LIPOSARCOMA OF THE RETROPERITONEAL SPACE – CASE REPORT

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Liposarcomas are rare neoplasms most often localized in the retroperitoneal space. Due to their localization, slow growth, and long latency period they often attain large sizes, infiltrating surrounding tissues and organs, before being diagnosed.

The study presented the case analysis, diagnostics, multistage surgical treatment and chemotherapy and recurrence rate, as a consequence of the primary tumors’ properties. Surgery remains the main method of treatment considering patients’ diagnosed with liposarcomas. The role of chemotherapy in the management of the above-mentioned remains an unsolved issue.

Key words: liposarcoma, retroperitoneal space soft tissue sarcoma

Liposarcoma for the first time was reported by Virchow in 1660’s (1, 2).

Liposarcomas are malignancies originating from mesenchymal tissues. Soft tissue sarcomas constitute only 2% of all malignancies. Approximately 10-20% of these tumors are located in the retroperitoneal space (3).

Liposarcoma is the most common sarcoma of the retroperitoneal space. According to various estimates, it accounts for 5-30% sarcomas and therefore is one of the most common soft tissue malignancies in adults (4, 5). Liposarcomas also occur in the mediastinum, lower limbs and abdominal cavity. Early diagnosis of this tumor is extremely difficult, in particular in obese subjects. Due to its location favoring growth, this tumor often achieves gigantic dimensions resulting in compressive symptoms before their diagnosis is made. Their size and location usually make radical surgical treatment impossible (3, 6). Relatively often these tumors infiltrate adjacent tissues and organs such as kidneys, large intestine, pancreas, duodenum and even large abdominal blood vessels. Often “en bloc” procedure is required, involving removal of infiltrated tissues and organs (3).

Sarcomas very rarely result in lymph node metastases (3, 7). Remote metastases (most commonly in lungs and liver) occur most commonly in later stages of development of highly malignant sarcomas.

CASE REPORT

A 51-year old patient, M.B., underwent multiple surgical procedures due to recurrent liposarcoma of the retroperitoneal space in 2nd Chair and Clinic of General and Oncological Surgery, Medical University in Wroclaw, between 2005-2009. Between 1989-2005 he had been diagnosed and treated outside the academic center.

In 1980’s the patient noted sporadic symptoms (reddening of his face, perspiration, hand tremor) suggesting arterial hypertension. When he bent, he experienced heatstroke, sudden increase of blood pressure to 200/160.

The patient was hospitalized many times (for the first time in 1989) in the department of internal medicine, cardiology and arterial hypertension of his regional hospital. Despite multiple tests, diagnostic procedures, treatment attempts, the patient has still been
complaining of unstable, uncontrolled arterial hypertension reaching as high as 250/170. Numerous imaging studies performed during this time (hospitalization every 2 years with complete panel of basic studies – chest X-ray, abdominal X-ray, US imaging of abdominal cavity, ECG, echocardiography, gastroscopy, colonoscopy, exercise tests, laboratory tests) did not reveal any abnormality.

During this time the patient’s well-being worsened dramatically. Despite following the recommended treatment, the patient complained of high blood pressure increases causing sudden, profuse sweating, face reddening, hand tremor, he experienced abdominal pain at the slightest bending. Face reddening is commonly regarded as a sign of alcohol abuse and the patient withdrew from social life for this reason. Sudden increases of blood pressure result in breakage of small blood vessels in the eyes, causing hemorrhages, conjunctivitis, conjunctival reddening.

US imaging performed in April 2005 demonstrated “…a lipid mass filling left half of his abdominal cavity from the diaphragm to the iliac fossa that crossed the midline. The mass was composed of three lobes: the main lesion had a shape of an egg (was palpable through the abdominal wall), in its lower pole, deeply reaching muscular layer, another part had dimensions of 6.5x7x3 cm and typical fluid spaces and calcifications (regressive changes) and from the medial side – the third, shaft-like part of the mass, reaching its right side”. CT imaging performed two weeks later demonstrated a lobular mass with dimensions of 14.4x17.2x14 cm in the abdominal cavity, in left umbilical and hypogastric region, reaching the pelvis minor.

For the first time the patient was admitted to 2nd Clinic of General and Oncological Surgery, Medical University in Wrocław, in 2005. Physical examination at admission demonstrated a quite hard, modestly painful mass, mobile upon respiration, with smooth surface, filling the left subcostal region, lateral abdomen and the lumbar region. Lower pole of the mass reached the iliac crest. After the patient preparation and performance of laboratory tests (that revealed no abnormality), the patient was operated for the first time. Three gigantic, capsulated, equipped with rich blood supply masses located in the abdominal cavity and retroperitoneal space on both sides of the descending and sigmoid colon, were removed. Two smaller masses, located medially, with approximately 10x10 cm in diameter each and a mass with 20x15 cm in diameter, reaching the spleen hilum and adhering to the left kidney, were resected completely. The surface of the mass section was homogeneous, fatty, grayish-yellow, without foci of necrosis or hemorrhages. Histopathological examination demonstrated liposarcoma lipoma like dedifferentiated type (M 8850/3). The postoperative period was uncomplicated. The patient left the hospital on day 11 after the surgical procedure. He was regularly followed-up in an outpatient setting and felt well.

