

CASE REPORT

GIANT RETROPERITONEAL LIPOSARCOMA: A CASE REPORT

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Abstract: *Background:* Liposarcoma is a neoplasm of mesodermic origin derived from adipose tissue and represents the most frequent histopathological variety of the retroperitoneum. Retroperitoneal liposarcoma may grow to a large size without symptoms. Approximately 20% of the tumours are > 10 cm at the time of diagnosis and may reach extremely giant dimensions.

Case presentation: We present the case of a 58-year-old woman with a giant retroperitoneal liposarcoma. The patient presented at our clinic with diffuse abdominal pain and enormous abdominal distension. She had swellings and redness of the right leg. CT scan revealed a 50 × 25 cm sized enhancing soft mass. Ultrasound guided biopsy was negative for the presence of malignant cells. A double "J" ureteral stent was placed in the right ureter. We performed surgery with complete resection of the tumorous mass without multiorgan resection and with macroscopic free margins. A well differentiated, lipoma-like subtype of retroperitoneal liposarcoma, weighing 13.4 kg, was diagnosed in the histopathological report. At 6 months after surgery a new CT scan was done and there was no evidence of recurrence. Currently, after 12 months of follow-up, the patient is asymptomatic and disease free.

Conclusion: Surgery is the gold standard for treatment of retroperitoneal liposarcomas, well differentiated retroperitoneal liposarcomas, that have a minimal metastatic potential. It is preferable to place a ureteral stent before surgery for minimizing the risk of intraoperative lesions of ureters. Symptoms in lower extremities, such as swelling, redness and others, that are due to compression in the retroperitoneum by giant tumours, can disappear after their surgical removal.

Key words: retroperitoneum, well differentiated liposarcoma, giant tumor, surgery.

Background

Liposarcoma is a lipogenic tumour of large deep-seated connective tissue spaces. Liposarcomas represent neoplasms of mesodermic origin derived from adipose tissue and correspond to 20% of all soft tissue sarcomas [1, 2]. These tumors, which are known to occur in the deep soft tissues, are of primitive mesenchymal cell origin, differentiating into mature adipose tissue and rarely arising from a preexisting lipoma. Liposarcoma is the most frequent histopathological variety of the retroperitoneum [2]. Retroperitoneal liposarcomas alone comprise 0.07–0.2% of all neoplasias [3]. In 42% of cases, liposarcomas occur in the retroperitoneum, 41% in a lower extremity favoring the thigh, 11% in an upper extremity, and 6% in the head and neck region [4]. No race predilection is reported and a slight male preponderance is described. Liposarcoma is primarily a tumor of adults and the average age has been calculated variably at 42 years and 53 years.

Histologically, liposarcomas are classified in five distinct categories: (1) well differentiated, which includes the lipoma-like adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell; and (5) pleomorphic [5, 6]. The anatomic distribution of liposarcomas appear to be closely related to the histological type: myxoid/round cell and pleomorphic liposarcomas have a predilection for the extremities, whereas well-differentiated/dedifferentiated liposarcomas occurs predominantly in the retroperitoneum⁷. These tumors may grow to a large size without any symptoms and about 20% of the tumors are > 10 cm in diameter at the time of diagnosis.

The clinical presentation depends on the site of the tumours, most of which are painless palpable masses. Retroperitoneal liposarcoma presents with inherent characteristics in relation to its deep localization and slow expansive growth. There is compromise of the adjacent organs in up to 80% of the cases [3]. The symptoms of liposarcoma are nonspecific and depend largely on the site of the tumour. Clinically, these tumours tend to present with diffuse abdominal pain accompanied by anorexia and weight loss and an increase in abdominal girth. Most symptoms develop as a result of displacement of the nerves and vessels or as a result of compression of adjacent organs or structures, such as the urinary outflow tract and the gastrointestinal tract [3, 8]. Retroperitoneal liposarcomas have a low metastatic potential and surgery is the gold standard for treatment.

Case Presentation

A 58-year-old woman presented at our clinic with a chief complaint of abdominal mass associated with abdominal discomfort. She had first noticed the symptoms a year before in the form of dyspeptic symptoms accompanied with

of the resection, knowing that extensive en bloc resections might be required to achieve complete excision. Before surgical intervention we sent the patient to a urologist for the placement of a double "J" stent in the right ureter for better identification and to minimize the risk of intraoperative iatrogenic lesions.

We entered the abdominal cavity with the patient in dorsal decubitus through a midline incision made from the xyphoid to the pubis, revealing a giant tumor that encompassed the entire abdominal cavity arising from the right retroperitoneum, displacing the right colon anteriorly and to the left.



