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Case Report

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Cases of giant Retroperitoneal Liposarcomas

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Summary

Aim Liposarcomas are the most common type of retroperitoneal tumours. The course of the disease is determined by histological subtype, grade, size of the tumour and completeness of resection. Surgical treatment is the basic therapy for retroperitoneal liposarcomas.

Case Description Two cases of large retroperitoneal liposarcoma are reported. In both cases, the only sign of the disease was enlargement of the abdomen. Large masses filling the abdominal cavity were detected by ultrasound scans and by computed tomography. In both cases the abdominal organs were involved, with infiltration of the renal capsule, making unilateral nephrectomy necessary.

Results The histology result for the first patient showed a well differentiated liposarcoma while the result for the second patient was liposarcoma arising from the renal capsule. The postoperative course was uneventful and the latest follow-up tests showed no signs of recurrence.

Conclusions Surgical treatment is method of choice for liposarcoma tumours.

Key words **liposarcoma • retroperitoneal tumours**

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BACKGROUND

Liposarcomas are malignant neoplasms deriving from the fatty tissue and are one of the most common soft tissue tumours diagnosed in adults (peak incidence is in the 5–6th decade of life) [1–3] accounting for 0.1% of all malignant neoplasms and 10% of soft tissue tumours [4]. Men are more frequently affected than women (2.5:1). Normally, the tumours are sized 5–10 cm, although cases larger than 15 cm have been described [5]. The most frequent location is a lower limb (50%) though 10–15% of all liposarcomas arise in the retroperitoneal space. Other reported sites include the upper limbs, mediastinum, orbits and oral cavity [6].

CASE DESCRIPTION

Case 1

Patient A.N. (case No. 3496), aged 25, was admitted to the 1st Oncological Surgery Department, Great Poland Cancer Centre, on the 6th of May 2004 for diagnosis and treatment of a giant abdominal cavity tumour. The patient had been observing a gradual increase of the circumference of her abdomen for a year. The patient had not complained of other ailments and had not previously consulted a doctor. On admission, the patient's abdomen was found to be hard and domed above the chest. Bilaterally enlarged inguinal lymph nodes were also observed.

Ultrasound examination revealed that the whole of the abdominal cavity was filled with abnormal masses. Numerous hypoechogenic foci were present, the largest of which measured 83×67 mm and neighboured the lower pole of the right kidney. Changes extended into the pelvis (*p. minor*) where a large hypoechogenic structure had formed, on the left side, whose dimensions were difficult to estimate. The omentum was found to be thickened. A non-homogeneous structure measuring 120×60 mm was located near the navel and found to be hyperechogenic in the centre while being hypoechogenic in the periphery. The liver and spleen were not enlarged and their structures were normal. The left kidney appeared normal while the right kidney was compressed leading to a small degree of widening in the pyelocalyceal system. On the left side, intestinal loops were segmentally widened.

Computed tomography also revealed the presence of growths of varying size occupying the whole of the abdominal cavity. After oral administration of a contrast medium a slight improvement in the images of the changes could be observed. The stomach was retracted and loops of the small intestine, filled with contrast medium, were displaced into the rear section of the left half of the abdominal cavity. The kidneys and spleen were also pushed backward. The pancreas could not be visualized. The kidneys secreted urine normally and the ureters were retracted. The loop of the large intestine could not be localized. The uterus was retracted and pressed on the rectum, nearly touching the sacral and caudal bones. The liver appeared normal.

The results of laboratory investigations were normal and the patient qualified for exploratory laparotomy. This procedure was carried out on the 17th of May, 2004. During the operation, a liposarcoma-like tumour arising from the right renal capsule was discovered. This partly encapsulated tumour, measuring 35×35×25cm and weighing 14 kg (Figure 1), was removed. Histopathological examination revealed that the kidney, measuring 9×5×5 cm and with an attached ureter, was present eccentrically within the tumour. The neoplasm infiltrated the renal capsule superficially. At the upper pole of the kidney the adrenal gland was present, but was separated from the kidney by a layer of tumour tissue. No neoplastic changes were found in the renal parenchyma, pelvis, ureter or adrenal gland. The histopathological diagnosis was of a well differentiated liposarcoma (G1). The postoperative course was without complication. The patient was discharged from the hospital, in good general condition, on the tenth day after the operation. At the most recent check-up, on the 11th of August 2004, the patient showed no signs of recurrence.

