

An unusual case presentation of a palpable abdominal wall mass: extragastrointestinal stromal tumor with literature review

Rzadki przypadek palpacyjnego guza jamy brzusznej: stromalny guz wywodzący się spoza przewodu pokarmowego. Przegląd piśmiennictwa

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Abstract

Gastrointestinal stromal tumors represent the majority of mesenchymal tumors of the gastrointestinal tract. A palpable abdominal mass on physical examination is an extremely rare situation in this diagnosis. These tumors are generally located in the stomach and proximal part of the small intestine, but they can also be found beyond the alimentary tract, i.e. the omentum, mesentery or peritoneum. In this article we present the case history of a 63-year-old female with irrelevant past medical history admitted to the University Hospital for surgical treatment of a palpable abdominal wall mass. Histopathological examination revealed characteristic features for gastrointestinal stromal tumor with unspecific localization. Postoperative hospital stay and 3-month follow-up were uneventful with no reported complaints. We present this case due to the rare occurrence of this kind of lesion and the interesting presentation of this particular case.

Introduction

Stromal tumors are rarely occurring mesenchymal neoplasms. Most of them are localized in the gastrointestinal tract (GI), mostly in the stomach and the proximal small intestine. In 1990, from a wide group of rhabdomyosarcomas a new class was created: gastrointestinal stromal tumor (GIST). It was indicated that this group has common genetic, immunological and mor-

Streszczenie

Nowotwory podścieliskowe są najczęstszymi nowotworami mezenchymalnymi przewodu pokarmowego. Niezwykle rzadko zdarza się, żeby tego typu zmiana została wykryta przy badaniu palpacyjnym. Guzy te zwykle znajdują się w żołądku i jelicie cienkim. Czasami jednak diagnozuje się je poza układem pokarmowym – w sieci większej, okolicach krezki oraz otrzewnej. W niniejszej pracy przedstawiono przypadek 63-letniej pacjentki, bez wcześniejszej istotnej historii chorobowej, którą przyjęto do Akademickiego Szpitala Klinicznego w celu operacyjnego leczenia palpacyjnej zmiany w jamie brzusznej. Ocena histopatologiczna preparatu tkankowego wykazała charakterystyczne cechy dla nowotworów podścieliskowych z lokalizacją poza układem pokarmowym. W pooperacyjnym przebiegu oraz 3-miesięcznej obserwacji nie stwierdzono żadnych powikłań. Autorzy zaprezentowali ten przypadek ze względu na jego rzadkość oraz trudności, jakie może przysporzyć podczas procesu diagnostycznego.

phological features with precursor interstitial cells of Cajal (ICC), pacemakers which create the basal electrical rhythm leading to contraction of the muscle [1]. It is also known that some of them have a different origin, because they can be found outside of the GI, in the omentum, mesentery and peritoneum, where ICC is not observed [2]. This group is called extragastrointestinal stromal tumor (EGIST). Although they are the most fre-

quent nonepithelial tumors involving the alimentary tract, they constitute only 1% of all primary GI neoplasms [3]. Epidemiological studies suggest that the incidence of stromal tumors is approximately 1-2 cases per 100 000 population per year.

They are slightly more often seen in men, mostly over the age of 50 years. Statistically more frequently they occur between the ages of 55 and 65, rarely under 40 years and very rarely in childhood [4]. Stromal tumors are built of spindle cells, epithelioid cells or a mixture of both, usually, but not consistently expressing the KIT protein – transmembrane receptor tyrosine kinase (RTK) [5]. Minor subsets (3% to 5%) of GISTs are negative for KIT by typical immunohistochemical examination. Those are more likely to be located in the stomach. Furthermore, this type of GIST generally has a low mitotic activity and relatively favorable prognosis [6]. The CD117 antigen is part of the KIT transmembrane receptor tyrosine kinase (RTK) that is the product of the KIT proto-oncogene. Discovering this was a great breakthrough and allowed identification of the whole GIST group and its near-universal expression of the CD117 antigen in contrast to leiomyomas, true leiomyosarcomas and other spindle-cell tumors of the GI, which were typically CD117-negative [7]. Stromal tumors used to be classified as benign tumors according to their histopathological features. However, it is now believed that, after a long follow-up, all GISTs have malignant transformation potential [8]. In 2001, Joensuu *et al.* observed a very good impact and good tumor response to imatinib therapy. Since then imatinib mesylate has been a standard treatment therapy in patients with advanced, inoperable or metastatic disease [9].

