รายงานผู้ป่วย

Giant Retroperitoneal Liposarcoma Presenting as Indirect Inguinal Hernia

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Abstract **Giant Retroperitoneal Liposarcoma Presenting as Indirect Inguinal Hernia.**
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The rare case of giant retroperitoneal liposarcoma with scrotal sac extension was herein reported in a 56 year-old man. The initial symptoms began with abdominal distension subsequently left inguinal and scrotum enlargement. The ultrasonography demonstrated diffuse echogenic abdomen and nearly entire fluid-filled bowel loops were displaced into upper abdomen. The computed tomography revealed a huge fat attenuation tumor occupied in the retroperitoneum and extended to left sided pelvic cavity, left inguinal region and left scrotum. Intraoperative found rather well defined tumor within retroperitoneal space that extended through the left inguinal canal into left scrotum. The tumor weighing 11 kg was excised. Left herniorrhaphy and left orchidectomy were also performed. Histopathological diagnosis showed liposarcoma of the retroperitoneum; well-differentiated subtype. The patient has received neither radiation nor chemotherapy after surgery.

Introduction

Liposarcoma is a rare malignant tumor of mesenchymal origin. Its incidence varies from 10-12 percent of all soft tissue sarcoma between fifth and seventh decades. The two major sites of liposarcoma are the extremities, particularly the thigh and the retroperitoneum which account for about two-third of all cases[1]. Because of the inguinal region communicates directly with the retroperitoneal cavity, retroperitoneal liposarcomas occasionally extend through the inguinal canal into the scrotal sac or through the obturator foramen into the thigh[1]. Liposarcomas in the retroperitoneum are usually slow to be recognized and at the time of operation tend to be of much larger size those than in the extremities. Its clinical presentation characterized by symptom which usually late and nonspecific[1, 2]. The author reports the case of a huge retroperitoneal liposarcoma, which extends through left inguinal canal into the left scrotum.

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Case Report

A 56 year-old man was referred to our hospital with abdominal distension for 1 year that has been growing in size without pain. In 3-4 months before admission, his left inguinal region and left scrotum has been enlarged with palpated mass. The mass had gradually continued to grow since he first discovered it. The physical examination discovered mark distension of abdomen, soft, no tenderness. The left inguinal was swelling and palpated left scrotal mass with cystic consistency. (Fig 1)

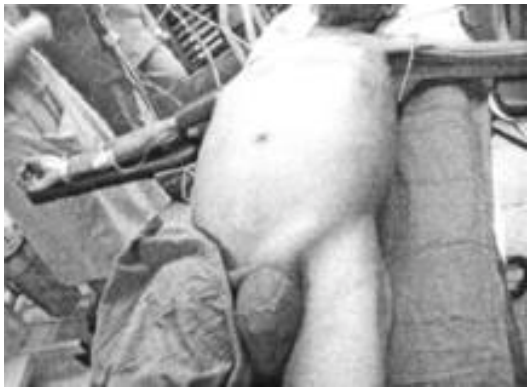


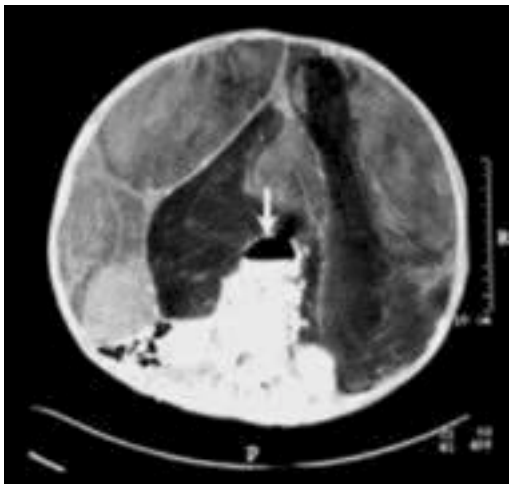
Fig 1 View the patient with abdominal distension and left scrotal mass.

This mass was suspected of being an inguinal hernia in left groin. There was abnormal liver function test and high tumor marker level (LDH=712 U/L, normal 313-618 and CEA=60.73 ng/ml, normal 0.14-6.05)

The chest x-ray and plain supine abdomen illustrate elevated hemidiaphragms and totally opaque abdomen. The ultrasonography showed diffuse echogenic abdomen and multiple fluid filled bowel loops displaced to upper abdomen. (not shown)

The computed tomography of the whole abdomen with oral and intravenous contrast revealed a huge heterogeneous mass with predominantly fat attenuation occupying in the retroperitoneum which extended to left sided pelvic cavity, left inguinal region. The left scrotum also had mass with the same attenuation as in the retroperitoneum. Its size was measured at about 14x26x35 cm. This tumor also had internal thickened, irregular septa and minor nodular components with attenuation on CT scans approximating that of skeleton muscle. This tumor displaced nearly entire bowel loops into upper abdomen. (Fig 2 A-B), (Fig 3 A-D)

