CASE REPORT

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Giant oesophageal leiomyoma as a diagnostic and therapeutic problem – a case report

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Abstract

Oesophageal leiomyoma is a rare benign tumour of the oesophagus, which does not cause clinical symptoms in more than half of cases. Below we present the case of a symptomless oesophageal tumour. Due to the unequivocal result of imaging examinations and no histopathological diagnosis the patient was qualified for thoracotomy. During the operation the oesophageal tumour was suspected of passing through the hiatus into the abdominal cavity. The specimen revealed a neoplasm of mesenchymal origin without distinct traits of malignancy. The patient was admitted to the Surgical Department, where she was qualified for surgery, which was carried out by a team of surgeons and thoracic surgeons. The encapsulated tumour and oesophagus were resected. The histopathological and immunohistochemical examinations corres ponded to leiomyoma oesophagi. The lesion was radically resected. The size of the lesion was 22 × 14 × 13 cm. Three months after the surgery no traits of relapse were found.

Key words: oesophageal, leiomyoma, surgery.

Streszczenie

Mięśniak gładkokomórkowy przełyku jest rzadko występującym nowotworem łagodnym przełyku, który w ponad połowie przypadków nie powoduje objawów klinicznych. W pracy zaprezentowano przypadek bezobjawowego guza przełyku wykrytego u pacjentki podczas badań okresowych. Zmiana obejmowała cały piersiowy odcinek przełyku. Pacjentka została przyjęta na oddział torakochirurgii, gdzie wykonano biopsję zmiany. Z uwagi na niejednoznaczny wynik badań obrazowych oraz brak rozpoznania histopatologicznego chorą zakwalifikowano do zwiadowczej torakotomii, podczas której stwierdzono guz obejmujący cały odcinek przełyku z przejściem przez rozwór do jamy brzusznej. Rozpoznano nowotwór pochodzenia mezenchymalnego, bez wyraźnych cech złośliwości. Na dalszym etapie, po uzupełnieniu diagnostyki, chora została zakwalifikowana do zabiegu chirurgicznego, wykonanego przez zespół chirurgiczno-torakochirurgiczny, podczas którego usunięto otorebkowany guz wraz z przełykiem. Obraz histopatologiczny uzyskany na podstawie wyników badań immunohistochemicznych odpowiadał *leiomyoma oesophagi*. Wymiary zmiany wynosiły 22 × 14 × 13 cm. Przebieg pooperacyjny był niepowikłany. Obecnie pacjentka pozostaje pod kontrolą Poradni Chirurgii Onkologicznej, w wykonanych badaniach obrazowych po 3 miesiącach od zabiegu bez cech wznowy.

Słowa kluczowe: przełyk, mięśniak gładkokomórkowy, zabieg chirurgiczny.

Introduction

Leiomyoma is a benign neoplasm of mesenchymal origin. The most common location in the alimentary tract is the oesophagus, where the tumour is responsible for most diagnoses of benign tumors of the organ [1]. The frequency of occurrence ranges between 0.005% and 7.9% of the population [2-4]. It is chiefly diagnosed between the ages of 30 and 59. The tumour grows slowly. It is often symptomless and is accidentally diagnosed in tests. If symptoms occur, usually it is dysphagia and pain in the epigastrium and chest [5, 6]. The basic method of treatment of oesophageal leiomyoma is its radical resection. Depending on the size and location of the lesion patients are qualified for a particular type of surgery. At present a wide range of operations are available, from minimally invasive submucosal resections carried out during endoscopy, through surgery involving opening of the thorax, to subtotal oesophageal resection with the transhiatal method or thoracotomy.

Although oesophageal leiomyoma is a benign neoplasm, its diagnostics and treatment are often a serious challenge

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for the whole team of physicians. Below we present a case study of a patient whose considerable oesophageal leiomyoma was both a diagnostic and therapeutic problem.

