CALCIFYING FIBROUS TUMOR OF THE SMALL BOWEL MESENTERY IN A 27-YEAR OLD MALE PATIENT – CASE REPORT

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Calcifying fibrous tumor is a rare disease entity, usually concerning the soft tissues of the limbs, neck, trunk, or scrotum. Cases of the above-mentioned pathology have also been reported considering the pleural and peritoneal cavity, and small bowel mesentery. The essence of the disease, whose etiology and pathogenesis remains unclear, is the fibrous tissue infiltration and diffuse inflammation with focal calcifications. The study presented a case of a 27-year old male patient subjected to surgical intervention, due to an abdominal cavity tumor. The tumor was radically removed, and its character and definitive diagnosis were established postoperatively. After a seven-year follow-up period, recurrence was not observed.

Key words: small bowel mesentery tumor, CFT

Calcifying fibrous tumor (CFT) is a benign, usually single lesion of mesenchymal origin of uncertain etiology. The above-mentioned most often is diagnosed in children and young people. Its location in the gastrointestinal tract is very rare. Literature data has described isolated cases of CFT of the small bowel mesentery (1, 2).

The study presented a case of a 27-year old male patient subjected to surgical intervention, due to an abdominal cavity tumor.

CASE REPORT

A 27-year old male patient (history nr: 14362/2006) without a previous surgical history was admitted to the Department on November 27, 2006 for the diagnosis and surgical treatment of an abdominal cavity tumor. The patient complained of dull abdominal pain located around the naval, which was present for the past seven months. He complained of lack of appetite and progressive weakness. No other symptoms were observed. Abdominal ultrasound and CT showed a retroperitoneal lesion at the level of the hip bone plates, in the vicinity of the aorta and vena cava inferior. Abdominal CT was as follows: in the retroperitoneal space at the level of the hip bone plates, visible well-demarcated tumor, 5.6x5x6 cm in size, with numerous calcifications, being in favor of a proliferative lesion (fig.1 and 2). In its vicinity visible enlarged lymph nodes, their size reaching 2 cm, enlarged lymph nodes along the mesentery artery. The liver was normal in size without focal lesions. The biliary ducts without pathology. The gall-bladder, normal in size, without calcifications. Normal spleen and pancreas. Both kidneys normal in size, with a 1 cm cyst located in the right upper calyx. No other pathologies were observed.

The remaining diagnostic examinations (chest X-ray, gastroscopy, colonoscopy, testicular ultrasound) showed no pathology. Laboratory results (morphology, urine sample,
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CRP and tumor markers (AFP, Ca 19-9, CEA, HCG) were within normal limits. The patient was qualified and prepared for surgery. After opening the abdominal cavity by means of a midline incision we observed the presence of a small bowel mesentery tumor, 6 cm in size, located 80 cm from the suspensory ligament of the duodenum. In the vicinity of the lower pole of the tumor we observed the presence of adjacent upper mesenteric vessels and enlarged lymph nodes (fig. 3). The liver and remaining abdominal cavity organs showed no pathology, both macroscopically and during intraoperative palpation. The superior mesentery artery and vein were prepared. The tumor was subjected to resection including a healthy tissue margin with segmental resection of the small bowel, due to ischemia. End-to-end anastomosis was performed, the mesentery closed by means of interrupted sutures. Due to lack of previous verification and macroscopic suspicion of a neoplastic lesion, the site of organ removal was marked by titanium clips, in order to perform future radiation therapy.

Antibiotics, analgesics, and anti-thrombotic therapy were used during the postoperative period. On the second postoperative day the nasogastric tube was removed, while on the fifth the abdominal cavity drain. Ten days after the operation the patient was discharged from the hospital in good general condition with normal peristalsis and the postoperative wound healing by first intention.

The postoperative histopathological examination (nr 138768-88) showed the presence of a tumor, 5.5x4x4.5 cm in size, well-demarcated, within the mesentery numerous enlarged lymph nodes. The texture of the tumor showed elements of fibrous tissue with central calcifications, and peripheral abundant lymphocytic infiltrations with the development of lymphoid follicles and presence of multinucleated cells. The texture of the tumor was location to eosinophils, few granulocytes and plasma cells. Twenty-three lymph nodes were of normal texture. Diagnosis: calcifying fibrous tumor, radical surgery.

The seven-year observation period and numerous abdominal CT examinations should no signs of tumor recurrence. Currently, the patient is feeling well, actively working, reporting no complaints.

DISCUSSION

For the first time in 1993, Fetsch et al. (3) described the above-mentioned disease entity in 10 patients, aged between 1 and 33 years, identifying them as calcifying fibrous pseudotumors.