2 A



2 B

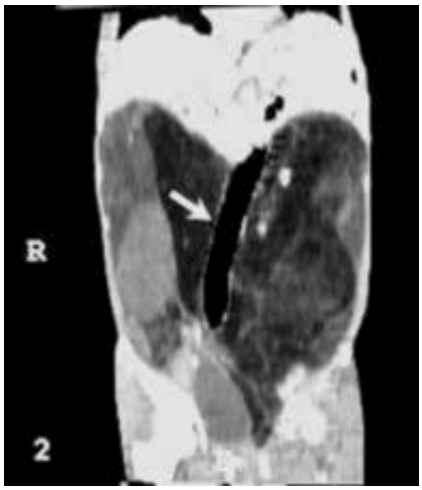
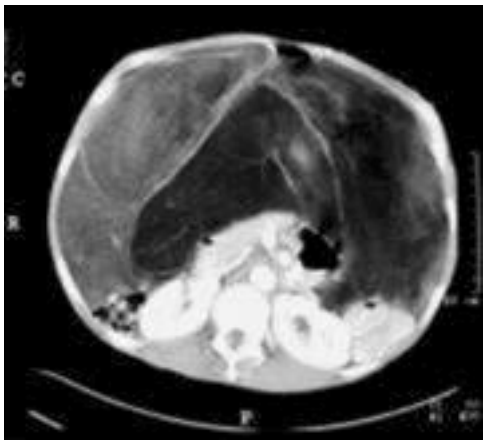
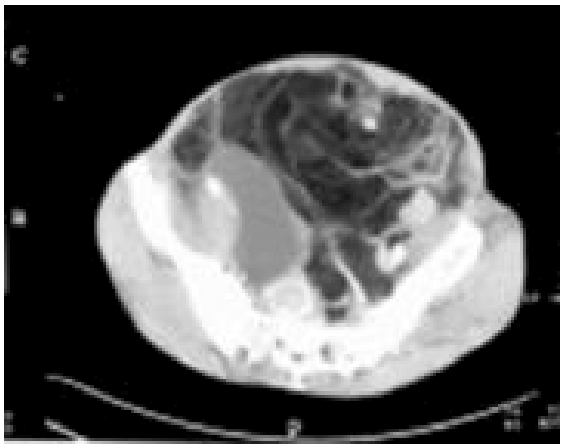


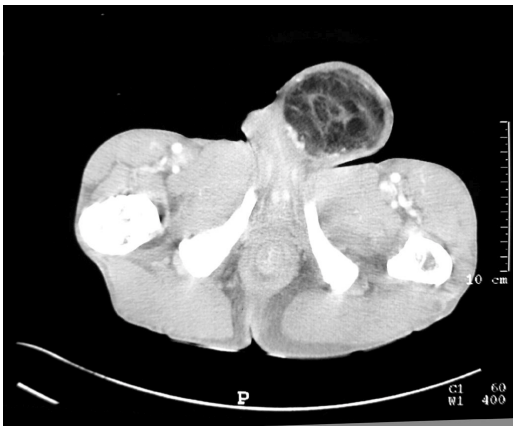
Fig 2 (A and B) The computed tomography (axial and sagital views) reveals huge tumor which has pressure effect to the descending colon (arrow) (antero-medial displacement of descending colon), determine whether the tumor is locating within the retroperitoneal space.



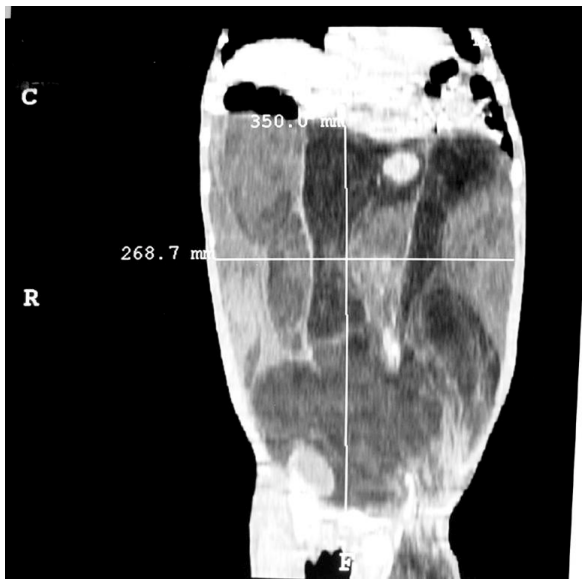
3 A



3 B



3 C



3 D

Fig 3 (A, B, C and D) The computed tomography (axial and coronal views) of the whole abdomen including inguinal region and scrotum shows a huge heterogeneous tumor with predominantly fat attenuation occupying in the retroperitoneum which extended to left sided pelvic cavity, left inguinal region and into the left scrotum

At laparotomy, rather well defined-margin of huge tumor was found within the retroperitoneal space, which displaced the entire bowel loops to the upper part of abdomen. (Fig 4) This tumor was extended to left pelvic cavity through left inguinal canal into the left scrotum. The tumor was resected as well as left herniorrhaphy and left orchidectomy. The gross removed tumor measured 14x30x35 cm in dimensions and covering weighed 11 kg. (Fig 5)

Fig 4 Intraoperative reveals rather well defined–margin of huge tumor within the retroperitoneal space.

