Recurrent Retroperitoneal Liposarcoma: a Case Report and Literature Review

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Abstract

This is a case report of a 61-year-old male patient, presented with fifth recurrence of retroperitoneal liposarcoma and operated for six times. Literature review on retroperitoneal liposarcoma biology, recurrence pattern, role of surgery in primary tumor, and recurrent tumor, adjuvant therapy, role of multivisceral resection, and palliative surgery.

Keywords Retroperitoneal liposarcoma · Recurrence · Multivisceral resection

Introduction

Liposarcoma is a common type of soft tissue sarcoma [1], which occurs most commonly in the extremities (52%), followed by the retroperitoneal space (19%) [2]. Liposarcoma can be histologically subdivided into five subtypes: well-differentiated liposarcoma (WDLPS), myxoid, round cell, pleomorphic, and poorly differentiated [3]. Generally, round cell, pleomorphic, and poorly differentiated subtypes are regarded as high-grade, while well-differentiated and myxoid liposarcomas are low-grade. Retroperitoneal liposarcoma is the most frequently observed subtype of retroperitoneal tumor, with an incidence of 2.5 per million people annually [4]. It usually occurs between 40 and 60 years of age, and affects the genders equally [1, 5]. Its occurrence within the retroperitoneal space is usually asymptomatic until the tumor is large enough to compress the surrounding organs [6]. The large space of the retroperitoneum permits the tumor to grow to a large size before exhibiting symptoms of compression or infiltration. These include abdominal distension, abdominal pain, gastrointestinal hemorrhage, early satiety, vomiting, and weight loss. In this article, we present a case of multiple times recurrent retroperitoneal sarcoma and review of literature on management.

Case Report

This is a case report of a 61-year-old gentleman, a known case of well-differentiated retroperitoneal liposarcoma, now presenting with fifth recurrence (i.e., for 6th surgery). He has undergone tumor excision with en bloc resection of various organs during the previous surgeries. The details of the same are as below.

He was first diagnosed in 2009 and underwent excision of a 30 × 25 cm retroperitoneal tumor and received adjuvant radiation. He had a recurrence and underwent another excision in May 2011. The second recurrence was excised en bloc along with the left kidney and a segment of the descending colon with primary colocolic anastomosis being done in July 2013. He developed anastomotic leak and operated 1 week later with end transverse colostomy and distal stump closure, which was still present at the present admission. The third recurrence was operated with tumor excision and en bloc splenectomy and posterior abdominal wall resection in July 2015. The fourth recurrence was operated with excision with distal pancreatectomy with proximal duodeno-jejunal short-segment resection anastomosis. Except for the anastomotic leak of the colon during the second recurrence, the rest of the postoperative courses were uneventful. Stoma closure was deferred during the subsequent surgeries as per patient's wishes. Figures 1, 2, and 3 show CT scan images in 2015, 2016, and 2017. The patient presented all these times with symptoms of abdominal distention, abdominal pain, and fatigue. The patient is non-comprehensive to follow-up in postoperative period.

In May this year, he presented with a large abdominal mass and clinical features of IVC compression. PET scan showed a

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Fig. 1 CT scan of the abdomen in 2015 showing retroperitoneal tumor, nephrectomy status, and colostomy

large 28×25 cm fat and soft tissue attenuating retroperitoneal mass lesion compressing the inferior vena cava, displacing D1 and D2 medially with mass effect on it, abutting the right renal vein at the hilum, the uncinate process of the pancreas, and segment IV of the liver, with loss of plane with the gall bladder. There was no metastatic disease.

In view of the present location of the tumor and single-kidney status, after detailed discussion with radiologists, urologists, and cardiovascular surgeons, en bloc excision of the tumor with simultaneous right renal transplant and right nephrectomy (which implied life-long dialysis) and IVC repair/reconstruction was planned; the same was explained to the patient and family. With informed consent, the surgery was performed on 05/06/2017.

Fig. 2 CT scan of the abdomen in 2016 showing retroperitoneal tumor to the left of the aorta

Fig. 3 CT scan of the abdomen in 2017 showing retroperitoneal tumor with loss of fat planes with gall bladder and pushing the duodenum medially

Midline laparotomy along previous incision with left subcostal extension was performed. Postoperative adhesiolysis was done—small bowel injury during laparotomy primarily closed with vicryl 3-0. Stoma was dismantled to gain access to the tumor. Tumor was separated from the liver and the duodenum and portal structures. Iatrogenic rent in the duodenum was repaired. The gall bladder was resected en bloc with tumor as adherent. Fortunately, the tumor was free from the underlying vessels (renal vein, IVC) and kidney, hence was completely resected without the need for resection of any other viscera. The distal colonic stump was identified, and stoma was reconstructed with a colocolic anastomosis. Two drains were placed—one each in the Morrison's pouch and pelvis. Hemostasis was achieved and the abdomen was closed.