Figure 2 – Intraoperative findings. Giant tumour occupying the entire abdominal cavity and displacing the right colon anteriorly and to the left

We founded lax adhesions to the ascending and transverse colon and successfully dealt with them. The tumour was adherent to both ureters, kidneys and retroperitoneal vessels with neovascularization. We managed to find the right dissection plane and to perform a complete resection of the tumor without multiorgan resection and with macroscopic free margins. The patient had a satisfactory evolution and was discharged after 5 days postsurgery without any complication.



Figure 3 – The excised surgical specimen, which was identified on pathological examination as a well-differentiated liposarcoma

The specimen was sent to a pathologist. The final histopathological report showed an atypical lipomatous disease, a lipoma-like subtype of well-

differentiated liposarcoma of the retroperitoneum consisting mainly of gross fat, with dimensions $50 \times 30 \times 25$ cm weighing 13.4 kg.

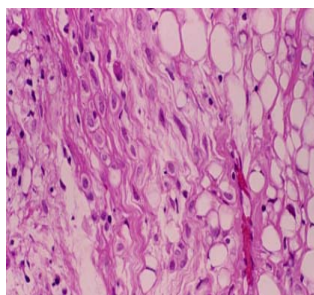


Figure 4 – Liposarcoma, well-differentiated, consisting of lipomatous differentiation in the right field and fibrous area in the left with atypical cells

The patient is being followed up every 3 months. At 6 months post-surgery, a new CT scan was done and there was no evidence of recurrence. Her blood pressure is being stabilized and she has no need of antihypertensive drugs. Swelling and redness of the right lower limb is completely withdrawn. Currently, at 12 months of follow-up, the patient is asymptomatic and disease-free.

Discussion

Liposarcoma is the most frequent histological type of retroperitoneal sarcoma, corresponding to 41% of these tumours [3, 9]. Because there are few clinical findings and specific laboratory abnormalities, these tumours may grow to a large size without any symptoms. Approximately 20% of the tumors are > 10 cm at the time of diagnosis and because of the high degree of adipocyte differentiation it is difficult to distinguish them from normal retroperitoneal fat [8]. The biggest retroperitoneal liposarcoma was reported by Yol *et al.*, weighing 42 kg [10]. Clinically, the symptoms are related to the size of the mass and compression that it makes to the adjacent structures. These tumours present with abdominal symptoms including diffuse abdominal discomfort and pain, accompanied by anorexia and an increase in abdominal girth. The swelling of the lower right limb in our patient was due to the compression of the iliac blood and lymphatic vessels. It is obvious that our patient presented all the signs and disease-specific symptoms due to the size of the abdominal mass.

Our decision, with patient's agreement, was to perform surgery. Complete resection of the tumour was carried out. There are a plenty of reports that describe the experience in the surgical management of primary retroperitoneal liposarcoma and the significance of complete resection of all gross disease in improving local control and disease-specific survival. In most reports, complete

resection can be achieved in up to 70% of cases [2, 8, 11, 12]; however, in up to 50% of these cases, multiorgan resection is necessary in order to achieve this goal [2]. The most frequent organ that needs to be resected is the kidney in 30% of cases [12]. A routine CT that we performed gave us useful information on the positioning and morphology of the tumour, but did not completely resolve whether the vessels or adjacent organs were involved. Determining whether adjacent organs will be attached to or freely separable from the tumour can be difficult based on preoperative imaging. In the case we report here, even with the giant size of the tumour, organ resection was not necessary, because there was no infiltration into neighboring structures or organ involvement. The size of the tumor itself is not associated with surgical resectability.

The ability to completely resect a retroperitoneal sarcoma remains the most important predictor of local recurrence and overall survival [7, 8, 14]. Retroperitoneal liposarcomas represent a distinct category that may justify more aggressive approach including multiple resections for repeated recurrences and even occasionally incomplete resections. Liposarcomas in this location have been observed to have a lower incidence of distant metastases (7%) than that of other histological subtypes of sarcomas (15–34%) [15].

The histology of the mass is vitally important to treatment. In general, liposarcomas are well circumscribed and multilobular. Low grade tumours displaying extensive gross fat are atypical lipomas, that are also referred to as well-differentiated liposarcomas which consist of > 75% of fat and histologically these tumours show irregularly shaped cells with hyperchromatic nuclei called lipoblasts, and lipocytes [16].

With the possible exception of low-grade well-differentiated retroperitoneal liposarcomas, no major survival benefit has been observed when incomplete resection is undertaken [8, 12, 17]. Well-differentiated liposarcomas often recur locally but have minimal metastatic potential, with a 5-year survival probability of 90%. By contrast, the 5 year survival rate of pleomorphic subtypes has been reported to be 50%, for myxoid and round cell subtype 60% and for de-differentiated up to 75% [18].

