CASE REPORT AND THE REVIEW OF LITERATURE

The recurrent primary retroperitoneal liposarcoma

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Abstract: Aim: Describe a patient with multiple recurrences of the primary recurrent liposarcoma.
Clinical case: A 60-years-old man complained of weight loss (BMI 18.4) with a palpable huge retroperitoneal tumour, which displaced left kidney, and was confirmed on USG and CT. Laboratory examination showed anaemia and pathological blood tests. Chest X-ray initially showed a negative finding. A complete transperitoneal surgical extirpation of the tumour with left side nephrectomy was performed on June 28, 2007. The tumour mass weight was 1900 g. It was lying on the posterior face of the kidney in diameters 170x120x120 mm, completely capsulated by thin grey-pink capsula with peripheral fat tissue on the section grey-pink, lobulary shaped, in ⅔ parts with central necrotic changes. Histopathologically was confirmed the primary dedifferentiated (non-lipogenous) liposarcoma low grade of malignancy. Nephrectomy specimen was confirmed as age related finding. There was no evidence of positives surgical margins. Despite oncological and surgical treatment, followed repeated recurrence with eight transperitoneal surgeries in the retroperitoneum and abdomen with extirpation of the metastases, left side hemicolecotomy, splenectomy and repeated extirpation tumour metastases from abdomen and radix mesenterii. Last tumour weighed 2900 grams. Patient died on January 9, 2011, after the eight surgeries on multigons failure due to hemorrhagic shock and persistent atrial fibrillation by cardiopulmonary insufficiency.

Retroperitoneal liposarcoma is an infrequent locally aggressive and recurrent malignancy developed from mesenchymal tissue. We report a seldom case of a huge primary retroperitoneal liposarcoma on the left side, which invaded and displaced the left kidney with multicentric intraperitoneal and retroperitoneal repeated recurrences, adjuvant chemotherapy and eight surgeries.

Case presentation

The 60-years-old man complained of weight loss 10 kg in three month (weight 52 kg, height 168 cm, BMI: 18.4) with some vague symptoms. Physical examination revealed a palpable abdominal mass on the left side. Preoperative USG and CT imaging study showed a huge retroperitoneal tumour. On USG examination, there was seen a clear burdened hypoechoic retroperitoneal mass. CT on June 20, 2007 demonstrated on the left side retroperitoneal tumour 167x124x119 mm, a slightly heterogenous mass with native density +22 to +31 HU, and postcontrast enhancement from +29 to +58 and + 60 HU. Tumour invaded and displaced the left kidney without hydronephrosis (Fig. 1). Laboratory data showed anaemia and pathological blood tests (Hb11.5 dg/l, RBC 3,91x1012/l, PVC 0,34, WBC 15,6x109/l, Tr 426 000, N 87% seg, Eo 2%, Mo 3%, Ly 8%, elevated Fibrinogen 8,55 (N 1.8–3.5 g/l), FW 88/first hour). S-Na140 mmol/l, S-K 4,0 mmol/l, S-Cl 104 mmol/l, S-Creatinine 52,3 μmol/l, GMT 0,64, ALP 2,87, PSA 1,01 ng/ml were within normal limits. Chest X-ray showed initially a negative finding without metastasis (MTS). A complete transperitoneal surgical extirpation of the tumour with left side nephrectomy without adrenalectomy was performed on June 28, 2007. The tumour mass weighted 1900 g. Macroscopically the tumour was lying on the posterior face of the kidney with diameters 170x120x120 mm and was completely capsulated by thin grey-pink capsula with peripheral fat tissue (Fig. 2). On the section, the tumour was grey-pink, lobulary shaped, in ⅔ parts with central necrotic changes. Histopathologically, tumour was confirmed as dedifferentiated liposarcoma (non-lipogenous) low grade of malignancy; nephrectomy specimen was histologically confirmed as age related finding. There was no evidence of positive surgical margins. After oncologist consultation, he received postoperatively no adjuvant therapy. CT three months later on October 3, 2007 showed no MTS. February 11, 2008 on haematology examination was established diagnosis of heterozygous MTHFR congenital thrombophilia without elevation homocystein acids, hyperuricemia, hepatopathia with elevation ALP, deficiency of vitamin K depending factors and Factor VIII, and sideropenia. On February 20, 2008, eight months later, CT
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confirmed first retroperitoneal MTS on the left side which were resected on March 7, 2008 (Fig. 3). Similarly, CT on October 10, 2008 showed recurrence of a new MTS nearby m. psoas and colon descendens. Followed extirpation of this tumour on October 28, 2008. On November 2, 2009 due to new MTS, left hemicolecctomy with tumour extirpation was done. Because recurrence of the liposarcoma continued, the surgical treatment was performed on March 3, 2010. During this surgery, multiplices MTS were found on radix mesenterii and also completely extirpated (Fig. 4). Followed operations on April 8, 2010, then on June 15, 2010, and finally on December 28, 2010 because of excessive huge recurrence in the whole left part of the abdomen and retroperitoneum with tumour weight 2900 grams (Fig. 5). The position of this metastasis was the same as the first time surgical resected tumour in year 2007. Patient died on January 9, 2011, after the eight surgeries on multiorgans failure due to hemorrhagic shock and persistent atrial fibrilaton and cardiopulmonary insufficiency. As a speciality, he