Case report

A 63-year-old female with irrelevant past medical history was admitted for surgical treatment in our Department of General and Oncological Surgery. Before admittance she was being diagnosed in one of the regional hospitals of Lower Silesia. She complained of pain in the lower abdomen for a long time as the only symptom. During the ultrasound (USG) examination of the abdominal cavity a tumor mass was found on the right side and a suggestion of colon cancer was made. Diagnostics was extended and abdominal computed tomography (CT) was performed. It revealed a large tumor mass, approximately 10 cm in diameter (Figure 1), which had contact with the right liver lobe and was oppressing the ascending colon and hepatic flexure but had no primary location in the gastrointestinal tract. In gastroscopy nothing except gastric reflux was noted, and during colonoscopy the whole way up to the cecum was free of any pathological changes coming out of the colon wall. She was discharged and sent to our University Hospital. On the admittance day, during the physical examinations no deviation from the norm was noticed except a mass, palpable through the skin, and abdominal pain. Routine blood test results were unremarkable. All oncological markers were within correct limits: AFP: 2.89 (1.09-8.04 ng/ml); CA 125: 21.2 (0-35 U/ml); CA 19-9: 11.48 (0-37 U/ml); CEA: 1.04 (0-3 ng/ml). On the next day the operation was performed. An incision was made over the tumor, revealing a large, white-tan, well-capsulated mass, not attached to any intraperitoneal structures, which was described as an EGIST (Figure 2). It was located close to the hepatic flexure. It was removed as one block (Figure 3). Because of the



Fig. 1. Computed tomography of the abdominal cavity with hyperdense mass 10 cm in size

Ryc. 1. Hiperdensyjna zmiana o wielkości około 10 cm w badaniu tomografii komputerowej



Fig. 2. Intraoperative image of tumor resection

Ryc. 2. Śródoperacyjny obraz z wycięcia guza

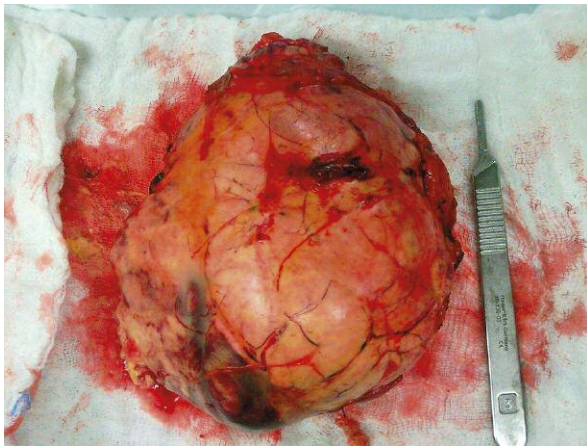


Fig. 3. Large, white-tan, well-capsulated mass evacuated from abdominal cavity

Ryc. 3. Duża, biaława, dobrze otoczona zmiana usunięta w całości z jamy brzusznej

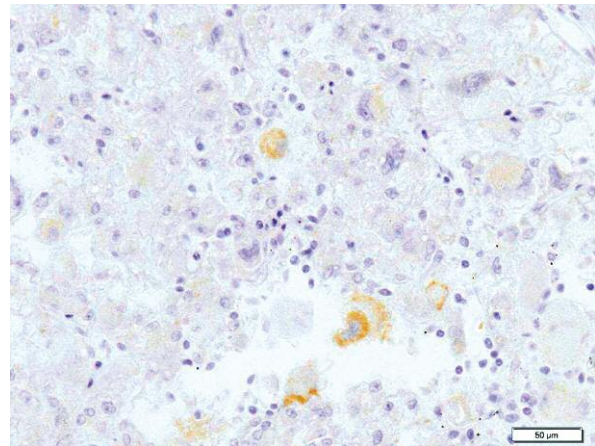


Fig. 4. Strong positive expression of CD 117 in tumor cells

Ryc. 4. Silna ekspresja CD117 w komórkach guza

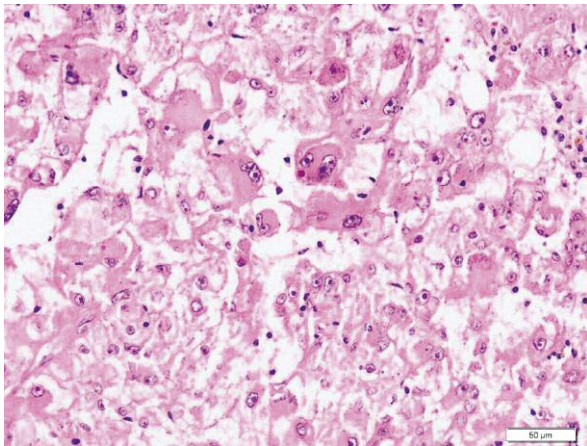


Fig. 5. Pleomorphic type of gastrointestinal stromal tumor (GIST) (hematoxylin and eosin staining)

