left femoral head, measuring 120x60x70 mm and presenting heterogeneous contrast uptake predominantly peripheral and central areas without any acquisition that could correspond to necrosis (Imaging 1 and 2). Therefore, MR imaging showed a heterogeneous hypointense signal on T1 sequence with losing fat plane in the left iliopsoas muscle as well as high intensity and heterogeneous signal in T2 sequence with fat saturation after having administered gadolinium heterogeneous enhancement. Scintygraphy didn´t show metastatic spread and arteriography didn´t display the formation of new blood vessels. Image-guided percutaneous multiple needle core biopsy was performed.

Based on the history, physical examination, and imaging studies, what is the differential diagnosis at this point?

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Differential Diagnosis

Soft-tissue sarcoma: liposarcoma, leiomyoma, malignant fibrous histiocytoma

Metastatic tumors

Infective process

Haemorrhagic process

Benign and malignant neural tumors

The patient underwent an extraperitoneal approach with extension for infra-inguinal exposure and the tumor was resected and evaluated histologically.

Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, what is the diagnosis and how should the patient be treated?

Hystology Interpretation

Histopathological examination of the specimen measuring 12 centimeters revealed a mesenchymatous tumor consisting of spindle cell with pleomorphic, enlarged nuclei demonstrating medium to high mitotic activity. A detail inmunohitochemical analysis demonstrated that tumor cells were stained strongly positive for cd68 and vimentin, whereas a negative staining for c-kit, S-100 protein, cd57, desmin, myogenin, and alpha smooth muscle actin was noted. Tumor grade was classified as high grade with a proliferative index (Ki67) about 40%-50%. Microscopic margins were negative.

Histologic analysis of three femoral lymph nodes showed absence of tumoral infiltration

Diagnosis

Malignant Fibrous Histiocytoma.

DISCUSSION AND TREATMENT

In this patient, the pertinent findings supporting the diagnosis of malignant fibrous histiocytoma (MFH), CT scan was very helpful to diagnose, to evaluate the extent of tumor and the involvement was shown by enlargement of the muscle with altered attenuation. Histopathological examination confirmed the diagnosis of MFH.

Retroperitoneal soft-tissue sarcomas are rare tumors with an incidence of 0.5 to 1 new cases per 100.000 inhabitants per year [1]. MFH is one of the most common soft-tissue sarcoma [2,3,4,5] and this generally occurs in 50-70 year-old people [2,6]. This entity affects both sexes, most of the tumors are located at the extremities, its predilection for the thigh was reported in various studies[4,7], almost half are subcutaneous [3],and only 16% appears in the retroperitoneum [3,8]. Depth and size are the most important factors for local recurrence and distant metastasis [6].

The differential diagnosis based on clinical examination and radiological imaging is broad and it includes various soft-tissue sarcoma, metastatic tumors, abscess, haemorrhagic process, and benign and malignant neural tumor. The differentiation of iliopsoas lesions even using ultrasound, CT and MRI can be difficult [9,10]. CT has led to an increase in recognition of psoas pathology in general and malignant involvement in particular. Image-guided percutaneous core needle biopsy is strongly recommended. The most common malignant tumors to occur in this area are metastatic tumors such a testicular, renal, cervical, bladder and prostate tumors [12]. On MRI, metastatic involvement is usually low signal on T1-weighted images and high on T2-weighted images. These tumors affect older patients more frecuently. Benign and malignant neural tumors are common because of the intimate relationship to the lumbar plexus and sympathetic trunk [11,12]. MRI is helpful where spinal canal/vertebral involvement is suspected and axial images provide data to determine the origin. Abscess may include fever and chills. Blood test can be helpful. CT features includes postcontrast enhancement of the peripheral abscess wall and gas bubbles are often found in abscess but rarely with tumor [10]. MRI findings include low signal and enhancing areas on contrast on T1-weighted images, heterogeneously high signal on T2-weighted images. It is possible to confirm the presence of purulent material by diagnostic needle aspiration. Haemorrhage is rare and usually occurs secondary to trauma or from involvement of the psoas muscle in abdominal aortic aneurysm rupture [10,11]. The diagnosis of rupture of the abdominal aortic aneuryms involves acute abdominal pain and high attenuation on CT images. Soft-tissue sarcoma in the iliopsoas compartment often appears in an indolent manner with non-specific signs and symptoms. Differentiation from lipomatous tumour, leiomyosarcoma, and lymph node metastasis, is also difficult. CT-guided biopsy is the best alternative due to the convenience in the retroperitoneal space [1,8] and the diagnosis is mainly based on histologic and immunohistochemical findings. In dedifferentiated liposarcomas histologic criteria is defined by the presence of a clear-cut, well-differentiated liposarcoma component, separated from the poorly differentiated component [14]. immunohistochemical study shows positivity for mdm2 and cdk4. Leiomyosarcoma expresses a myogenic line differentiation: eosinophilic splindle cells in a fascicular pattern. This tumor also shows extensive positivity for smooth-muscle actin and desmin.