Fig. 1. Preoperative CT with a huge retroperitoneal tumour (arrows) invading the left kidney (A-C). Postoperative CT after first surgery with nephrectomy showed no tumour (D-F).

Fig. 2. Operative preparation – encapsulated retroperitoneal tumour with the left kidney (tweezers).

Fig. 3. Tumour recurrences (arrows) on CT imaging on February 20, 2008 (A-B), and on October 10, 2008 (C-D).
was treated without transfusion because as Jehovah’s witness refused blood derivates. On May 2008, started well tolerated first-line oncological adjuvant chemotherapy only by sole application of Adriblastina (adriamycine) 100 mg to eight cycles to September 2008 due to solitary kidney and restricted calculated GFR 0,810, and renal TRR 0,966. Bone scan on September 2008 was negative, without MTS. Control PET/18 FDG on April 4, 2008 found some small deposits in mediastinium, pulmonary hilum and right supraclavicular region. Similarly, suspect finding was found in skeleton nearby joints without evidence of metabolic metastatic activity. Because of the malignancy continued in recurrence from November 29, 2008 to May 31, 2009, the second-line of the che-

Fig. 4. Repeated recurrency of the liposarcomas on CT imaging on February 22, 2010.

Fig. 5. Huge retroperitoneal metastases on CT on December 10, 2010.
mothers were used. The patient’s treatment included four cycles of Ifosfamid (Holoxan 8000 mg, Uromitexan 8000 mg). Despite recurrence after repeated surgery continued, on June 2009 started the third-line of chemotherapy by application of Dacarbazine 1800 mg as monotherapy. Despite intensive oncological third-line chemotherapy, the patient underwent after an initial operation due to recurrent malignancy totally seven surgeries (Tab. 1) (see list of the operations). Finally, totally thirteen CT studies were performed. CT confirmed osteolytic pelvic and pulmonary metastases. Histopathological examination of the recurrent tumour recurrence operated on June 6, 2010 revealed spindle-cell proliferation set in a variable dense fibrous, collagenous and myxoid stromal tissue with sporadic mixture of the atypical hyperchromatous scattered multinuclear cells with sporadic mitotic activity without lipogenous component. Histological examination confirmed a relapse of the well-differentiated liposarcoma, spindle cell variant and low grade liposarcoma dedifferentiation.

