GIGANTIC SOLITARY FIBROUS TUMOUR OF EXTRA-PERITONEAL SPACE. A CASE REPORT AND REVIEW OF THE LITERATURE

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Solitary fibrous tumour (SFT) is a rare soft tissue tumour which has been described in pleura by Klemperer and Rabin in 1931 (1). According to the WHO classification SFT is a mesenchymal tumour that belongs to fibroblastic/myofibroblastic group. Clinically is a slow growing neoplasm that appears in mid-aged patients, independently of sex. Course of disease is unpredictable. The most common localisation is pleura but SFT was also described in different localisation such as peritoneum and extraperitoneal space or mesentery (2).

CASE REPORT

71-years old patient was admitted to the Department of General, Gastroenterologic and Oncologic Surgery because of abdominal mass revealed by palpation of abdominal cavity. The main complain was constipation and leg swelling for about 2 months before admission. Rising abdominal diameter forced patient to perform GP visit. Tumour in abdominal cavity was revealed by ultrasonography, computer tomography (CT) scan confirmed its large diameters: 224x136x256 mm. CT scan also showed necrotic sites and calcinosis, strong impression on abdominal part of aorta (fig. 1). Radiologist’s suggestion was sarcoma with no defined source in abdominal cavity. After on-
cologic consultation patient was transferred to the Department of General, Gastroentero- logical and Oncologic Surgery for further examination. On admission he was in good general condition, pain negative, with anemia (Hb 11.6 g/dl) and increased Fibryn level up 505 mg/dl and C-reactive protein (CRP) up 14.3 mg/l. On examination atrial flotation was confirmed. After cardiologic consultation and echocardiography (ECHO) investigation patient was qualified for operation on schedule. Before operation patient got antithrombotic and antibiotic prophylaxis as follows: Delta-parin 5000 iu (1 day before), Cefazolin 1 g (30 minutes before incision).

Surgical treatment

During operation abdominal mass of about 6-8 kilograms weight filling all abdominal cavity was confirmed (fig. 2). There was no other internal organs infiltration. Tumour’s surface was coated by many pathologically enlarged vessels with heavy bleeding on touch (fig. 3). Tumour was extirpated, it came out of retroperitoneal space (fig. 4). On the same day, late night, patient was reoperated because of heavy intra abdominal bleeding. On investigation during laparotomy source of bleeding was not found but many vessels were assessed to be at high risk of recurrent bleeding and preserved. Patient got 6 units of fresh frozen plasma and 6 units of packed red blood cells. During post-operative period there was prolonged bowel immobilisation, full oral diet was implemented on day 6th, diet was well tolerated. Patient was discharged on 28th day of hospitalization and 19th day after operation, clinically in very good condition. Histopathological report of excited tumour revealed: solitary fibrous tumour, immunohistochemically CD34(+) (fig. 5), CD117(-), actin (-), desmin (-), S100 (-), CD31(-), CD99(+) (fig. 6), bcl2 (+). Low mithotic activity was confirmed by proliferation index MIB 3%, focuses of necrosis (fig. 7), nuclear polymorphism could indicated malignant disease in this case. Follow-up period is negative, up till April 2013 patient is in good condition, with no complains.
DISCUSSION

First histopathological solitary fibrous tumour characteristics as a pleural tumour was published in 1870 by Wagner. In 1931 Klemperer and Rabin described SFT as differentiated serosal origin and in 1942 Stout and Murray as mesothelial tumour. Today, it is widely confirmed that SFT is fibroblastic mesodermal neoplasm with its characteristic branched vessels that makes it similar to hemagiopericitoma (HPC) tumours. Despite similar immunophenotype SFT is histopathologically different than HPC, has no organized architecture. Typical for SFT is glazing in matrix and around vessels, rich presence of collagen fibre. Similar to HPC it shows expression of CD34 (80-90%), CD99 (70%), rarely bcl-2 (30%), epithelial membrane antigen (EMA, 30%) and smooth muscle actin (SMA, 20%). Usually, there is no expression of S100 protein, desmin and cytokeratin. Probably, because of unclear SFT and HPC differentiation previously, many HPC tumours would be recognised as SFT today. During last 10 years, because of high interest in these types of tumours, they became better recognized, new classifications, subclassification and prognostic markers were established. There were also different types of SFT distinguished, such as fibrous, cellular, adipose or gigantocellular tumour (3).

SFT origin can exist not only in serosal localisation like pleura, pericardium or peritoneum but in soft tissues and visceral organs also (4). During last 20 years this kind of tumour was diagnosed in extra-pleural localisation mainly (5).

Clinically, SFT of soft tissue is a slow-rising tumour, expressed in medium-aged person, with no sex relationship (6). It can reach over 10cm diameter, that can cause pressure and dysfunction of different organs. Sometimes it can give paraneoplastic signs as hypoglicemia because of insulin-like growth factor (IGF) secretion (7). The most common extra-pleural localisation is retroperitoneal and peritoneal space, soft tissues of extremities and head. Disease progression remains to be unpredictable. 10-15% of all cases are malignant disease, with metastases to lungs, liver and bones. Malignancy signs of tumour are diameter over 5 cm, high proliferation index MIB (over 2%), nuclear polymorphism, focuses of necrosis, high mitotic index (over 4 mitoses/10 FOV).

Unfortunately, in described case histopathological investigation revealed almost all of these signs what claim to be a negative prognostic factor.

Solitary fibrous tumours are published rarely. Yamashita et al. described tumour 15x14x10 cm confirmed as SFT. There was low nuclear polymorphism and mitotic activity (0-2 mitoses/10 FOV), proliferation index 2. During 26 months follow-up period authors didn’t confirmed recurrent or metastatic disease (8). Similar tumour, in 24-years old woman described Cristi et al. (9). Despite benign neoplasm confirmed by histopathology examination authors proposes very strict and long lasting follow-up period because of unpredictable disease progress and high risk of very late recurrence rate. Lau et al described 53-years old man who was suffering from groin pain, with nausea, lack of appetite, diarrhoea. CT scan revealed 22x18x11 cm size tumour in right iliac fossa. During operation investigation showed tumour in distal ileum mesentery (10).
Similar tumour was described by Bouhabel et al. in 71-years old woman. Both authors maintain that these were two first publications about SFT in mesenteric localisation.

Thanks to improvement of diagnostic methods there are much higher extra-pleural SFT identification. Solitary fibrous tumour is a low-signal and potentially benign neoplasm but there is no assessed malignancy conversion rate, metastases appearance and recurrence rate risk. Patients after curative SFT operation should be under strict oncologic supervision, in the same way as patients with malignant disease confirmation.

REFERENCES


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