The use of radiation therapy as adjuvant treatment for retroperitoneal liposarcomas has been discussed, but until now, no randomized trial has evaluated the role of adjuvant radiation. Several retrospective studies have suggested that postoperative radiotherapy may yield better outcomes than surgery alone, although other similarly designed studies revealed no advantage [19]. Although mesodermic tumours are radioresistant, liposarcoma is more sensitive [13, 20]. It has been noted that radiotherapy may increase survival and a disease-free interval [8, 20]; other authors have reported that, in the case of complete macroscopic resection, adjuvant radiotherapy has not demonstrated long-term improvement in overall survival [21]. The large size and extent of these tumours make treatment with tumoricidal doses of adjuvant radiation difficult and are often

associated with substantial morbidity. Postoperative radiation with doses that are more likely to be effective can be associated with acute and delayed bowel toxicity [22]. When the giant tumour mass has been removed from the retroperitoneum, the bowels tend to fall into the resection bed. These fixed loops of bowel are trapped within the radiation treatment volume and even if standard radiation doses of 50 Gy are utilized, gastrointestinal toxicity is significant. There is an agreement for its palliative use in non-operable tumours or in cases of incomplete resections [20, 23].

The role of chemotherapy in the treatment of retroperitoneal sarcomas remains controversial. For the low-grade, well-differentiated LS, adjuvant chemotherapy has little to offer, given the very low mitotic rate of these lesions. The use of adriamycin and ifosfamide for high grade liposarcoma, such as round cell, myxoid and pleomorphic, results in partial responses in up to 50% of patients without improvement in overall survival [24]. The Sarcoma Meta-analysis Collaboration evaluated the impact of chemotherapy in 14 trials that included 1568 patients with resectable soft tissue sarcomas and revealed a modest improvement, with complete response rates to chemotherapy in less than 10% of patients [25]. Complete resection is thus the most important component of treatment, regardless of tumour size or adjacent organ involvement.

From this point, combined with the fact that we achieved complete resection with negative surgical margins and the tumour was of a favourable histopathological grade and subtype, we predict a low rate of local recurrence and good overall survival without adjuvant therapy.

Conclusion

The most important factor in long-term success' and the gold standard for treatment of primary retroperitoneal liposarcomas, is complete surgical margin free resection regardless of tumour size or adjacent organ involvement. The resectability of the tumour and histopathologic grades are associated with disease prognosis. Well-differentiated retroperitoneal liposarcomas represent a distinct situation that may justify a more aggressive surgical approach, including, when necessary, multiple resections for repeated recurrences and even occasionally incomplete resections. Adjuvant therapy has little to offer in increasing the overall survival in patients with a well-differentiated lipoma-like subtype of retroperitoneal liposarcoma.

It's preferable to place a ureteral stent before surgery to minimise the risk of intraoperative lesions of the ureters.

The symptoms in lower limbs, such as swellings and rednesses, that are due to retroperitoneal compression by giant tumours, may withdraw after surgical removal of the tumour.

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Резиме

ГИГАНТЕН РЕТРОПЕРИТОНЕАЛЕН ЛИПОСАРКОМ: ПРИКАЗ НА СЛУЧАЈ

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Вовед: Липосаркома е тумор од мезодермално потекло кој се создава од масно ткиво и претставува најчест хистопатолошки варијетет во ретроперитонеумот. Ретроперитонеалните липосаркоми може да постигнат големи димензии без

да дадат никакви симптоми. Околу 20% од туморите се над 10 cm во моментот на дијагностицирањето и може да постигнат енормно големи димензии.

Приказ на случај: Презентираме случај на 58-годишна пациентка со гигантен ретроперитонеален липосарком. Пациентката се јавила на нашата клиника со дифузна абдоминална болка и енормна дистензија на абдоменот, со отоци и црвенило на десната нога. На КТ е откриена хиподензна сенка со димензии 50 × 25 cm. Ехотомографски водената биопсија беше негативна за присуство на малигни клетки. Дупла „J“ сонда беше поставена во десниот уретер. Пациентката е оперирана со комплетно отстранување на туморот без ресекција на друг орган и со слободни ресекциони рабови. Хистопатолошки е докажан добро диференциран липосарком, со тежина од 13.4 kg. По 6 месеци од операцијата направена е контролна КТ на абдоменот и не се забележани знаци за рецидив. Во моментот, по 12 месеци пациентката е асимптоматична и без знаци на повторно појавување на болеста.

Заклучок: Хирургијата е златен стандард за третман на ретроперитонеални липосаркоми, особено добро диференцираните липосаркоми со минимален метастатски потенцијал. Препорачливо е да се постават уретералните сонди пред операцијата за намалување на ризикот од интраоперативна лезија на уретерите. Симптомите на долните екстремитети, како што се отоци и црвенило кои се должат на компресија во ретроперитонеумот од гигантни тумори, може да се повлечат по хируршкото отстранување на туморот.

Клучни зборови: Ретроперитонеум, добро диференциран липосарком, гигантен тумор, хирургија.

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