Discussion

With their embryological origin in the mesoderm and in nerve structures of the neuroectodermic layer soft tissue, sarcomas represent only 1.5 % cases in the National Registry of Malignant Tumours in Mexico (Pérez-Ponce et al, 2008). Liposarcoma is a malignancy of fat cells and is the most frequent soft tissue sarcoma localized in the retroperitoneum often with multicentric origin (Sato et al, 2004). Retroperitoneal sarcoma is a rare tumour that accounts for 10–15 % of soft tissue malignancies. The incidence is 2.5 per million and liposarcoma accounts for approximately 20 % of all retroperitoneal sarcomas. As a slow-growing tumour with vague symptoms, the most frequent symptom is nonspecific abdominal pain, sometimes anorexia, loss weight and diffuse abdominal enlargement, but rarely it was presented by another symptoms, e.g. lower gastrointestinal bleeding or hemoperitoneum, with a picture of acute abdomen resulting from a massive tumour haemorrhage (Wanchik et Lucha, 2009; Echenique -Eli- zondo et Amondarain-Arratibel, 2005; Choi et al, 2010; Guzmán Martínez-Valls et al, 1996). Sometimes primary liposarcoma can arise from the renal capsule (Terakawa et al, 2005), and mesente- rium (Moyana, 1988). Rare exists synchronous primary large B-cell gastric lymphoma and huge retroperitoneal liposarcoma with inguinal hernia (Ghimire et al, 2011), retroperitoneal liposarcoma extending into the thigh (Salemis et al, 2011), retroperitoneal li- posarcoma invading an abdominal aortic aneurysm causing rupture (Paravastu et al, 2010), paraneoplastic retinopathy (Kondo et al, 2010), or tumour presenting as an incarcerated femoral hernia (Bognár et al, 2009). Sometimes a suspect huge preoperative diagnosed retroperitoneal liposarcoma can be commute for adenral or retroperitoneal myelolipoma (Nishio et al, 2007; Takahashi et al, 2004). Retroperitoneal liposarcoma can occur with reactive pleural effusion (Funahashi et al, 2004), or well-differentiated liposarcoma of the retroperitoneum and spermatic cord, contigu- ous through the inguinal canal was presented clinically as a scro- tal mass (Longbotham and Joyce, 1987). In literature are reported extremely rare cases of paraneoplastic syndromes or ectopic production of proteins associated with liposarcoma, as a production of Granulocyte-Colony Stimulating Factor, alpha-fetoprotein, paraneoplastic pemphigus and leucocytosis, Acrokeratosis para- neoplastica (Bazex’s syndrome) or cases of retroperitoneal lipo- sarcoma associated with small plaque parapsoriasis (Tartaglia et al, 2007). Liposarcoma can occur in the extremities with unusual pattern of metastatic spread (Vassilopoulos et al, 2001).

If we used the words „abdominal and retroperitoneal lipo- sarcoma“ for searching in Pubmed, we have found more than 70 articles with this topic from 1954 to June 22, 2011. Many of them described a case report or small series on a huge retroperitoneal or abdominal liposarcoma in both, men and women. In some cases, tumour mass occupies the whole retroperitoneum, pelvis and part of the abdomen. Sometimes can reach substantial proportions (Hann et al, 2010; Echenique-Elizondo and Amondarain-Arratibel, 2005; Maámouri et al, 2005; Pascaul Samaniego et al, 2003; Antinori et al, 2002; Farese and Palasciano, 2002; Azpiazu Arnaiz et al, 2000). Removal of a huge retroperitoneal or abdominal mass, sometimes with concomitant resection of adjacent organs were needed, but half of the patients developed tumour recurrence, mainly limited to the retroperitoneum or abdominal cavity. The mean recurrence-free survival was 43.3 months, with 3 and 5-year overall survival rates of 79 % and 61 %, respectively (Fernández- Ruiz et al, 2010). At surgery, pelvic and abdominal organs were displaced and liposarcomas invaded surrounding organs as uterus, adnexa (Smrkolj et al, 2010; Susini et al, 2000), mesenterium, small intestine or colon (Choi et al, 2010) etc. including the right or left kidney, in which cases hemicolectomy, and nephrectomy as in our patient were performed. The resected weight of the tumour reached from some decagrammes, but more frequently nearby from 1000–2000 grams, or some kilograms, sometimes 5.5–6 kg (Izumi et al, 2010; Ebbe, 1993), 15 kg (Tsang et al, 2003) an extremely large liposarcoma weighted 18 kg (Inoue et al, 2005), and 29 kg (Hsiao et al, 1998).

As the first preoperative imaging study, except history and physical examination, was used USG, sometimes with incidentally founding of the liposarcoma, but the best diagnostic tool before surgery in decisive majority was mainly CT, or MRI, with histological confirmation during or after surgery (Shoji et al, 2009; Takahashi

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**Tab. 1. List of the operations and hospitalisations.**

