

Retro-Peritoneal Liposarcoma: Diagnostic Difficulties and Therapeutic Attitudes

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Abstract

Background

Retro-peritoneal liposarcoma is a rare tumor which preoperative diagnosis and staging are essential for planning its management.

Methods and results

Case 1: A 60-year-old woman complaining of jaundice. Ultrasonography and Computed tomography revealed a dilatation of the common bile duct and the wirsung upstream of a tumor of the pancreatic head. The tumor involved the entire upper retroperitoneal space. The lesion was unresectable. A choledocoduodenal anastomosis and a biopsy of the lesion were done. Histo-pathological examination was indicative of a well differentiated liposarcoma. A post-operative chemotherapy has been done.

Case 2: A 75-year-old man complaining of abdominal distension. Ultrasonography and Computed tomography revealed a huge tumor that involved all the right retroperitoneal space associated with two intra peritoneal lesions. At surgery, a right hemi colectomy and a right nephrectomy were performed for complete resection of the tumor. The final histopathological report showed undifferentiated liposarcoma of the retro peritoneum.

Case 3: A 55-year-old lady who had six iterative laparotomy and iterative resections of a retroperitoneal liposarcoma. This attitude has prolonged survival over eight years despite five recurrences.

Conclusion

Total surgical resection provides the patient best chance for cure. It may extend to the adjacent organ for an R0 resection. Recurrences require multiple resections or multi organ resection.

Keywords: Retro-peritoneum; Liposarcoma; Treatment; Recurrence

Introduction

Liposarcoma are neoplasms of mesodermic origin derived from adipose tissue or becoming from mesenchymal multi potent stem cells with special proliferation of lipoblasts [1]. It's the single most common soft tissue sarcoma which corresponds to 10–14% of all them [2,3] and the most common retroperitoneal sarcoma [2]. Histologically, they are subdivided into four subgroups based on morphology and cytogenic abnormalities: well differentiated (WDL), dedifferentiated (DDL), myxoid/round cell, and pleomorphic [4-7]. This tumor raises several diagnostic and therapeutic problems. Preoperative diagnosis and staging are essential for planning its management. These data are mainly provided by the CT which must be carefully interpreted. Complete surgical excision is the gold standard for treatment of liposarcoma. Recurrences are frequent and often require a more aggressive surgical approach, including multiple resections or multi organ resection.

We experienced three cases of retroperitoneal liposarcoma treated at our medical institution. We discuss modalities of preoperative diagnosis, surgical procedure for primitive tumor and for recurrence.

Case Presentation

Case 1

A 60-year old lady, without significant pathological antecedents, presented with jaundice for the last two months, which was insidious in onset and gradually progressive in nature. At local exam, we found a palpable, not clearly delimited, lump of 15 × 10 cm occupying left lumbar, and left iliac fossa. Blood analysis results revealed hyperbilirubinemia at 175 mg/l.

Ultrasonography of abdomen showed, first, stones in gallbladder with marked dilatation of the common bile duct and the wising. Second, an echogenic and heterogeneous lesion (Figure 1A) occupying the left retroperitoneal area nearby vertebral column and iliac vessel. This tumor measured 7 × 4 cm. All other organs appeared normal.

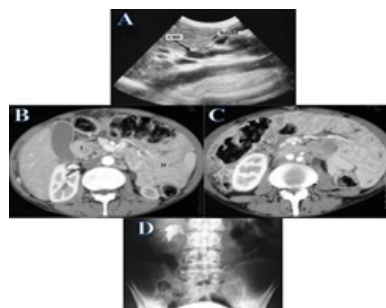


Figure 1: Marked dilatation of common bile duct upstream an echogenic lesion of the head of pancreas in ultrasonography (A). On CT-scan: Tumor involved the head of the pancreas (I), extended on either side of the lumbar column and involved all the upper retroperitoneal space (II) (B). Tumor involved left kidney and left suprarenalian gland (C). Intravenous urography showed a left renal mutism (D).

Contrast Enhanced Computed Tomography of abdomen revealed a dilatation of the common bile duct and the wirsung upstream of a tumor of the pancreatic head. The tumor was extending on either side of the lumbar column and involved all the upper retroperitoneal space (Figure 1B). It was a low density and heterogeneous lesion with multiple linear septa. This lesion was causing compression and displacement of adjacent bowel loops. This tumor seems to involve left kidney, the left lumbar ureter (with no renal secretion and excretion) and left suprarenalian gland without possibility to delimitate it from the tumor (Figure 1C). Intravenous urography showed a left renal mutism (Figure 1D).

Surgical intervention was transperitoneal with retroperitoneal access after left parietal-colic detachment. The tumor was giant, encompassed the entire left retroperitoneal cavity from pancreas to the pelvis involving, too, with close adhesions to the descending colon, left kidney and ureter and adherence to the aorta and iliac vessels. In the right, the tumor involves the cephalic part of the pancreas, inferior vena cava and mesentery. Structure was lipomatous with a thin capsule around the most part of the tumor.

There were also two nodules on the gastro-colic ligament and on the transverse colon. The lesion was unresectable. A choledocoduodenal anastomosis and a biopsy of the lesion were done.