Signs and symptoms of arterial hypertension in the patient resolved after the first surgical procedure. The patient’s well-being was getting better and better after the procedure, his eye hemorrhages were resolving as his face redness, excessive perspiration, sudden, accidental increased of his blood pressure.

A follow-up CT imaging performed in September 2005 demonstrated no malignant lesions. Four months later, in January 2006, US imaging demonstrated a local recurrence, only in the left iliac fossa, that was composed of three parts as previously. First of these three parts was located 2-3 fingers below the umbilicus, peripherally, closer to the iliac bone. This mass was located superficially and was palpable through the abdominal wall. Oval, whetstone-like, solid, capsulated mass with diameters of 10x9x2.5 cm. Moving deeper from this mass and in the direction of iliac blood vessels, another mass was found, which was whetstone-like too, with central calcifications, with 7x3x5 cm in diameter, without evidence of infiltration. A third lesion was directly adjacent to the previous lesions, located medially, was whetstone-like too, with a long tail reaching in the direction of blood vessels at the border between abdominal aorta – left iliac artery, with 6x3x8 cm in diameter, without any evidence of infiltration. Intraoperative examination performed two weeks later demonstrated numerous liposarcoma lesions, extending from the mesentery of the sigmoid colon laterally to the intestine, deeply to the retroperitoneal space along the aorta. The largest lesion, with dimensions of 10x10 cm, was located in the mesentery of the sigmoid colon while a smaller, hard lesion, with a diameter of 5-8 cm, was located immediately
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beneath the former lesion. After these lesions were removed, another lesions were visualized, located in packs along the aorta, each having approximately 6 cm in size, as well as several other smaller lesions. All lesions were resected completely and sent for histopathological analysis. Histopathological analysis demonstrated Liposarcoma lipoma like dedifferentiated type (M 8850/3).

In February 2006 the patient received his first cycle of chemotherapy (1st course AP3 (Doxo, DDP). US imaging performed in March 2006, 10 days after his second course of chemotherapy, demonstrated another recurrence in the same location. Typical focus of adipose tissue was located deeply retroperitoneally on the left side and had dimensions of 7x5.5x6 cm. Its center contained an amorphous fluid space. No other lesions were found.

Between April and June 2006 the patient underwent successive cycles of chemotherapy, a total of six. At the end of June 2006 the patient was admitted to department of internal medicine of his regional hospital due to marked pancytopenia, general weakness, severe bacterial infection with purulent lesions in his throat, to improve his blood cell counts and his general condition before another chemotherapy cycle. The patient was hospitalized for 6 days and was discharged home in good general condition.

A follow-up US imaging performed in July 2006 did not demonstrate any new liposarcoma lesions. The lesion found in US imaging in March 2006 did not change. Follow-up CT imagings of the abdominal cavity performed in September and December 2006 confirmed the result provided by US imaging of the abdominal cavity. Subsequent follow-up US imaging of the abdominal cavity from February 2007 did not show any lesion progression.

In August 2007 the patient was admitted to the hospital emergency department in Regional Hospital due to progressive jaundice, abdominal pain, abdominal pain and diarrhea. Due to progression of the malignant process the patient was transferred to Department of Internal Medicine, Academic Clinical Hospital in Wroclaw where acute pancreatitis (AP) with accompanying mechanical jaundice was found. After surgical consultation, the patient was qualified for ERCP and further medical treatment of AP. In September the patient was admitted to 2nd Chair and Clinic of General and Oncological Surgery, Medical University in Wroclaw, to undergo surgical treatment of cholecystolithiasis and appendix vermiformis. Histopathological examinations did not demonstrate any pathology.

Numerous follow-up US imagings of the abdominal cavity performed until January 2009 did not demonstrate any local recurrence. In January a retroperitoneal whetstone-like adipose structure was found between the aorta and ½ upper left kidney, 5.5x4.5x12 cm in size.

The patient was reoperated in March 2009. Intraoperatively two lesions were found (one on another), each 15x6x8 cm in size, infiltrating the descending colon and proximal part of the sigmoid colon. Furthermore, several nodules of various size were removed from the pelvis minor. Unfortunately, the procedure was not radical. The postoperative period was uncomplicated. After healing took place, the patient was discharged home and advised to continue his follow-up at the Clinic. Histopathological examination confirmed the previous diagnosis – liposarcoma lipoma like dedifferentiated type.

