CASE REPORTS

INFERIOR VENA CAVA LEIOMYOSARCOMA IN THE RETROPERITONEAL SPACE – DIAGNOSTIC AND THERAPEUTIC PROBLEM

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Inferior vena cava leiomyosarcoma is a malignant neoplasm rarely diagnosed. The study presented two cases of inferior vena cava leiomyosarcomas subject to treatment at the Department of Oncology, Center of Oncology in Warsaw. The first patient underwent surgery after a two-year thorough gastroenterological diagnostic process, being suspected of pancreatic cancer. Proper diagnosis was established intraoperatorically, the tumor was completely excised. After ten months the patient was subject to chemotherapy, due to the presence of distant metastatic lesions. Chemotherapy was stopped after 24 months, due to disease progression. The patient died six months, thereafter. The second patient underwent surgery after a short diagnostic process, being suspected of an adrenal gland tumor. Proper diagnosis was established intraoperatorically, and the tumor was completely excised. The patient remained under close oncological control. Three years after the procedure disease recurrence was not observed. Retroperitoneal space tumors pose a significant clinical problem. Rapid diagnosis and proper therapeutic decisions often determine the future of such patients. The five-year survival rate of patients subject to radical surgical treatment, due to inferior vena cava leiomyosarcoma ranges between 33 and 55%.

Key words: inferior vena cava, leiomyosarcoma, retroperitoneal space tumor, surgical treatment, diagnostics

Soft tissue sarcomas are rarely diagnosed and constitute 1% of all diagnosed malignant neoplasms (1). In spite of the dynamic development of non-invasive imaging diagnostic methods and constant progress in oncological therapy, tumors localized in the above-mentioned site pose a significant problem for the surgeon.

The study presented two patients treated at the Center of Oncology, due to retroperitoneal space tumors. Proper clinical diagnosis was established intraoperatorically. Inferior vena cava leiomyosarcoma diagnosis was established postoperatorically on the basis of the histopathological examination.

CASE REPORTS

1. A 53-year old female patient was admitted to the Department of Surgery, due to suspicion of pancreatic head tumor, 9x7 cm in size. Diagnostics of the ambiguous lesions observed during abdominal ultrasonography was performed in other medical centers, and began two years ago. During the prolonged period of diagnostics numerous examinations were performed, such as endoscopic ultrasonography and fine-needle biopsy, computer tomography, somatostatic receptor scintigraphy, and blood sampling (neoplastic markers). The patient was qualified for surgical treatment. Intraoperatoratively, proper diagnosis was established-pancreatic head tumor infiltrating the intra-abdominal segment of the inferior vena cava. Due to the macroscopic character of a sarcoma the tumor was completely excised, together with part of the inferior vena cava. The vena cava was reconstructed by means of a vascular prosthesis. The histopathological examination revealed leiomyosarcoma.
result was as follows: leiomyosacoma partim pleomorphicum venae cavae G3, the tumor 7 cm in diameter was completely excised.

Ten months after the operation control CT and MRI examinations showed the presence of isolated metastatic lesions localized in the liver, without features of local recurrence. The patient was qualified to palliative chemotherapy, based on doxorubicin, dacarbasin, and ifosphamid schedules. During chemotherapy one observed the stabilization of lesions in the imaging examinations followed by progression and deterioration of the patients’ general condition. Chemotherapy was stopped after 24 months. Six months later the patient died, due to cachexia. Since the beginning of diagnostics to patient death, 5 years and 5 months had elapsed, and since surgery to death – 3 years and 5 months.

2. A 20-year old female patient was admitted to the Department of Surgery with diagnosis of a right adrenal gland tumor (10 cm in diameter) infiltrating the inferior vena cava, established during ultrasound and CT examinations. The tumor was hormonally inactive. The patient was qualified for surgical treatment. Intraoperatively, the diagnosis was established: a 10 cm tumor in contact with the right adrenal gland and kidney, without visible infiltration. The tumor infiltrated the intra-abdominal segment of the inferior vena cava, being a sarcoma. The tumor was completely excised with part of the inferior vena cava. The vena cava was reconstructed by means of a vascular prosthesis (fig. 1). The postoperative course proved uneventful. The histopathological examination result was as follows: leiomyosarcoma G2, partim G3, the tumor 9 cm in diameter was completely excised.