Histopathological examination was indicative of a well differentiated liposarcoma. Post-operative course was uneventful and the patient was discharged on the 5th postoperative day. A post-operative chemotherapy has been done. Patient died nine months later.

Case 2

A 75-year-old man presented with complaints of painless distension of abdomen for the last six months, which was insidious in onset and gradually progressive in nature. There was history of weight loss for the past 6 months.

Local exam discover two homogenous abdominal tumor of 15 cm size, occupying umbilical region. Rest of systemic examination was within normal limits.

Abdominal ultrasonography showed a 15 cm diameter tumor on the right side of the abdominal cavity from the abdominal centromedian. The lesion was hypo echoic and hetero genius. Tumoral process involved, too, right kidney lodge with posterior translation of the right kidney and compress inferior vena cava. The second lesion had the same characteristics of the previous and was located on the left side of the abdominal cavity and seems to be intrapérítoneál.

A CT scan detected a 24 × 23 × 13 cm huge tumor that involved all retroperitoneal space from the right lobe of liver to the pelvic level with the right kidney, liver, cephalic portion of the pancreas and the right colon significantly displaced to the left side (Figure 2). There were two other lesions located on the para-umbilical region strongly enhanced that suggests secondary locations. We assumed it was a retroperitoneal lipomatous tumor with intra-peritoneal carcinosis and decided on surgical exploration.



Figure 2: On CT-scan: Huge tumor (T) involved all the right retroperitoneal space with a marked displacement of the right kidney (K), cephalic portion of the pancreas and the right colon (C) to the left side.

The surgery revealed first two intra-peritoneal tumors. The first, adjoining the omentum was resected. The second involved the ascending colon which excision had required a right hemi-colectomy (Figure 3A). The right retroperitoneal tumor was giant, multi-lobular and uniformly yellow. Because the tumor grossly encased the right kidney, we performed a nephrectomy for complete resection of the tumor (Figure 3B).

The surgical specimen (Figure 3C) measured 30 × 25 × 6 cm and weighed 7 kg.

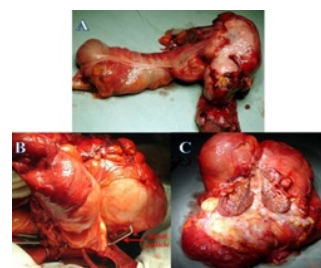


Figure 3: A) Right hemicolectomy was mandatory to respect the entire tumor. B) Right nephrectomy was mandatory for complete resection of the tumor. C) Surgical specimen of retro-peritoneal liposarcoma containing right kidney.

The final histopathological report showed undifferentiated liposarcoma of the retro-peritoneum. Postoperative evolution was simple. He declined adjuvant therapy. Currently, at 12 months of follow-up, the patient is asymptomatic and disease free.

Case 3

A 55-year old lady underwent a laparotomy in another centre in 1995 for a giant retroperitoneal liposarcoma with an intimate contact with the left ureter. She had a large tumor “en bloc” resection with the left kidney. Histopathological examination was indicative of a well-differentiated liposarcoma. She didn't have adjuvant therapy.

She was re-operated three years later for a left retroperitoneal recurrence associated with peritoneal nodules nearby the recto sigmoid junction. Tumor excision with sigmoid resection was made.

She was admitted in our department one year later for a new recurrence. Masses were located in supra duodenal area, in the right iliac fossa and in the left retroperitoneum. Tumors resections were made.

Two years later, she underwent a laparotomy for peritoneal carcinosis. Nodules were nearby right kidney, the first duodenum and the stomach. Careful and complete resection of the mass was done and no organ resection was required.

She was re-operated one year later for intra and retroperitoneal recurrence. Tumor resection was made. On post operatively, she had 3 cycles of chemotherapy (Adrea, haloxan).

She was re-operated two years later for a new intra and retroperitoneal recurrence (Figure 4).

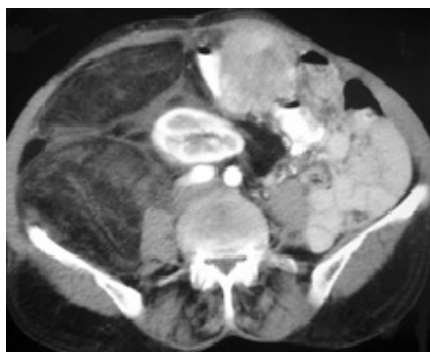


Figure 4: CT-Scan of the 5th intra and retroperitoneal recurrence.

Partial resection was made leaving residual tumor on the mesentery and in contact of the pancreas.

The patient was followed for another 3 months but after that there is no information about him.

Discussion

Location of this tumour in the retroperitoneal area gives it some clinical particularities. The tumor is often deep-seated and large at the time of diagnosis as the retroperitoneum space provides a large potential volume allowing sizeable growth prior to development of signs and symptoms. Thus, clinical presentation is nonspecific and often absent until the last stages of evolution.