MFH is a sarcoma of mesenchymal origin, composed of tumor cells without evidence of an specific differentiation, both histiocytic and fibrous elements [15]. Based on its histopathologic features, MFH has been categorized into stori-pleomorphic, mixoid, inflammatory, giant cell and angiomatoid variants. Among these types, the stori-pleomorphic is most common [4,14,16]. It is composed of splindle-shaped cells arranged in short fascicles in a storiform pattern, along with plump histiocyte cell, mitotic figures and multinucleated giant cells. Inmunohitochemical analyses shows tumor cells positive for vimentin, and negative for cytokeratins, desmin, S-100 protein, actins, c-kit and CD34.

The diagnosed of MFH was favored by histologic pleomorphism, degree of vimentin staning, absence of the smooth mucle marker desmin, and the macroscopic and microscopic findings of the resected tumor of our patient.

In the retroperitoneal space, MFH represents from 7 to 30% of sarcomas. They have varying clinical courses depending on their histological subtype and grade and are usually very large at the time of presentation. Subfascial tumors are bigger at the time of diagnosis and have longer duration of symptoms. Therefore, they carry the risk of infiltrating neighboring structures and metastatic spread. In an institutional review, almost all patients presented with indolent tumor [3]. The most common symptoms reported is a painless mass and complain about fatigue, weight loss and abdominal, back or hip pain [2]. This symptoms are clinically undetectable very often and the tumor can rise for longer periods of time [9,17]. In our case, symptoms encompassed lower extremity pain, abdominal distension, and femoral nerve involvement presented as instability of the knee when coming downstairs and loss of hip flexion. The prime cause of death is distant metastases and the most common site is the lung with about 80%, the majority of the metastases occurred within the first 2 years. Overall prognosis is influenced by tumor size, grading and resected margins, being negative microscopic margins the most favorable prognostic factor.

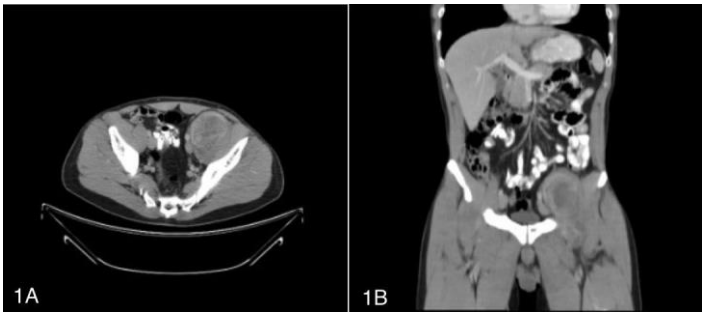

Figure 1A-B: (A) Axial CT scan image shows a mass in the left iliopsoas compartment presenting heterogeneuos contrast uptake predominantly peripheral and central areas without any acquisition that could correspond to necrosis.(B) Coronal CT scan image demonstrating the tumoral extending to the root of the tight

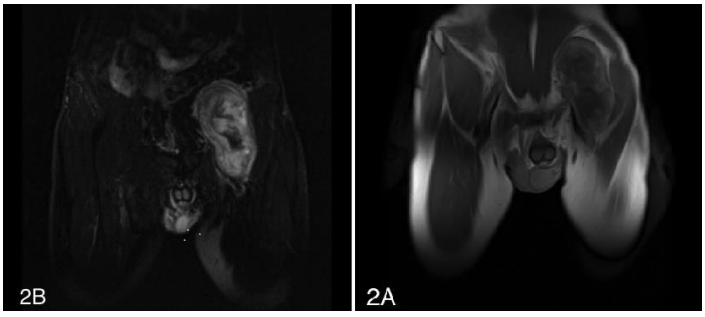

Figure 2A-B: (A) MR imaging showed a heterogeneous hypointense signal on T1 sequence with losing fat plane in the left iliopsoas muscle. (B) High intensity and heterogeneous signal in T2 sequence with fat saturation after administered gadolinium heterogeneous enhancement