In September 2009 a follow-up US imaging of the abdominal cavity demonstrated a local recurrence. The imaging demonstrated a gigantic adipose mass extending from the costal arch on the left side to 3 fingers below the umbilicus. The lesion had approximately 35x18 cm in size. No other pathologies were found.

In October 2009 the patient underwent another operation and intraoperatively a nodular-adipose lesion was found, in a mesentry of the small intestine, in the region of splenic flexure and descending part of the large intestine. Its size was approximately of a handful. Macroscopic lesion was resected “en bloc”. A second similar lesion was found retroperitoneally, lying on and adherent to the lower pole of the left kidney. The lesion was resected along with a part of the lower pole of the left kidney. The pole of the left kidney was sutured with single sutures and a spongostan was put there. A follow-up US imaging demonstrated that size of the left kidney was normal and no urine retention was found in the renal collecting system. A hematoma, 5x7 cm in size, was found in the region of the lower pole of the kidney. After his wounds healed, the patient was discharged home. One month later the patient was admitted to hospital emergency department with signs and symptoms of pneu-
monia and fever 38.5-40°C. Later he was transferred to Department of Internal Medicine in Wrocław where medical treatment resulted in improvement of his condition and resolution of his complaints. CT imaging (December 2009) demonstrated “purulent lower pole of the kidney, 5 cm in size. Purulent reaction was also found in the left paraspinal canal. The patient was admitted to department of nephrology of Medical University where physical examination at admission demonstrated pain upon palpation in the projection of the left kidney and numerous postoperative scars but was otherwise unremarkable. Laboratory tests demonstrated elevated CRP levels and sterile pyuria. The patient was transferred to 2nd Chair and Clinic of General and Oncological Surgery, Medical University in Wrocław to undergo further surgical treatment. The patient underwent US-guided puncture of the paraspinal region and a bloody contents was obtained in the syringe, suggesting an “old” hematoma after the previous surgical procedure.

Histopathological examination (no. 311966) of the resected lesions demonstrated liposarcoma – liposarcoma bene dedifferentiatum.

DISCUSSION

It seems that in an era of development of imaging modalities, diagnosis of sarcoma of the retroperitoneal space should not pose problems. However long latency period, slow tumor growth and its location result often in sarcomas reaching gigantic sizes before such tests as US imaging of the abdominal cavity, computed tomography (CT) imaging or nuclear magnetic resonance (NMR) imaging are undertaken. Therefore patients for many years are unaware of their disease or for many years are searching for a cause of their complaints before correct diagnosis is made, which is supported by the reported case. This is a consequence of low incidence of sarcomas as compared to other abdominal tumors.

Surgical treatment is the mainstay therapy for sarcomas. The best results of surgical treatment are achieved when a malignant lesion is radically resected with a margin of healthy tissues. Most of the authors believe that resection of the tumor with a margin of healthy tissues is often impossible (8, 9). Therefore they recommend multiple resections of recurrent sarcomas believing that even non-radical procedures or partial tumor resection markedly improves patient survival (3, 8, 10). Clinical observations indicate that local recurrence rate closely depends on the method of tumor resection and degree of radical operation (3, 8, 9, 11). Basing on available literature, Storm et al. (3, 11) showed that despite generally poor prognosis, long-term survival can be achieved in more than half of patients undergoing radical operation. He estimated that local recurrence after resection of a sarcoma of the retroperitoneal space (SRS) occurs in 40% of patients within 2 years of surgical treatment, in 72% of patients within 5 years and in 91% of patients within 10 years of the tumor resection. Most authors estimate that recurrence rate after SRS resection ranges from 33% to 86% (12, 13). For liposarcoma, 5-year survival ranges from 85% (liposarcoma lipoma like dedifferentiated), 10-year survival is by 10-15% lower (14, 15). According to the literature, liposarcoma of the retroperitoneal spaces carries particularly poor prognosis (due to impossible radical resection) – 5 year survival is approximately 4%.

The above presented case seems typical – a relatively common lesion, giving local recurrences. Before the correct diagnosis was made, the patient was treated for many years. No metastatic lesions were observed despite five operations. Unfortunately, despite efforts of the surgical team, radical procedure could not be performed which meant that probable reoperation would be required in the future.

Results of chemotherapy in our patients do not recommend its wider application. In 1980’s several authors published their experiences with chemotherapy before (11) and after (2) surgical treatment. No clear beneficial of such management on patient survival was found. Its efficacy is not clear and feasibility of such treatment is debatable.

CONCLUSIONS

1. Surgical treatment is the mainstay therapy for sarcoma.
2. Local recurrence after the treatment of the primary tumor increases the risk of another recurrence and reduces the chance of complete cure.
3. Role of chemotherapy in the treatment of liposarcoma is unclear.
REFERENCES


Received: 23.06.2010 r.
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