Abdominal symptomatology is due to compression of the organs or to local invasion of them. They tend to present with diffuse abdominal pain, urinary symptoms, gastrointestinal obstruction, vessel compression and lower extremity swelling or neurological signs accompanied by anorexia and weight loss and increase in abdominal girth [1,3,8]. Abdominal mass can be detected in almost 80% of cases [3].

EUS can suspect the diagnosis in 30 to 50% of cases showing a heterogeneous and hyperechoic lesion with hypo echoic area signing intra-tumoral hemorrhage or necrosis [9].

However, preoperative diagnosis is often made on the abdominal CT scan data. This exam demonstrated the retroperitoneal origin of the tumour, could assess the relationships with other organs, and could discover possible intra-abdominal metastasis or bone invasion [1].

Typically, CT scan showed a heterogeneous lesion, combining a mainly fatty attenuation and muscle densities with thick septa containing higher density nodules [8,10,11].

These radiologic aspects had been found in our three cases, but preoperative diagnosis was established in only the second and third case. The diffuse and infiltrating aspect of the lesion in the first case was unusual.

In cases where doubt exists, CT or endoscopic ultrasound-guided fine-needle aspiration have been advocated by some and can make the diagnosis with high sensitivity [8-10].

During tumor progression, multiple retroperitoneal organs could be invaded. There is compromise of the adjacent organs in 60-85% of the cases [4,3,7,12].

However, complete resection is thus the most important component of treatment and provides a hope of a cure, regardless of tumor size or adjacent organ involvement.

This will be made a dilemma about the extensive surgical resection and its benefit.

Complete excision is often difficult; since the margins are not grossly apparent thus often treatment usually involves an “en bloc” surgery to remove tumor and adjacent viscera for an R0 resection. Kidney is the most interested organ in extended resections (30%) [3]. Colon, duodenum, adrenal gland, pancreas, spleen, and even inferior vena cava may be also interested [13]. In our first case, invasion of mesentery root was against indication of resection. For the second case, extensive resection was interesting all the organs invaded and potentially resectable which has achieved R0 resection. Failure to achieve macroscopic clearance was often due to the size of the tumor. Patrik et al. [3] demonstrated that in liposarcomas >10 cm, complete resection can be carried out in up to 70% of cases; however, in up to 50% of these cases, multiorgan resection is necessary in order to reach this goal. This attitude may prevent remnant tumor tissue and decrease the risk of recurrence [7,9]. These extended resections improve survival. However, morbidity is higher varying from 20 to 30% with a risk of operative mortality of 4 to 11% [2,10]. However, despite complete resection, most patients developed local recurrence [7,14]. According to different publications, the 5-year local recurrence rates are varying from 35-85% [4,9,12] and in some cases, the primary and recurrent retroperitoneal liposarcoma of the same patient showed different histopathologic subtypes [7].

The recurrence interval had a tendency to shorten as the disease recurred again and the recurrence interval was significantly shorter in

recurrent retroperitoneal liposarcoma cases than in primary cases. This finding was demonstrated by the reports of Na JC et al. and Kim EY et al. [7,11]. The median interval to first local recurrence was varying from 24 to 35 months of the initial surgical resection [6,11].

Many factors were incriminating in the tendency of recurrence: de-differentiated subtype [2], tumor size at initial evaluation (>10 cm) [2,3,13] and involved surgical margins [2,3] were associated with a high risk of local recurrence.

Given the prognosis of this tumor is significantly better than other sarcomas, these patients will be included in new resections. This raises the second dilemma of the frequency of these iterative resections knowing that's lead to mortality.

In our third case, our attitude was aggressive and based on resection of all local and iterative recurrence. This attitude has prolonged survival over eight years despite five recurrences. However, the last three resections had consisted on reductions of tumor and not R0 resections.

Our attitude is not unique since other authors have advocated such iterative surgery but it seems that our case is unique in the number of reoperation for recurrence. This attitude had helped prolong survival in these patients relapsed [2,6,7,11].

The high recurrence rate of retroperitoneal liposarcoma after surgery has prompted the investigation of new combined-modality treatment approaches [14].

Radiotherapy and chemotherapy were proposed by some authors but has not demonstrated long-term improvement in survival or specific disease in cases of complete macroscopic resections [3]. There is no prospective randomized controlled trial confirming the potential benefit of radiotherapy that emerges from retrospective studies [2,13].

In some series, this adjuvant treatment extends the interval between recurrences, in others; it reduces tumor volume and leads to R0 resection in neo adjuvant modality [4,10,14].

Conclusion

Because of great tolerability of retroperitoneal space, retroperitoneal liposarcomas had a long asymptomatic evolution that is correlate with their dimensions. Their diagnosis is mainly based on morphological exam whose signs must be known in order to suggest this type of lesion.

Its treatment is mainly surgical, based on a complete resection and if necessary combined with an "en bloc" excision of adjacent organs to achieve an R0 resection as it reduces the risk of recurrence and

improves survival. However, these recurrences are frequent. Each recurrence should be included in a program of iterative resection as this is the only Guarantee of prolonged survival. Radiotherapy and chemotherapy must find their indication in these forms to improve the results of surgery.

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