Giant recurrent retroperitoneal liposarcoma presenting as a recurrent inguinal hernia

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Abstract

Retroperitoneal liposarcoma presenting as an inguinal hernia is a rare entity. We present *the first case of Giant recurrent liposarcoma presenting as a recurrent inguinal hernia* in a 40-year-old male. Physical examination showed an irreducible lump in the right inguinal region and a scar in the right lumbar and right inguinal region. Computed tomography (CT) scan of abdomen revealed it to be a retro peritoneal mass extending into the right inguinal region along and involving the cord structures. Wide local excision of the tumour with right orchidectomy and inguinal hernioplasty was performed. Histo-pathology confirmed it to be a liposarcoma. Patient received postoperative radio therapy. Follow up of two years has shown him to be disease free. Retroperitoneal liposarcoma can grow along cord structures into the inguinal canal and mimic an irreducible indirect inguinal hernia.

Introduction

Liposarcoma is a rare malignant tumor of mesenchymal tissue. The two major sites of liposarcoma are the extremities, particularly the thigh and the retro peritoneum.[1] The retroperitoneal cavity communicates with the inguinal region through cord structures, and hence retroperitoneal lipo sarcomas can occasionally extend through the inguinal canal into the scrotal sac, presenting as an indirect inguinal hernia or through the obturator foramen into the thigh. Lipo sarcomas in the retro peritoneum grow slowly and present with complex, usually late and non-specific symptoms.[2,3] We present a rare case of a giant recurrent retroperitoneal liposarcoma presenting as an irreducible inguino scrotal hernia in a previously operated case of right-sided indirect inguinal hernia.

Case Report

A 40-year-old male presented with a swelling in the right inguinal region of six months duration. The patient had been previously operated for a retroperitoneal tumour eight years ago in a different hospital and was diagnosed to be liposarcoma according to his family physician's note. However, there were no details about the level of resection with respect to margins. Patient did not follow up with the previous hospital as he had changed his residence. He also had history of a right inguinal hernia repair three years ago. Physical examination showed a lump in the right inguinal region, which was irreducible and a scar in right lumbar and inguinal region. (Figure 1). This lump was in continuity with a large lumbar mass. Hematologic investigations were within normal limit. Computed tomography (CT) scan of abdomen revealed a 21×14×12 cm ill-defined mass in the retro peritoneum with fat attenuation, pushing the right kidney and ureter anteriorly, and extending into the right inguinal region. The right scrotum also had a mass with the same attenuation as in the retro peritoneum (Figure 2). FNAC showed liposarcoma. Tumor marker LDH was 503 IU/L (normal 100-190 IU/L) and CEA was 3.65 ng/mL (normal 0-2.5 ng/ mL). Exploratory laparotomy revealed a huge retroperitoneal mass with well defined margins extending into the right scrotum through right inguinal canal and involving the cord structures. Wide local excision of the tumor with a resection margin of more than 20 mm along with right orchidectomy was performed. Right inguinal hernioplasty was carried out to repair the defect.

Grossly the tumor measured 47×14×12 cm and weighed 12.7 kg (Figure 3A). Cut surface of the specimen showed yellow lobulated fat tissue (Figure 3B). Microscopically revealed areas of mature adipose tissue containing, however, atypical lipoblasts with cellular fibrous septa suggestive of well differentiated liposarcoma (Figure 4). Histopathology confirmed tumor free margins *i.e.* R0 resection. The patient received postoperative radio therapy and is asymptomatic after two years of follow up.

Discussion

Liposarcoma are rare tumors of mesenchymal origin, with the most common sites being the extremities and retro peritoneum.[1] Liposarcomas account for less than 0.1% of all human tumors as per literature.[2,3] Retroperitoneal liposarcoma is primarily a tumor of adults with a peak incidence between 50 and 65 years. It accounts for at least 10-12% of all sarcoma in adults with male predominance but no racial predominance. The neoplasms are de novo malignant, with only a few cases originating from benign lipomas. The dominant sites of origin are reported as the extremities (particularly the thigh at 13-60%) and the retro peritoneum (10-36%).[1]

Retroperitoneal liposarcoma is among the most common primary retroperitoneal tumors, along with malignant fibrous histiocytoma and leiomyosarcoma.[4,5] It has three principle forms: i) atypical lipomatous tumor well differentiated (50-55%); ii) myxoid-round cell variety (40%); and iii) pleomorphic (5%).[3,4] Most patients with retroperitoneal liposarcoma present with an asymptomatic abdominal mass, often after the tumor has reached a considerable size.[3,6] Diffuse abdominal pain is the most common symptom present in 50% of the patients and less common symptoms include early satiety, nausea, vomiting, weight loss, lower extremity swelling and GI hemorrhage. Our patient presented with a mass lesion.

Retroperitoneal tumors usually grow slowly to a larger size due to absence of anatomical barrier in the retroperitoneal cavity. As retroperitoneal space communicates directly with the pelvic cavity and inguinal region, the retroperitoneal tumor tends to extend into the pelvic cavity and inguinal region.[5] These tumors follow the gonadal vessels through the retro peritoneum into the inguinal region. This is further augmented by the deep ring which is a defect in the fascia transversalis through which cord structures enter the inguinal region. Hence the presence of deep ring and cord structures entry through it provide a potential pathway through which retroperitoneal sarcomas enter the inguinal region and mimic inguinal hernias.

CT and magnetic resonance (MR) imaging provide information about the size, extent of

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the tumor mass and its relation to major vascular structures, and also the presence of metastatic disease. In the present patient, CT scan showed a large lobulated fat attenuation tumor within the retro peritoneum, which displaced nearly entire bowel loops superiorly. It was extending through the right inguinal region along the cord structures into the scrotum.

