CLINICAL CASE

Adv Clin Exp Med 2004, **13**, 6, 1055–1058
ISSN 1230-025X

WOJCIECH WITKIEWICZ, JAN GNUS, WILLY HAUZER, PIOTR PŁAWSKI

Diaphragm Leiomyosarcoma – Diagnostic Difficulties

Mięśniakomięsak gładkokomórkowy przepony – problemy diagnostyczne

General and Vascular Surgery Department, Regional Specialistic Hospital of Wroclaw, Poland

Abstract

The study presents a case of a 61-year-old woman with unspecific intra-abdominal pain. Based on ultrasonography and computer tomography she was diagnosed with gastric tumour and qualified for surgery. The histopathological examination demonstrated a leiomyosarcoma originating from the right side of the diaphragm. Three years following surgery abdominal USG and CT demonstrated tumour recurrence. The patient was reoperated and diagnosed with a hepatogastric ligament tumour, which required excision. Five years since the first surgery the patient was diagnosed with tumour of the hilum, requiring reoperation. Histopathology revealed the presence of leiomyosarcoma. The patient has been free of symptoms ever since. Surgery is the treatment of choice in case of such tumours, being the only chance of survival (**Adv Clin Exp Med 2004, 13, 6, 1055–1058**).

Key words: leiomyosarcoma, diaphragm, treatment, surgery.

Streszczenie

W pracy przedstawiono przypadek 61-letniej kobiety z nieswoistymi dolegliwościami bólowymi jamy brzusznej. Na podstawie badania ultrasonograficznego i tomografii komputerowej rozpoznano guz żołądka, a chorą zakwalifikowano do leczenia operacyjnego. Śródoperacyjnie stwierdzono guz prawej odnogi przepony, a badaniem histopatologicznym rozpoznano mięśniakomięsak gładkokomórkowy. Po trzech latach od operacji na podstawie USG jamy brzusznej i tomografii komputerowej stwierdzono wznowę guza. Pacjentkę ponownie operowano i stwierdzono guz więzadła wątrobowo-żołądkowego, który wycięto. Po pięciu latach od pierwszej operacji u chorej rozpoznano guz umiejscowiony we wnęce wątroby – chorą reoperowano. W badaniu histopatologicznym rozpoznano mięśniakomięsaka gładkokomórkowego. W przypadkach guza tego rodzaju leczenie operacyjne jest leczeniem z wyboru (**Adv Clin Exp Med 2004, 13, 6, 1055–1058**).

Słowa kluczowe: mięśniakomięsak gładkokomórkowy, przepona, leczenie chirurgiczne.

The first case of leiomyosarcoma of the diaphragm was described by Kirschbaum in 1935. Only few such cases have been reported from then on [1, 3, 5, 6]. Unsymptomatic and poor histologically-differentiated leiomyosarcomas that are situated in the thorax or abdomen as the pedicle tumour of the right part of the diaphragm are found in the adult only occasionally. Soft tissue sarcomas are considerably rare and stand for only approximately 0.7% of the malignancies in the entire population [9]. The most prevalent histological types of sarcoma are liposarcoma and leiomyosarcoma. Leiomyosarcomas can be situated in different sites yet above half of cases occur in the organs of the abdomen and retroperitoneal space. They metastasise to the liver and lungs via blood or less common the lymphatic vessels. The treatment of choice in the early stages of sarcoma is surgical operation [4, 7, 8].

Case Report

61-year-old woman – non-smoker, white-collar worker – once suffering from rheumatic disease was examined because of non-specific gastric complaints. Laboratory results and clinical examination were normal. The ultrasound and contrast X-ray of abdomen revealed the tumour originating from the gastric wall at the site of the lesser curvature.

Despite the smooth borders, the specific type of vascularisation described during the Doppler-USG examination as well as the above-mentioned results created considerable fear regarding the histological type. Endoscopy showed protrusion of the cardiac part of the stomach and the lesser curvature without macroscopic mucosal changes. An X-ray of the chest was within normal limits. CT scans detected 5-cm tumour located under the diaphragm next to the lesser gastric curvature.