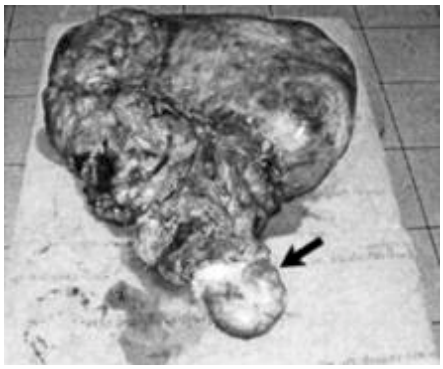


Fig 5 The excised tumor weighed 11 kg shows large retroperitoneal part connecting with small scrotal part (arrow).

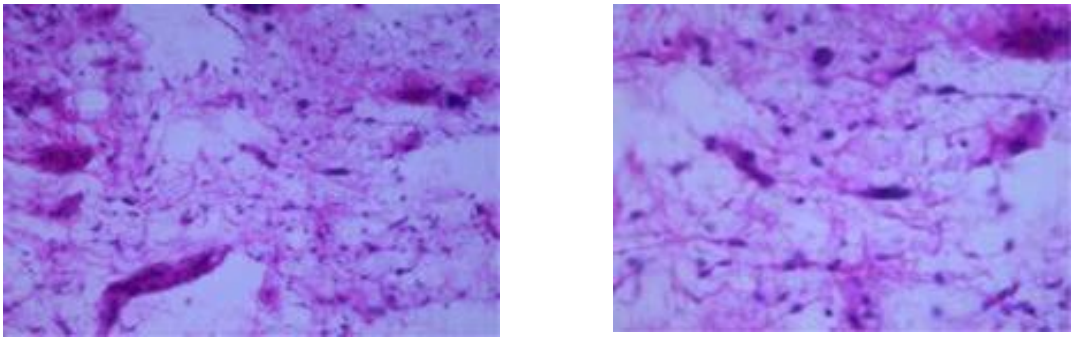


Fig 6 The microscopic findings, low power (A) and high power (B) show areas resembling mature adipose tissue but containing cellular fibrous septa with atypical lipoblasts.

The cut surface of the tumor was showed yellow and brown lobulated fat. The microscopically revealed lipoblasts. (Fig 6) A–B The left testis had few spermatogenesis. The spermatic cord was hemorrhage. The stump was free from tumor. The histopathological diagnosis was well–differentiated liposarcoma, according to the WHO classification.

No adjuvant therapy was performed. One month after surgery this patient was admitted in the hospital again with cachexia and after discharged he was loss from the medical record.

Discussion

According to literature liposarcomas account to less than 0.1 percent of all human tumors[1,7] . However liposarcoma is one of the most common soft tissue sarcomas found in adult. The dominant sites of origin are reported as the extremities (particularly the thigh comprising 13–60 percent) and the retroperitoneum (comprising 10–36 percent)[2].

Retroperitoneal liposarcoma is among the most common primary retroperitoneal tumors, along with malignant fibrous histiocytoma and leiomyosarcoma[7,8,9]. It is slow growing and has a propensity to displace rather than invade adjacent structures[9].

Because of there is no anatomic barrier

within the retroperitoneal space. They are usually slow to be recognized and at the time of operation tend to be of much large size[5,7]. In additional, the retroperitoneal space communicates directly with the pelvic cavity and inguinal region[2,4]. So the retroperitoneal tumor tends to growth into pelvic cavity and inguinal region.

However the author traced only few papers in the literature reported about retropeitoneal liposarcoma with inguinal/scrotal extension; Serdar Y. et al, reported a case of retroperitoneal and scrotal giant liposarcoma[1]; Hirofumi N. et al, reported a case of retroperitoneal liposarcoma presenting a indirect inguinal hernia[2]; Maj. Jacqueline H. Longbotham. et al, reported a case of retroperitoneal liposarcoma presenting as spermatic cord tumor[3] ; Elizabeth M. et al, reported incidental liposarcomas identified during hernia repair operations[4].

To my knowledge, this is the first report of a huge retroperitoneal liposarcoma with left inguinal and scrotal sac extension.