- 28.6.2007 Transperitoneal extirpation of the retroperitoneal tumour with left side nephrectomy, the weight of extirpated specimen was 1900 grams
- 7.3.2008 Relaparotomy, extirpation of the retroperitoneal MTS
- 28.10.2008 Relaparotomy, extirpation of the paraaortic MTS
- 4.11.2009 Relaparotomy, revisio, extirpation MTS in toto, hemicolectomy on the left side, extirpation MTS from scar
- 3.3.2010 Laparotomy, extirpato retroperitoneal multiples MTS and from radix mesenterii
- 8.4.2010 Extraperitoneal MTS extirpation in toto
- 15.6.2010 Relaparotomy, extirpato MTS multiples e retroperitonei l. sin. et radicis mesenterii
- 9.–12.7.2010 Hospitalisation due to subileous status only with infusion therapy.
- 28.12.2010 Extirpation MTS retroperitonei et radicis mesenterii multiples, splenectomy, with tumour weight 2900 grams
- Exitus letalis on Januar 9, 2011
et al, 2009; Song et al, 2007; Funahashi et al, 2006; Kuroasaki et al, 1998; Guzmán Martinez-Valls et al, 1997; Dieckmann et al, 1997).

The histological subtype and margins of resection are the most important prognostic factors for survival in primary retroperitoneal liposarcoma. Sato et al (1999) described nine primary intra-abdominal or retroperitoneal liposarcoma, of which eight were recurrent tumours. Histopathological examination of primary tumours revealed that the number of well-differentiated, pleomorphic and myxoid type was two, four and three respectively. In two recurrent cases, histological differentiation changed from well-differentiated type into myxoid or pleomorphic types. It was seen also by other authors. Spindle cell liposarcoma is presently regarded as a rare variant of well-differentiated liposarcoma, which has the potential for aggressive clinical behaviour (Shioi et al. 2010). Dedifferentiated histological subtype and the need for contiguous organ resection was associated with an increase risk of local and distant recurrence. Nephrectomy may be needed to achieve complete resection was considered by all authors as the treatment of choice.

Sato et al, 2001). The main problem after surgery is local or distant recurrence. Local recurrences are frequent, especially in the first three years, often in the absence of distant metastases. Farma et al (2003) in 4 patients for retroperitoneal liposarcoma performed 15 operations, in 4 patients a second operation was performed for recurrence, in 3 a third operation and in 1 a fourth. Fotiadis et al (2000) described two patients in whose tumour recurred 5 times. In our patient, we have done eight surgeries. When the tumour recurs locally, the best therapy is still to remove the mass. In the treatment of liposarcoma the conventional chemotherapy does not seem effective, while radiotherapy has a little improvement on survival. Generally, the prognosis was poor with overall 5-year survival of 15–50 % (Lauretti et al, 1998). Yoshida et al (2007) in a 53-year-old patient after surgery with multiple unresectable abdominal metastases histologically proven as dedifferentiated, and well-differentiated liposarcoma unsuccessfully used as a first-line therapy (VCR 1.5 mg, ACD 0.5 mg, CPA 900 mg) chemotherapy with disease progression. As second-line chemotherapy, weekly IFM (2g)+CDDP (30 mg) was given. A partial response against peritoneal dissemination was achieved. However, hepatic metastases increased, and the patient died 6 months after surgery. They concluded, that this case suggested that IFM+CDDP might be useful in dedifferentiated liposarcoma. Baratti et al (2010) assessed the outcome of clinically and pathologically homogenous subsets of 37 patients with peritoneal sarcomatosis, from which 13 patients had retroperitoneal liposarcoma (RPLP), uniformly treated by cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) with cisplatin and doxorubicin or mitomycin-C. Overall, the results of CRS and HIPEC did not compare favorably to those of conventional therapy. In a subgroup analysis, the combined approach did not change RPLP natural history. In our patient, the progression of the malignant diseases continued despite third-line chemotherapy, and patient developed new metastases.

Also, our case showed multiple recurrences and surgeries and unsuccessful adjuvant chemotherapy with limited survival. As review of literature showed, curative resection remains the main treatment for primary and recurrent liposarcomas. No standardized guidelines have been established for its treatment because too small series of the patients have been reported and surgical resection was considered by all authors as the treatment of choice. Some success on the field of the adjuvant chemotherapy must be proven by well controlled, blinded, randomized studies.

**Conclusion**

Primary retroperitoneal liposarcoma is a serious malignancy without standardized treatment. This diseases to prejudice QoL because multiple surgeries and unsuccessful adjuvant therapy with unpleasant consequences for patients. Curative resections remains the main treatment for primary and recurrent liposarcomas, and the best therapy is still to remove the mass. Surgical treatment of the primary recurrent liposarcoma without effective adjuvant oncological therapy had limited effectivity.

**References**