Case study

A patient, aged 54, was admitted to the Department of General Surgery and Surgical Oncology, Wielkopolska Cancer Centre, where she was to be treated for a posterior mediastinal tumour, which filled the left half of her chest. The lesion was diagnosed in routine periodic examinations at her place of work in March that year (chest radiograph). The patient had not reported any ailments which may have been related to the presence of the lesion. The patient had no chronic diseases diagnosed; her oncological history was insignificant. The patient was referred to the Thoracic Surgery Outpatient Clinic and then admitted to the Thoracic Surgery Department, Wielkopolska Center of Lung Disease and Thoracosurgery in June that year to undergo diagnostics and treatment. The patient underwent computed tomography of her thorax, where a pressed left lung lower lobe modelled on an enormous mass was described. The tumour came out of the stomach and caused concentric thickening of its wall, mainly around the body. The lesion was adjacent to the pericardium, modelling the left lower pulmonary vein, lifting the left main bronchus and pressing the basal segmental bronchi of the left lung lower lobe. The maximum size of the infiltration was 12 × 13 × 18 cm (Fig. 1). Apart from that, there were no significant deviations from the description. In the conclusions from the examination the radiologist suggested that the diagnosis should take a lymphoma or myosarcoma into consideration, GIST being less likely. In order to obtain a histopathological diagnosis a fine-needle aspiration biopsy was carried out. As a result, no neoplastic cells were found; the smears had an extremely low cell count. Due to the results a transthoracic core-needle biopsy of the lesion was carried out, which brought no diagnosis, either. Its result only corresponded to relatively numerous very fine fimbriae of low cell connective tissue with individual fine vessels. Due to the lack of histopathological diagnosis and unequivocal image of CT examination the patient was qualified for exploratory thoracotomy with biopsy of the lesion. During the surgery the lesion was found to be more advanced than the CT examination indicated, because in practice, the tumour occupied the entire thoracic segment of the oesophagus and the gastric cardia, which was moved to the thorax. Part of the tumour seemed to enter the abdominal cavity through the oesophageal hiatus. A specimen collected in the thoracotomy produced the diagnosis of mesenchymal spindle cell carcinoma without distinct traits of malignancy. Later, in August 2012, the patient was admitted to the Surgical Department, Greates Poland Cancer Centre, Poznań, Poland. The patient had diagnostics supplemented with computed tomography of the abdominal cavity, which confirmed the presence of an oesophageal tumour filling the left pleural cavity. Apart from that, the examination did not reveal pathology within the organs and structures beneath the diaphragm. The patient underwent gastroscopy, which did not reveal any lesions in the oesophageal or gastric mucosa. The patient was qualified for surgery carried out by a team of surgeons and thoracic surgeons. First the patient underwent laparotomy and the region of the oesophageal hiatus was assessed. The tumour was not found to enter the peritoneal cavity. Next, the thorax was opened on the left, in the scar after the previous thoracic surgery. The encapsulated tumour was resected by cutting off the oesophagus proximally in the upper thoracic segment and distally within the region of the gastric cardia relocated to the thorax. As was described in the CT examination of the thorax, the tumour was adjacent to the pericardium and the lower lung lobe base, which required careful dissection of the lesion. The resected thoracic segment of the oesophagus passed right through the middle of the tumour.