Case 2

Patient S.K. (case No. 1142), aged 51, was admitted to the 1st Oncological Surgery Department, Great Poland Cancer Centre, on the 9th of February 2004. The patient was admitted owing to the presence of a tumour mass filling a considerable portion of her abdominal cavity. The results of a needle biopsy indicated the presence of a neoplasm of non-epithelial origin. The patient had been observing a gradual increase in the circumference of her abdomen over a three year period but did not complain of other ailments. On admission, the patient's abdomen was found to be domed over the chest and distended, with tense integuments. In a standing posi-

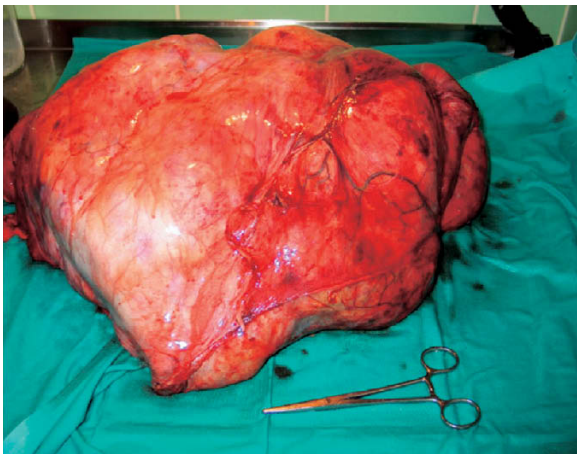

Figure1. Patient A.N. aged 25. Resected tumor. Size 35×35×25 cm and weight 14 kg.

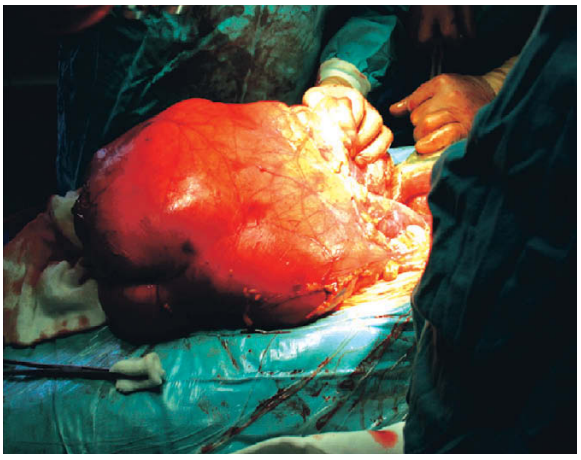

Figure 2. Patient S. K. aged 51 during resection. Size of the tumor 37×35×14 cm and weight 13 kg.

tion, a right inguinal hernia could be observed. In an interview it was revealed that the patient underwent removal of the uterus and adnexae in 2001, owing to the presence of a cyst on the left ovary. Ultrasound examination showed that the whole of the abdominal cavity was filled with a giant, solid tumour extending into the pelvis and covering the organs of the abdominal cavity. The liver had been translocated to a higher position, accessible for examination via the intercostal spaces, and was found to be homogeneous. The gall bladder was found to be normal and without stones. Neither the common bile duct nor the intrahepatic bile ducts were found to be widened. The pancreas was not accessible for examination. The kidneys were structurally normal and without stones. Urinary retention was observed in the joint systems of both kidneys and the proximal portions of both ureters. The spleen was not enlarged and was homogeneous. The bladder was symmetrical, with smooth walls.

Computed tomography of the abdominal cavity was commissioned. The investigation showed the presence of a massive lipoma-like change involving the intra-abdomen and hypogastrium and extending into the pelvis (*p. minor*). The liver, spleen and pancreas were found to be normal. Intestinal loops were moved to the right, to the paraspinal area. The left kidney was dislocated laterally on a lengthened pedicle. The bladder was displaced anteriorly. The patient qualified for exploratory laparotomy. The operation was performed on the 12th of February, 2004. A lobed, lipoma-like tumour, measuring 37×35×14 cm and weighing 13 kg, was excised along with the left kidney (11×5.5×4 cm) and adrenal gland (2.5×1.5×1 cm) which were embedded within it (Figure 2). The result of the histopathological investigation was: Lipoma-like sarcoma arising from the renal capsule. The postoperative course was without complication. The patient was discharged in good general health on the eleventh day after the operation. The most recent check-up on the 16th of August 2004, found the patient to be in good condition.