Well differentiated lipo sarcomas are low grade malignancies with a tendency for local recurrence but no distant metastases, while poorly differentiated malignancy carries increased risk for distant metastases.[7,8] Lymph node metastases by sarcoma are very rare (<5%). Well-differentiated lipo sarcoma can have two components: lipoma-like and sclerosing subtypes.[3,4] Prognostic factors include tumor size, histology and radiological density as per Reiten *et al.*[3]

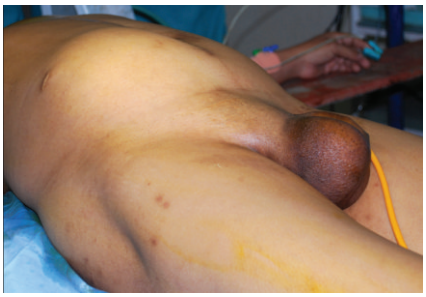

Figure 1. Photograph showing lump in right lower abdomen extending into right inguinoscrotal region.

Complete surgical resection is the most effective treatment for primary or recurring retro peritoneal liposarcoma. Local recurrence after complete resection of sarcoma is very common, usually in the tumor bed. One study has demonstrated that resection margins over 10 mm decreased risk of local and distant recurrences in patients with soft tissue sarcoma.[9] In our case, we had a resection margin of more than 20 mm. The role of adjuvant chemotherapy and radiotherapy is controversial.[10] Some studies have shown that 5-year local control of 51-71% was achieved with radiotherapy for retroperitoneal sarcomas,[11,12] but none of the studies supported an increased survival.[10] Also there is no evidence that chemotherapy increases survival.[10]

Between 10-50% of retroperitoneal sarcomas are resectable and the local recurrence occurs in 40-80% of cases.[2] Close follow up is recommended due to high rates of recurrence.[10] In the present case, the patient underwent exploratory laparotomy with wide excision of tumor with orchidectomy and herniaplasty. Since our patient presented with a recurrent disease, he received postoperative radiotherapy and is asymptomatic after two years of follow up.

A literature review showed few cases of retroperitoneal liposarcoma presenting as an indirect inguinal hernia[1] or as spermatic cord tumor.[13] However, there has been no report of recurrent liposarcoma presenting as recurrent inguinal hernia. This shows that even after a posterior wall repair in a hernia, there is always a potential defect along cord structures which allows retroperitoneal tumors to grow. This fact is highlighted by our present case.

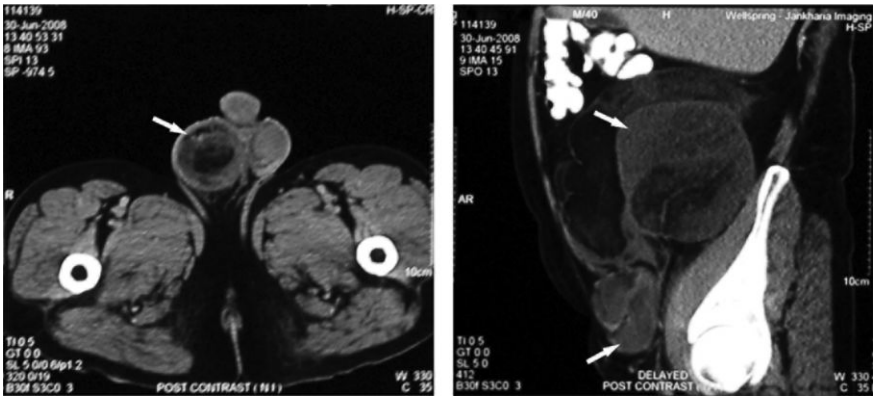

Figure 2. Computed tomography (axial and sagittal view) showed ill-defined mass of retro peritoneum (arrow) with fat attenuation, pushing the right kidney and ureter anteriorly and extending into the right inguinoscrotal region.

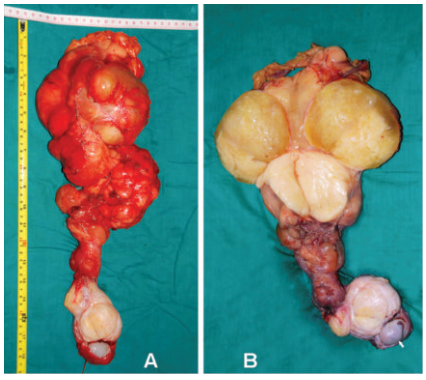

Figure 3A and B. Specimen showed yellow lobulated retroperitoneal fat tissue extending along the spermatic cord into scrotal sac.

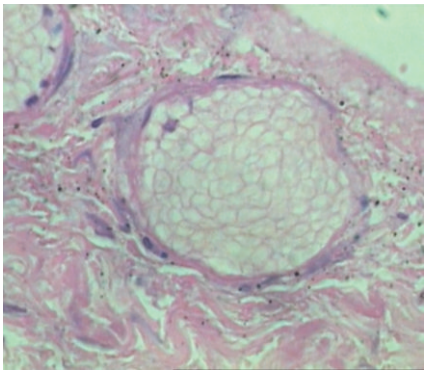

Figure 4. Histology reveals mature adipose tissue with atypical lipoblasts in areas containing cellular fibrous septa.

Conclusions

Retroperitoneal liposarcoma can be one of the differential diagnoses in a case of a recurrent inguinal hernia as cord structures provide the potential opening for these tumors to extend into the inguinal region.

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