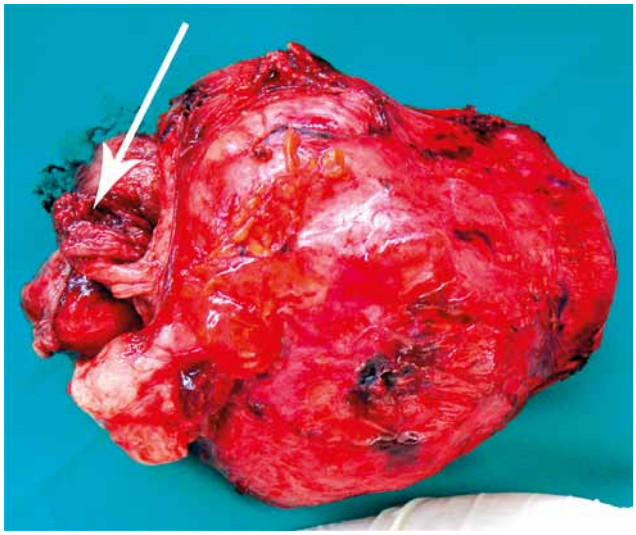

Fig. 1. Postoperative photograph of the tumour

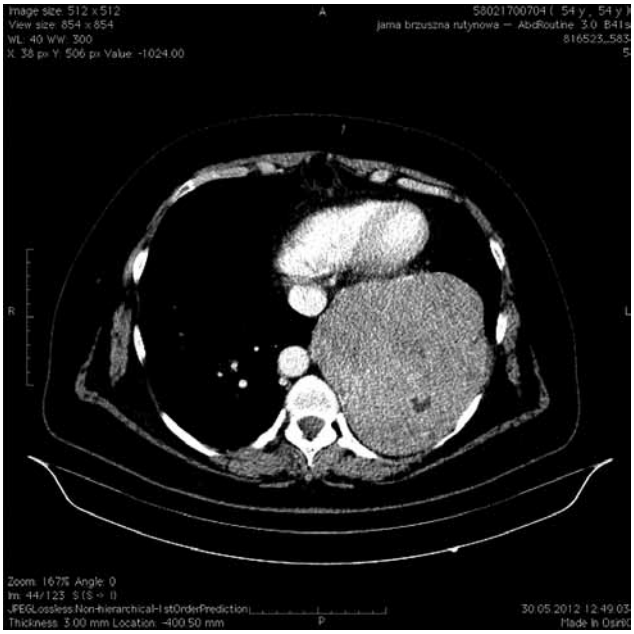

Fig. 2. CT scan with tumour

The alimentary tract was reconstructed by means of a gastric prosthesis, anastomosing it on the left side of the neck to the stump of the cervical segment of the oesophagus. The postoperative course was free from complications. On the 11th day after the surgery the patient was discharged from hospital with a recommendation of a follow-up examination at the Surgical Oncology Outpatient Clinic. The histopathological image based on the results of immunohistochemical examinations (SMA, Ki67, Calp, CD 117, S100, CD 34, DOG 1) corresponded to oesophageal leiomyoma. The lesion was radically resected. The largest size of the lesion was 22 × 14 × 13 cm. The patient remains under control of the Surgical Oncology Outpatient Clinic, where she has check-ups. Three months after the surgery no pathological lesions were found in computed tomography of the thorax.

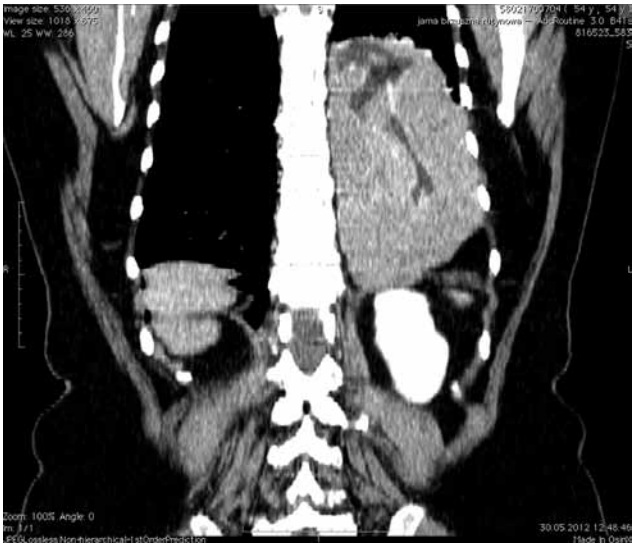

Fig. 3. CT scan with tumour

Discussion

Oesophageal leiomyoma usually occurs in the central and distal part of the organ. It is very rarely identified in the upper segment. The analysis of a group of 63 patients with the tumour, who were treated in one centre, revealed that the location in the central part of the oesophagus concerned 47.6% of the patients and the distal location 37.1% of the patients [7]. Similarly, in another study the most frequent location in the distal segment of the oesophagus concerned 46% of the patients. In the case under study the main part of the lesion was also located in the distal segment of the oesophagus. Due to the slow growth of these tumours the vast majority of them (59%) cause no ailments to patients. Some authors report a correlation between the size of the lesion and the occurrence of clinical symptoms. The average size of the tumour in patients with symptoms was 5.2 cm (0.8-14 cm), whereas in patients without symptoms the average size of the lesion was much smaller – 0.4 cm (0.1-1.0 cm) [8]. The literature describes different diameters of resected tumours, ranging from 0.1 cm up to the maximum size of 29 cm [1, 9, 10]. In the case under study, in the final histopathological examination the lesion was 22 cm. In spite of this size the tumour did not cause any clinical ailments and was diagnosed during routine periodic examinations at work, where the patient had her chest X-rayed.