Figures 1 and 2 both without contrast visualise subdiaphragmatic space with the tumour at the lesser curvature of the stomach wall.

Figures 3 and 4 show the same tumour after contrast enhancement.

The patient was operated on in December 1996. The abdominal cavity was open by median laparotomy and the 5-cm wide, pedicle tumour originating from the diaphragm without any infiltration of surrounding tissues but merely compressing the lesser curvature was found next to the esophageal hiatus. There were neither any enlarged lymphatic nodes nor macroscopic pathological changes of abdominal organs present. The tumour was excised with a 1-cm margin of healthy surrounding tissues. The void in the diaphragm was fixed with a continuous, absorbable suture Dexon 3/0 and the obtained material was sent to undergo histopathological examination.

On histological examination the tumour was found to be consisted of skeletal muscle fibres with addition of numerous, wide-lumen blood vessels and adhering and infiltrating leiomyosarcoma cells.

The postoperative period went uncomplicated and the wound healed by prime intention. During the third month after the operation USG and CT scans as well as thoracic cavity X-rays did not show any metastases or recurrence of the disease.

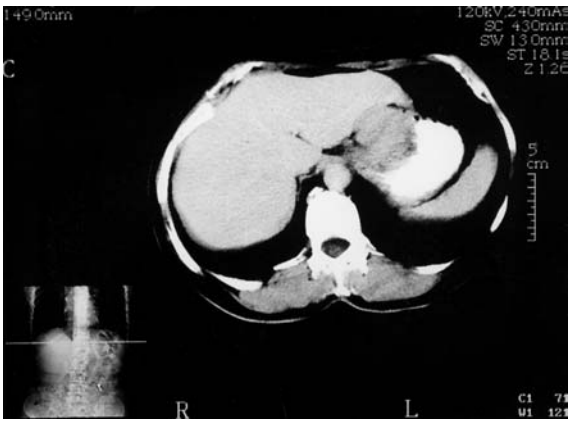

Fig. 1. Subdiaphragmatic space with the tumour at the lesser curvature of the stomach wall before application of contrast

Ryc. 1. Przestrzeń podprzeponowa z guzem na krzywiźnie mniejszej ściany żołądka przed podaniem kontrastu

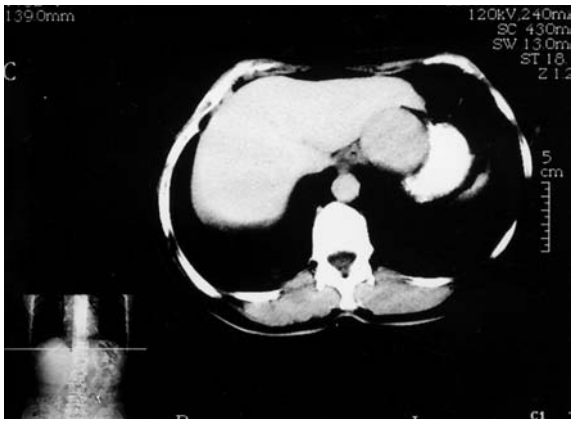

Fig. 3. Subdiaphragmatic space with the tumour at the lesser curvature of the stomach wall after contrast enhancement

Ryc. 3. Przestrzeń podprzeponowa z guzem na krzywiźnie mniejszej ściany żołądka po podaniu kontrastu

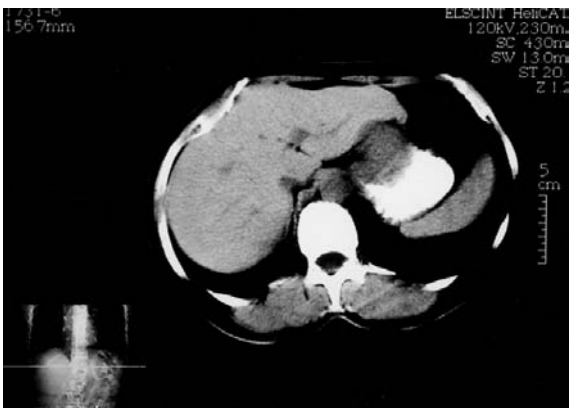

Fig. 2. Subdiaphragmatic space with the tumour at the lesser curvature of the stomach wall before application of contrast