Ryc. 5. Typ pleomorficzny guza podścieliskowego (GIST) (barwienie hemoksyliną i eozyną)

stone in the gall bladder's cervix a cholecystectomy was performed as well. Histopathological examination of the specimen revealed a well-circumscribed mass, about 11 cm in diameter, in cross-section pale pink, solid externally and soft in the middle. Extensive hemorrhage, foci of myxoid degeneration and focal necrosis were present. Immunohistochemical analysis showed strong positive expression of CD117 in tumor cells and negative staining for CD34 (Figure 4) (which can be negative in 25% cases of GISTs [10]), vimentin (negative in 5% of GISTs [10]), desmin, CK7, and CK20. The mitotic activity was 3-4 mitoses/50 high-power fields (HPF). The histopathological diagnosis was GIST, pleomorphic type (Figure 5). The patient had a regular, uneventful hospital stay and was

discharged after 7 days. We can report 3 months of follow-up with no complaints.

Discussion

Extragastric stromal tumors are quite a unique group, which shares the same features with the regularly occurring gastrointestinal tract tumors. Their pathology remains undiscovered. It is known that tumor size, mitotic rate and location of a primary mass are crucial for the long-term prognosis. According to the 2002 consensus virtually all GISTs have malignant potential. Mitotic count over 5/50 HPF, and tumor size more than 5 cm are two very strong risk factors [11]. Miettinen *et al.* decided to extend the 2002 consensus criteria by describing the primary location of a tumor in relation to risk of disease recurrence [5]. They observed that intestinal GI tumors are more malignant than those located in the stomach. They reviewed the data collected from 1765 cases of patients with gastric GIST (mortality was 17%) and compared them with 906 patients with small bowel GIST (mortality was 39%) and the difference was statistically significant. The TNM staging system for GIST was developed and proposed by the American Joint Committee on Cancer (AJCC) and International Union Against Cancer (UICC) and published in the 2010 7th edition of the cancer staging manual [12].

Stromal tumors are very difficult to diagnose, because a lot of them remain asymptomatic for a long time and patients report no complaints at all. They may be revealed incidentally, during randomly performed abdominal ultrasonography or during endoscopic examination. Additionally, they may present some nonspecific symptoms such as early satiety or bloating unless they grow enough to cause pain, obstruction or GI

bleeding. Metastases are mostly found in the liver or peritoneum, sometimes in the regional lymph nodes.

Computed tomography (CT) or endoscopic ultrasound (EUS) may be very useful in the diagnostic process, but what is more important, those examinations may also define the recurrence risk. Tumors bigger than 5 cm, circumscribed, which have mesenteric fat infiltration, regional lymphadenopathy or an exophytic growth pattern, are more likely to metastasize [13].

A preoperative biopsy is not generally recommended for operable tumors which in the CT are already diagnosed as a GIST. It may be helpful in the evaluation of possible resection and the characteristics of metastasis. It can be useful to determine the CD117 presence and possibility of imatinib therapy especially in advanced tumors which after inhibitor treatment may change into operable masses. If a biopsy is undertaken, EUS-guided biopsy is preferred over a percutaneous biopsy [14].

Conclusions

Extragastric stromal tumors are very rare tumors. This group is poorly known and provides a lot of diagnostic and therapeutic problems. Despite our wide knowledge about tumors occurring in the gastrointestinal tract, there is a lack of data about patients with EGIST, which could help with providing standard guidelines. This is the reason we decided to publish this material. It seems that even without such standards we can treat patients with the available knowledge. CT or diagnostic biopsy is a good examination to assess the presence of CD117 or the usefulness of preoperative imatinib therapy. More studies are necessary to establish the prognostic factors related to the primary mass localization of the EGIST. Thus follow-up for a long period of time is required.

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