The diagnosis of leiomyoma is based on imaging examinations and biopsy in order to obtain a histopathological diagnosis. By choice endoscopy and endoscopic ultrasound are the examinations where the lesions can be seen as submucosal movable masses with untouched mucosa, which enables exclusion of the malignant process. In most cases the tumours did not cause lesions in the oesophageal mucosa, which also did not occur in the case under discussion [8]. Unfortunately, the centre treating the patient does not have a possibility to carry out endoscopic ultrasound (EUS). Therefore, the diagnosis was based on other imaging examinations. Apart from the EUS, computed tomography and magnetic resonance are the examinations that enable diagnosis. The computed tomography image shows a mass bound to the oesophagus, without traits of lymphadenopathy, which enables the diagnosis of a benign lesion. In the case under discussion there was a relatively large discrepancy between the computed tomography description and the actual image observed in exploratory thoracotomy. The collection of specimens for histopathological diagnosis is a subject of discussion [6, 11]. Some authors stress that multiple biopsies may cause adhesions in the thorax, which later make minimally invasive resection of the lesion much more difficult and which cause weakening of the mucosa, which is of key importance in an attempt to enucleate the tumour without resection of the oesophagus [7, 12]. Besides, the collected material is usually insufficient to make a final diagnosis, especially if it is only cytological smear. At present histopathological diagnostics also includes the immunohistochemical examination, but it is necessary to have an appropriate amount of material to make the examination. In the case under study neither the attempt of a fine-needle aspiration biopsy nor core-needle biopsy resulted in diagnosis. Even when a specimen was collected during exploratory thoracotomy, only an initial diagnosis of benign mesenchymal tumour was possible. The final histopathological diagnosis was made when the entire lesion was resected and a panel of immunohistochemical examinations was carried out.

The basic method of treatment of oesophageal leiomyoma is surgical resection of the lesion. The choice of surgical technique depends on the size and location of the lesion and on the occurrence of clinical symptoms. The smallest lesions that are described are smaller than 0.1 cm in diameter. Usually these lesions are accidentally diagnosed and they do not cause any clinical symptoms. Some authors do not recommend resection of the lesions. They only suggest an EUS check-up every one or two years [6]. There are no clear guidelines concerning the procedure of handling small lesions. Larger lesions, over 5 cm in size, which cause clinical symptoms, have a visible growing tendency, cause ulceration and their final histopathological diagnosis is uncertain, need to be radically resected [5, 13].

So far four cases of malignant transformation of leiomyoma have been described [14]. The tumour located in the oesophagus can be resected with a minimally invasive method during thoracoscopy. Patients whose tumours do not exceed 5 cm are qualified for minimally invasive surgery, although some authors suggest that during thoracoscopy it is possible to resect lesions sized even up to 8 cm [15]. It may be difficult to resect smaller lesions than 1 cm, because precise location of the lesion may cause problems. Jiang *et al*. described a group of 40 patients with oesophageal leiomyoma, where in two out of five patients whose tumours were smaller than 1 cm in diameter it was impossible to locate the lesions in thoracoscopy and it was necessary to apply conversion [8]. Resection of the lesion on its own is not safe for all patients. In as many as 10% of cases with the diagnosis of leiomyoma it is necessary to resect the oesophagus. Usually this concerns larger lesions than 8 cm, which occupy the entire oesophagus and strongly adhere to the mucosa [5, 14]. By expanding growth these tumours may cause symptoms resulting from the pressure on the neighbouring structures. There is a report of a case of oesophageal leiomyoma pressing the left atrium of the heart and causing symptoms of dyspnoea and fatigue. The ailments were caused by obstructed flow of blood to the left atrium and the left ventricle [16]. Similarly, in the case under study the tumour closely adhered to the pericardium and the left lobe base, which required careful dissection of the lesion. Resection of the oesophagus with the lesion causes elimination of all ailments and prevents the occurrence of reflux, which is sometimes described in the literature after operations where only the tumour was resected [17]. Besides, many authors note that a long-lasting pressure of the tumour causes thinning and impairment of the muscularis mucosae, which may cause healing difficulties and result in the formation of a fistula [18]. When large oesophageal leiomyomas are treated, a possible transformation into leiomyosarcoma should always be taken into consideration. Therefore, the best procedure with large lesions is resection of the oesophagus with the lesion.

Oesophageal leiomyoma, which is a benign neoplasm, is a serious diagnostic and therapeutic challenge. Smaller lesions sized 5-8 cm may be resected with minimally invasive methods, but lesions sized over 8-10 cm require precise preparation for the surgery, because resection of the oesophagus may be necessary. Combining the skills of a team of surgeons and thoracic surgeons provides an optimal therapeutic procedure to a patient with a large tumour. A complete resection of the lesion combined with resection of the oesophagus was the optimal therapeutic procedure, which led to the patient's recovery.

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