Ryc. 2. Przestrzeń podprzeponowa z guzem na krzywiźnie mniejszej ściany żołądka przed podaniem kontrastu

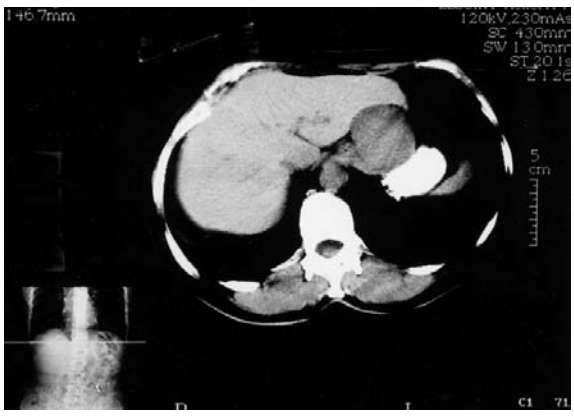

Fig. 4. Subdiaphragmatic space with the tumour at the lesser curvature of the stomach wall after contrast enhancement

Ryc. 4. Przestrzeń podprzeponowa z guzem na krzywiźnie mniejszej ściany żołądka po podaniu kontrastu

On oncologic consultation the patient was not qualified to specialised therapy. In December, 1999, USG and then CT scans of the abdomen showed 3 × 4 cm hypodense focus in the vicinity of the hepatogastric ligament. The change protruded into the liver and was not sharply separated from it. The findings suggested lymph nodes or liver metastases. The rest of abdominal organs were normal, no enlarged retroperitoneal lymph nodes were reported. The patient was qualified to be re-operated. The abdominal cavity was opened by the median laparotomy and the 3 × 4 cm tumour localised in the hepatogastric ligament that adhered to the left hepatic lobe sac was noted. The tumour was radically excised, control of abdominal organs did not show any pathology. The postoperative period was without complication, the wound healed by the prime intention.

On the ninth postoperative day the patient in a good physical condition was released from the hospital. The histological examination showed that the taken material consisted of: 1) nodule of hepatogastric ligament: *leiomyosarcoma*, 2) pedicle: *Infiltratio leiomyosarcomatis. Lymphonodulitis reactiva sine neoplasmate*.

Immunohistochemical studies revealed presence of muscle vimentin, desmin and actin in the neoplastic cells and morphology suggested leiomyosarcoma G2 stage. The studies were made in the Pathologic Anatomy Department of the Wrocław Medical University, by Prof. J. Rabczyński and Dr. M. Jeleń. No pathological changes were found in control examination during 6th and 9th month of convalescence. USG, CT and chest X-ray were normal.

The control USG of abdomen in March, 2001, revealed the liver hilum tumour – 20 × 18 × 12 mm – with a small fluid cavity suggesting a cyst – 15 × 10 mm – above. The retroperitoneal lymph nodes were not enlarged and the rest of abdominal organs – normal. Control CT of abdomen showed correct stromal organs, no enlarged lymph nodes were visualised. The patient was qualified to be re-operated again. The abdominal cavity was opened by median laparotomy and 20 × 20 × 15 mm tumour of the liver hilum was detected as well as the cyst of 20 × 10 mm adjacent to the tumour. The rest of abdominal organs were not macroscopically changed. The postoperative period went without complications, the wound healed by the prime intention, the patient in good physical state was dispatched home. Histological studies made in the Patomorphology Department of Regional Specialistic Hospital in Wrocław and in the Pathologic Anatomy Department of the Wroclaw Medical University detected leiomyosarcoma in the tumour but the tissues of the cyst were not neoplastically changed. Control USG of abdominal cavity made in December 2002 showed fluid cyst reaching 15 × 10 cm. The cyst was localized in the hepatogastric ligament located on the portal vein. The patient was qualified to be operated on.