Postoperative period was uneventful. Drains were removed on postoperative days 3 and 5. Patient was discharged on postoperative day 9 on normal diet. Figure 4 shows postoperative follow-up in OPD with previous laparotomy scars and colostomy closure. The final histopathology is retroperitoneal myxoid liposarcoma. Table 1 shows recurrences and surgery performed.

The initial five resection final histopathology reports came as well-differentiated adenocarcinoma, and fifth recurrence specimen report came as myxoid liposarcoma. All the time, we achieve complete resection of tumor en bloc without residual tumor. The patient received adjuvant radiotherapy after first resection as per tumor board decision and did not receive chemotherapy.

Discussion

Liposarcoma is the most common retroperitoneal sarcoma. It accounts for more than 20% of all sarcomas in adults and up to

Fig. 4 Postoperative follow-up in 2017 showing previous laparotomy incisions and colostomy closure

41% of all retroperitoneal sarcomas [7, 8]. Liposarcoma is biologically different from other sarcomas. Retroperitoneal liposarcoma can represent a more indolent subtype with death occurring almost invariably from local progression. Progressive cachexia and multifocal intestinal obstruction are frequent in this setting [9]. The incidence of distant metastases from retroperitoneal sarcomas reported in the literature ranges from 15 to 34% [4, 5, 8]. Liposarcoma appears to have less of a predilection for distant metastases to both the lung and liver than other histologic types [4, 10]. The rate of distant metastases was 7% in a series published, even when considering high-grade lesions. The principle of eradication of local disease and the effect of local treatment would seem to have more of an impact in this particular subtype. It would be reasonable to presume that a debulking or palliative procedure could potentially postpone the development of a critical tumor mass and its subsequent fatal effects.

Complete surgical resection remains the mainstay of therapy for retroperitoneal liposarcoma. Effective chemotherapy regimens have not been identified and external beam radiation therapy is limited by toxicity to adjacent normal structures. Advances in intraoperative radiation therapy designed to limit exposure of normal tissues have not resulted in improved survival in these patients [11]. Since presently there is no effective

adjuvant therapy, surgical resection should be attempted for retroperitoneal liposarcoma whenever feasible [12].

The rate of recurrence for a central WDLPS is higher than that for an extremity tumor. A study of 92 WDLPS cases found that 91% of retroperitoneal tumors recurred versus only 43% of extremity tumors. These investigators also noted that in 11 patients, dedifferentiation occurred on recurrence. Evans [13] found that LPS, especially the retroperitoneal location and (dedifferentiated liposarcoma)DDLPS histology, recurred frequently and often multiple times (usually <5 but up to 12 recurrences). Factors effecting recurrence are histology-dedifferentiated liposarcoma and complete or gross total resection (GTR) at the initial operation. Tumor size role in recurrence is controversial. When LPS recurs, it does not always have the same histologic classification as the original tumor. Frequently, WDLPS can recur as a DDLPS and even vice versa. The same holds true for subsequent recurrences. An important prognostic factor for retroperitoneal WDLPS and DDLPS is whether or not the tumor transformed when it recurred. Patients whose tumor became or remained a DDLPS had a worse outcome. Evans [13] found that only DDLPS, but never WDLPS, metastasized hematogenously (median time, 44 months). Local recurrence is best treated with resection. However, especially with retroperitoneal tumors, this can be difficult because of the proximity to vital structures and the frequency of recurrences. One study calculated the growth rate of locally recurrent retroperitoneal LPS and found that only patients with growth rates less than 0.9 cm per month had improved survival after aggressive resection of the local recurrence [14].

Macroscopic complete resection for recurrent retroperitoneal liposarcoma has been recommended [15]. Seventy-five percent of retroperitoneal liposarcoma requires multivisceral resection (MVR). Case series are showing gross tumor resection rate was higher in MVR cases than that in non-MVR cases. The most commonly resected organs are the kidney (32%) and the colon (25%).

It is believed that palliative resection is worthwhile for treating the troublesome symptoms of recurrence in patients who have little chance of gross resection [16]. If there was no possibility of gross resection, palliative resection was performed without multivisceral resection. Partial resections for

Table 1 Year of recurrence and management

Year	Surgery
Oct 2009	Resection + radiation
May 2011	Resection
July 2013	En bloc along with the L. kidney and descending colon
July 2015	En bloc splenectomy and posterior abdominal wall resection
June 2016	En bloc with distal pancreatectomy
	Duodeno-jejunal short-segment resection anastomosis
March 2017	GB resected en bloc with tumor

retroperitoneal liposarcoma should only be performed in patients with intolerable symptoms (e.g., extreme increasing intra-abdominal pressure, grave complications, and in some, emergency conditions). Multivisceral resection should be avoided in patients who have undergone partial resection because this does not result in cure and incurs greater morbidity. Studies are showing that surgical debulking palliates the symptoms and improves the survival [12].

Conclusion

In recurrent retroperitoneal liposarcoma, resection should be attempted whenever feasible, despite the frequent need for multivisceral resection.

Compliance with Ethical Standards

With informed consent, the surgery was performed on 05/06/2017.

Conflict of Interest The authors declare that they have no conflict of interest.

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