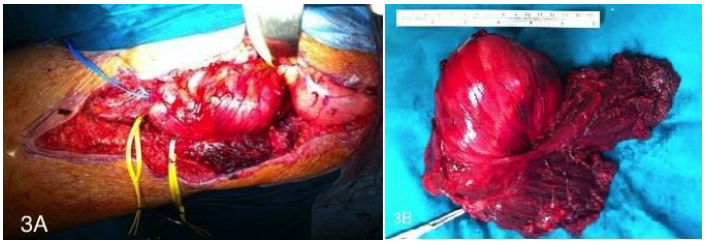

Figure 3A-B: (A) Intraoperative photographs show the infrainguinal and extraperitoneal approach preserving specific organs. (B)After removal de tumor, en bloc negative margin excision was confirmed encompassing the tumor and involved iliopsoas muscle

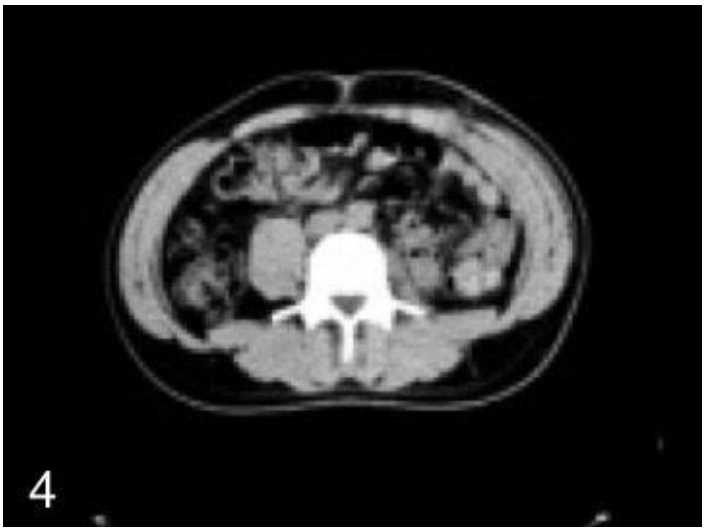

Figure 4: CT scan at the time of the final follow-up shows absence of left iliopsoas muscle with no signs of tumor recurrence

In the diagnostic assessment of the patient, adequate physical exam and images are very important. Ultrasound provides a diagnostic in only 60% of cases [10,11]. Contrast CT is the definitive initial investigation for evaluation and findings containing areas of low attenuation with loss of fat plane and mild contrast enhancement. In our patient CT showed a well-circumscribed mass with peripheral enhancement and hypodense central area necrosis. MRI showed a mass, hypointense on T1-weighted images and heterogeneous high intensity signal on T2-weighted images.

The optimal individual management plan should be determined in a multidisciplinary team preoperatively based on the imaging of the tumor and its surrounding structures . Surgery is the cornerstone of treatment and requires complete gross resection because resection quality is the most important risk factor for local recurrence. Debulking surgery is reserved for unresectable chemo-resistant tumors. Several studies have shown the value of adjuvant radiation either postoperatively or preoperatively in lowering the incidence of local recurrence [19,20]. Systematic chemotherapy can be useful when used in a neoadjuvant manner in selected patients with technically unresectable tumors that could potentially be rendered resectable by downsizing.

We decided a primary surgical approach as individual plan in the musculoesqueletical unit´s multidisciplinary team. A single incision was made combining infrainguinal and extraperitoneal approach [18]. Tumor excision was performed in a piece preserving specific organs. The defect of the inguinal ligament was repaired with Bard mesh (Imaging 3A). Intraoperative findings showed a single specimen of 190x130x90 mm encompassing the tumor and involved iliopsoas muscle, achieving macroscopically complete resection (Imaging 3B) and microscopically negative margins. Postoperative radiotherapy was offered in the adjuvant setting in the dose of 6o Gy. Now, our patient has been followed for 24 moths and is free of recurrence, with slight hypoesthesia in the left anterior thigh. After surgery a periodic follow up observation should be performed for the early detection of local recurrence and metastasis. A long-term follow-up with regular chest X-ray and CT scans of the abdomen is mandatory.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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