The computed tomography and magnetic resonance imaging (MRI) can demonstrate important characteristics of these tumors; diagnostic is often challenging for the radiologists. Diagnostic challenges include precise localization of the lesion, determination of the extent of invasion and characterization of the specific pathologic type. They also allowed us to detect residual tumor and recurrence[1,6].

The first step is to determine whether the tumor is located within the retroperitoneal space. Displacement of normal anatomic structure of the retroperitoneum is helpful in this regard[3,6]. In this patient, large lobulate tumor was displaced descending colon antero-medially; strongly suggests that the tumor arises in the retroperitoneum.

The next step is to identify the organ of origin. Before a tumor can be described as primarily retroperitoneal, the possibility that the tumor originates from a retroperitoneal organ must be excluded. There are some radiographic signs helpful in determining tumor origin such as **"beak sign"** *, the **"phantom (invisible) organ sign"** **, the**"embedded organ sign"** ***and the **"prominent feeding artery sign"** ****[6].

The herein-reported case showed no definite sign that suggests an organ of origin, the diagnosis of primary retroperitoneal tumor becomes likely.

To determine extension of tumor, CT and MRI were the most useful methods. In this patient, CT was performed which showed large

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* *Beak sign = when a mass deforms the edge of an adjacent organ to a "beak" shape, it is likely that the mass arises from that organ.*

** *Phantom (invisible) organ sign = when a large mass arises from a small organ, the organ sometimes becomes undetectable. This is known as the phantom organ sign.*

*** *Embedded organ sign = when a tumor compresses an adjacent plastic organ (eg. Gastrointestinal tract, inferior vena cava) that is not the organ of origin, the organ is deformed into a crescent shape.*

**** *Prominent feeding artery sign = hypervascular mass are often supplied by feeding arteries that are prominent enough to be visualized at CT or MR imaging, a finding that provides an important key to understanding the origin of the mass.*

lobulate fat attenuation tumor within the retroperitoneum, which displaced nearly entire bowel loops superiorly. It extended downwardly to left side pelvic cavity and left inguinal region. There was the same attenuation tumor seen in the left scrotum as seen in the retroperitoneum.

Familiarity with the specific features of various retroperitoneal tumors often allows accurate histological diagnosis and helps suggest proper management[6].

Some tumor contents can be clearly demonstrated at CT and MR imaging and provide strong clues that help narrow the differential diagnosis[6]. In this report, tumor contains predominantly fat attenuation that helps narrow the differential diagnosis of fat tumor. There were some irregular and high density of septa and nodules within the tumor which shown heterogeneous slightly increased enhancement after contrast study. So the diagnosis of liposarcoma should be considered.

The liposarcomas are classified into four histological subtypes; well-differentiated, myxoid, pleomorphic and round-cell subtypes[1,8]. The accurate classification of liposarcomas is important because of the differences between types in biological behavior and prognosis[2,5].

Tonsok K. et al[8] concluded that each histological subtype of abdominal liposarcoma showed different CT attenuation or MR imaging signal intensity characteristics. A clear under-standing of these findings should prove helpful in the diagnosis of liposarcoma.

The radiographic findings of this tumor should be categorized as well-differentiated liposarcoma. Well-differentiated liposarcoma can have two components; lipoma-like and sclerosing subtypes[7,8]. Radiographic evident fat comprises greater than 75 percent of well-differentiated liposarcoma. It tend to have smooth margins, a lobular contour and a predominate attenuation of fat. Internal septa of soft tissue attenuation are usually evident. The septa tend to be over 3 mm thick, are often nodular and demonstrate mild to moderate enhancement, helping distinguish them from benign lipomas[7,8].

At laparotomy, rather well define-margin of huge tumor was found within the retroperitoneal space, which displaced the entire bowel loops to the upper part of abdomen. This tumor was extended to left sided pelvic cavity through left inguinal canal into the left scrotum. The patient underwent an en-bloc excision of the tumor by abdominal route. The tumor was excised as well as left herniorrhaphy and left orchiectomy. The removed tumor measured 14x30x35 cm in dimensions and weighed 11 kg. The cut surface of the tumor was shown yellow and brown lobulate fat. The histopathological diagnosis showed well-differentiated liposarcoma, according to the WHO classification. The stump was free from tumor.

The aim of surgical treatment is to archive a wide resection margin, removing any adjacent organs and structures involved. In the literature, 10-50 percent of retroperitoneal sarcomas are respectable and the local recurrence rate ranges from 40-80 percent[1].

In our case, we didn't perform any adjuvant therapy. This was because the tumor consisted of well-differentiated liposarcoma, wide resection of the adjacent tissue was

performed and no malignant cells were seen histopathologically in any of the surgical margins. And the effectiveness of radiation and chemotherapy is sill controversial[2].

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