Discussion

With regard to the degree of a tumour's histological maturity, the WHO have established four histological types of liposarcomas, as follows:
1. Well differentiated liposarcoma;
2. Myxoid liposarcoma;
3. Pleomorphic liposarcoma;
4. Round cell/poorly differentiated liposarcoma.

Well differentiated liposarcoma without the traits of inflammation and sclerosis has been defined as lipoma-like liposarcoma [7].

The histological type, degree of advancement, size, and completeness of resection all influence the prognosis. Well differentiated and myxoid types are characterized by a low degree of malignancy though they have a tendency to recur locally (30%) [1]. Metastases are not observed if de-differentiation foci are not present. Liposarcomas with a high degree of malignancy are of the pleomorphic and poorly differentiated types. In these cases, the clinical course is much worse and metastases to the lungs are observed in 80–90% of cases. Thus, the survival rate for the pleomorphic type is 56%, 88% for the myxoid type and for the well differentiated type it is practically 100% [8,9]. However, a tu-

mour whose primary site is in the retroperitoneal space is associated with a worse prognosis (5 year survival rate averages 41%), chiefly owing to delayed diagnosis. The delay is frequently because of technically difficult resection and, in consequence, there is a tendency to recurrence and metastasis. Such tumours are the largest of all liposarcomas (average mass 7800 g, diameter 22 cm) and have a tendency to local expansion, causing symptoms induced by pressure on neighbouring organs [1]. Often they adhere to the renal capsule, though no invasion of the organ has been observed. The tumour mass often compresses the renal pelvis and ureter, leading to urine retention and hydronephrosis.

In the case of retroperitoneal tumours, enlargement of the abdominal circumference is often the first noticeable symptom. The abdomen may be found to be painful and tender upon examination, though the pain is rarely sharp (more frequently in the case of lipomas than liposarcomas). The enlarged abdomen is in contrast to more general emaciation; 25% of patients lose body mass, despite the increase in the circumference of the abdomen. Weakness and fatigue are frequently observed and 5% of patients complain of nausea, vomiting and constipation [10]. Kidney dysfunction may be observed in the majority of patients. Confounding diagnoses include lymphoma, neoplasms arising from the gametes and other soft tissue tumours (desmoid, malignant fibrous histiocytoma, leiomyosarcoma) [11]. Laboratory investigations may facilitate diagnosis: increased levels of alkaline phosphatase may point to lymphoma while increased levels of chorionic gonadotropin or alpha-fetoprotein accompany neoplasms of the germinal cells.

Radiograms are rarely diagnostic. Images may show unspecific masses of soft tissue. Usually, fatty tissue is not detected. Contrast examinations of the alimentary tract show the effects of pressure and displacement of the organs while ultrasound examination may confirm the presence of the tumour. Intravenous urography may reveal the displacement of the kidneys and ureters. Computed tomography allows for differentiation between well differentiated liposarcomas and other tumour types on the basis of higher fat content [12]. Using magnetic resonance the size of the tumour can be determined along with it's position, relative to the neighbouring organs. This technology may also be useful in the detection of recurrences and metastases. Based on findings from the literature, magnetic resonance should be considered to be the best method for the diagnosis of retroperitoneal tumours [13]. Fine needle biopsy allows for more accurate classification of the histological type of the tumour.

CONCLUSIONS

Surgery is a fundamental method in liposarcoma therapy. Adequate resection margins are necessary for recovery [14]. Complete resection is possible in 40–60% of cases of retroperitoneal tumours [5]. Recurrences are observed in 15–36% of cases and have been described even after periods of over 10 years and up to 30 years [15]. Liposarcomas are among the most radiosensitive of the sarcomas and radiotherapy (60–70 Gy) may be applied before, during or after surgery [4]. Large tumour masses will often move radiosensitive organs beyond the irradiated area. Preoperative radiotherapy has aroused the most enthusiasm as it allows for a reduction in tumour mass, facilitating resection [16–18] and, furthermore, lower radiation doses have proved to be most effective preoperatively. Some research centres have suggested the application of chemotherapy (doxorubicin, dacarbazine, cyclophosphamide, cisplatin) as an adjuvant treatment [4,19], though several multicentre research projects are studying survival rates after application of adjuvant chemotherapy.

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