The patient remained under close observation without adjuvant therapy. Nineteen months after the surgical intervention, the control computer tomography examination revealed the presence of a possible metastatic lesion, at the site of the previously removed tumor. The patient was once again qualified for surgery. The intraoperative examination showed no local recurrence, nor abdominal cavity metastatic lesions. At the moment (32 months after the operation) the patient is in good general condition. Computer tomography and chest X-ray examinations are free of recurrence.

DISCUSSION

Retroperitoneal space neoplastic lesions constitute a heterogenous group of disorders. In most cases these tumors are derived from the pancreas, kidney, and adrenal glands, as well as enlarged lymph nodes during the course of a lymphoma, metastatic lesions (testicle), and inflammatory processes. Fibromatosis, lipomas, and Schwannomas might also imitate retroperitoneal space tumors. Considering primary malignant mesenchymal tumors developing in the retroperitoneal space the following sarcomas dominate: liposarcomas, malignant fibrohistiocytomas, and leiomyosarcomas (2). In spite of the diversity of neoplastic diseases capable of developing in the retroperitoneal space one must not forget that the diagnosed tumor is in most cases malignant (65%-84%) (3, 4).

Inferior vena cava leiomyosarcoma is a rare disease entity. Only 300 such cases have been described, worldwide. The largest patient group can be found in the Memorial Sloan - Kettering Cancer Center (MSKCC) in New York. During a period of 20 years (1982 -2002) 25 patients with the above-mentioned diagnosis were subject to treatment (5). Based on our study and meta-analysis of other publications, one can come to the conclusion that inferior vena cava leiomyosarcomas mostly concern middle-aged (median 54 years) women (2/3 of patients). Disease progression is slow, and distant metastases appear late. First subjective symptoms, such as abdominal pain, palpable tumor through the abdominal
integuments, and lower leg edema, due to venous thrombosis usually appear relatively late. Imaging examinations revealed the size of the tumor, which attained an average of 10 cm (between 3 and 32 cm, according to the Memorial Sloan-Kettering Cancer Center). Imaging examinations show a large, diffuse tumor mass without a visible starting point. Fine-needle and thick-needle biopsy or sample collection (laparoscopy or laparotomy) have not found their place as standard diagnostic procedures. In case of atypical neoplasms fine-needle biopsy has little diagnostic value, while thick-needle or open biopsies are burdened with the risk of significant complications (the above-mentioned is not considered as optimal treatment in case of oncological asepsis).

The only effective method of treatment considering inferior vena cava leiomyosarcomas is the excision of the tumor with healthy tissue margins. Thus, the decision concerning surgery should be immediately undertaken after non-invasive diagnostics. In case of technical possibilities in the complete removal of the tumor, and lack of generalized disease symptoms, radical surgery should be undertaken.

During the past years radiotherapy has become a standard adjuvant therapeutic method following leiomyosarcoma surgery. In case of retroperitoneal tumor localization (vicinity of „critical” organs) intraoperative radiotherapy is initiated. The above-mentioned method reduces the number of local recurrences. However, in case of leiomyosarcomas there is no evidence, whether such treatment has any influence on survival.

There is no literature data demonstrating the efficacy of chemotherapy after surgery in case of patients diagnosed with leiomyosarcomas. Chemotherapy is used during clinical trials as palliative treatment in case of inoperable lesions or when distant metastatic lesions are observed.

The only oncological therapeutetic method after radical inferior vena cava leiomyosarcoma surgery consists in close observation combined with imaging examinations. Postoperative lesions at the site of the removed tumor are difficult to interpret. Thus, when suspecting tumor recurrence, patients should be subject too explorative laparotomy, which enables recovery. The estimated five-year survival rate after radical surgery ranges between 33 and 55% (6, 7).

REFERENCES


Received: 27.10.2010 r.
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