The World Health Organization (WHO) in 2002 established the above-mentioned as calcifying fibrous tumor (CFT). The lesion is a separate clinical and pathological entity,
originating from the soft tissues of children’s extremities, although may also be observed on the trunk, neck, arms, and pleural cavity. It is most often observed in case of young adults (1, 4, 5). Approximately, thirty cases have been described, considering patients diagnosed with abdominal CFT (4), confirmed by means of histopathological examinations. However, the incidence of CFT of the gastrointestinal tract is very rare (2). The average age of patients with abdominal CFT was 34 years, while in case of superficial soft tissues the average age was 16 years (4). The etiology and pathogenesis of calcifying fibrous tumors remains unclear. Clinically, one may observe the presence of an abdominal tumor, with a history of several weeks or months, without characteristic symptoms. If the lesion is large in size, it may provide compression symptoms to adjacent structures, such as pain or symptoms of ischemia. In the presented study case the tumor was located in the vicinity of superior mesenteric vessels, and its progression could lead towards significant consequences and intensification of clinical symptoms.

Literature data presented patients without symptoms for many years, despite the large size of the tumor. Imaging examinations showed no typical signs of CFT, results being unspecific of one given disease entity. The most common image of CFT includes thick, irregular or focal microcalcifications, which are better visible on abdominal CT and ultrasound, as compared to X-ray examinations. The MRI examination showed a fibrous tumor of a hypotensive signal in T1 and T2 dependent images (6). The image of the calcification may be variable during tumor growth, due to developing necrotic foci or hemorrhagic extravasations. Small lesions which are usually asymptomatic may be missed during imaging examinations and their diagnosis is often accidental.

Macroscopically, CFT is a well-demarcated, uniform tumor of different size, low in cellular content, and non-encapsulated. CFT can infiltrate surrounding tissues and the surface of the incision may be of granular structure. Microscopically, it presents the characteristics of all inflammatory pseudotumors, including benign vitreous collagen fibrous infiltrations, few fibroblasts, and healthy tissue plasmocytes presence. As compared to other inflammatory pseudotumors CFT is characterized by abundant calcifications (7). Microscopically, the tumor is characterized by low-cellular content fibroblast proliferation, focal plasmocytic, eosinophilic, and mast cell infiltrations. One may observe significant collagenisation with various degrees of calcifications and diffuse cellular dystrophy, psammomatous calcifications.

The histopathological differential diagnosis should include such lesions as myofibroblastic inflammatory tumor, idiopathic retroperitoneal fibrosis and associated fibrous-inflammatory sclerotizing infiltrations, desmoplastic type tumor - fibroblastoma and calcifying aponeurotic fibroma. Usually the disease is characterized by the presence of a single tumor, most common location being the mediastinum, subcutaneous tissue, deep soft tissues of the extremities, head and neck, as well as the stroma of different organs (6, 8). Literature data mentioned isolated cases of multifocal disease or family history (5).

Due to the rare occurrence of gastrointestinal CFT there is increased risk of diagnostic error. The above-mentioned is location too other, statistically more common calcifying fibrous tumors (3). Radiologically, these tumors might mimic features of malignancy, clinically presenting acute abdomen symptoms (2). The main diagnostic-therapeutic problem is to determine the character of the lesion, differentiating between its benign and malignant nature. Proper preoperative diagnosis is extremely difficult or often impossible, mostly due to the lack of a clearly established etiopathogenesis and atypical course. Initially the disease is asymptomatic revealing itself only when the tumor has reached a considerable size or when diagnosed accidentally. Available imaging examinations are also not capable too unambiguously determine the character of the lesion.

Considering the presented study case the patient reported to his family doctor, due to abdominal pain and initial diagnosis was established on the basis of an abdominal ultrasound. Although further evaluation failed to determine the character of the lesion, which was suspected of malignancy, only surgical excision enabled too establish the final diagnosis. Prior to surgery, biopsy was considered, but due to the location in the vicinity of the mesenteric vessels its implementation was abandoned. Lack of preoperative diagnosis did not alter further management, the patient being informed and such conduct obtained his consent.
Differential diagnosis of such lesions should include retroperitoneal space enlarged lymph nodes, gastrointestinal stromal tumors (GISTs), and neuroendocrine tumors (GEP-NEN) (1, 9). Considering non-neoplastic pathologies one should mention lipomas, myomas, cystic lesions, neurofibromas, small bowel desmoids (10), different connective tissue diseases, and Inflammatory Myofibroblastic Tumor, which was often considered in literature data as the preceding condition of CFT, but eventually was declared as a distinct disease entity (11). CFT treatment of choice consists in the radical excision of the primary and possible recurrent lesions. Long-term observation is necessary, both in case of incompletely excised or diffuse lesions, as well as those radically excised, due to the unpredictability regarding possible recurrence. Recurrence or metastasis are rarely observed (12). Diagnosis and thus, effective treatment requires the determination of the character of the lesion, which is not always possible, prior to surgical intervention. In the presented study case the character of the lesion was determined post-operatively, on the basis of the histopathological examination.

In conclusion, we presented a patient with a rare gastrointestinal CFT, which was accidently diagnosed and subjected to radical surgery. The seven-year observation period proved uneventful—there was no recurrence. In the absence of characteristic symptoms and specific markers diagnosis is only possible on the basis of the histopathological examination of the excised lesion. CFT resection is usually curative and ends further treatment, although due to the possible local recurrence the patient requires periodic follow-up visits. In the absence of characteristic calcifying features on imaging examinations, in order to distinguish between CFT and other rare abdominal soft tissue tumors, postoperative histopathological evaluation is decisive.

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