Discussion

Sarcomas of the abdomen cause no complaints for a long time or produce only mere symptoms. They are diagnosed in advanced stage, when surrounding tissues or organs have already been infiltrated. Considering clinical evidence it was decided to perform CT scans and ultrasound in this particular case. Primarily based on the results a cyst was suspected. Laboratory findings and an X-ray did not reveal anything specific. In described case of leiomyosarcoma was found as rare pedicular tumor coming from diaphragm. When tumor enlarged to considerable extent, the patient started to experience some unspecified gastric complaints. Modern diagnostic methods – endoscopy, ultrasound, CT scans of the abdomen X-ray and laboratory findings – did not result in accurate diagnosis. Only laparotomy enabled ultimate diagnosis. As agreed with other authors only surgical procedure may cure or at least prolong life and improve its quality [8]. Radical resection in which the entire anatomic compartment involved by the sarcoma should be provided. In this case such extensive procedure was impossible. Local lymphatic nodes were unchanged and no macroscopic infiltration of surrounding tissues was justifiable reasons of the range of procedure. The results of histopathological examination and over two-year-survival without local metastases and five-year-survival at present confirm rightness of the therapeutic scheme. There is no proof that chemio-, hormono- or radiotherapy have positive influence in such sarcomas. Mainly they are used as palliative treatment and may improve the condition of the patient for the next several weeks [1, 7].

The patient is under strict, regular control in authors' surgical clinic.

Malignant tumours originating from smooth muscle cells are very rare. There have been only few cases of primary leiomyosarcoma of the diaphragm described so far. The latest diagnostic methods including endoscopy, ultrasound and CT scans have important and key role in detecting leiomyosarcoma because the complaints of the patient were highly non-specific. The authors think that USG and CT of abdomen are very important and help detect the tumours of the diaphragm. Especially, USG of abdominal cavity is useful in monitoring during the follow-up period. Only surgical and radical operation enables the patient to survive.

References

[1] Blondeel PhN, Christiaens MR, Thomas J: Primary leiomyosarcoma of diaphragm. Eur J Surg Oncol 1995, 21, 429–431.
[2] De Vita: Cancer: Principles and Practice of Oncology. Lippincott-Raven Publishers 1997, 5th ed.
[3] Dionne GP, Beland JE, Wang NS: Primary leiomyosarcoma of the diaphragm of an asbestos worker. Arch Pat Lab Med 1976, 100(7), 398.
[4] Lang H, Nusbaum KT, Kaudel P: Hepatic metastases from leiomyosarcoma. Ann Surg 2000, 231(4).
[5] Kodriavstev VA, Ponomarieva OA: Malignant tumor of the diaphragm in a 2-month-old child. Vestin Khir 1978, 12, 85–86.
[6] McCoy WC: Leiomyosarcoma of the diaphragm. South Med J 1963, 642–647.
[7] Oakley R, Carty H, Cudmore RE: Multiple mesenchymoma of the chest wall. Pediatr Radiol 1985, 15, 58–60.
[8] Oparker MC: Leiomyosarcoma of the diaphragm – a case report. Eur J Surg Oncol 1985, 11(2), 171–175.
[9] Strauch JT, Aleksic I, Schorn B: Leiomyosarcoma of the diaphragm. Ann Thorac Surg 1999, 67, 1154–1155.

Address for correspondence:

Wojciech Witkiewicz
General and Vascular Surgery Department
Regional Specialistic Hospital
Kamieńskiego 73a
51-124 Wrocław
Poland

Received: 26.05.2004
Revised: 16.06.2004
Accepted: 16.06.2004

Praca wpłynęła do Redakcji: 26.05.2004 r.
Po recenzji: 16.06.2004 r.
Zaakceptowano do druku: 16.06.2004 r.