

CALCIFYING FIBROUS PSEUDOTUMOR OF THE SMALL BOWEL MESENTERY – CASE REPORT

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Calcifying fibrous pseudotumor (CFPT) is a benign mesenchymal tumor diagnosed in children and young adults, located in the subcutaneous tissue of the trunk and limbs. Its intraabdominal localization is a unique rarity. The Authors of the study presented a case of a 48-year old female patient with an accidentally diagnosed small bowel mesentery tumor during surgery.

Key words: calcifying fibrous pseudotumor, calcifying fibrous tumor

Calcifying fibrous pseudotumor (CFPT) is a benign lesion derived from fibrous connective tissue. It was first described in 1988 by Rosenthal and Abdul-Karim as “a fibrous tumor with psammoma bodies occurring in children” (1). Its presence in young adults has been confirmed for several years (2). The etiopathogenesis of the lesion remains unknown. Most literature data reports concerning treatment are based on single case studies. Prognosis is good and recovery is obtained after its surgical removal.

The study presented a case of a 48-year old female patient with an accidentally diagnosed small bowel mesentery causing chronic gastrointestinal obstruction.

CASE REPORT

A 48-year old patient (H.B.) (hist. nr: 2656/2012) was admitted to the Department of Gynecology, Pomeranian Trauma Center, due to acute hypogastric pain (myomatous

uterine) and loss of body weight. Abdominal ultrasound showed the presence of a solid, hypoechogenic lesion with polycyclic contours located in the small pelvis, 9 cm in diameter, suggesting the possibility of a myomatous uterus. Considering the physical and imaging examinations the patient was qualified for emergency hysterectomy with suspicion of torsion of a peduncled uterine myoma.

Laparotomy was performed by means of Pfannenstiel’s method showing a small uterus with an intramural anterior wall myoma, 3 cm in diameter, as well as a small peduncled myoma in the fundus of the uterus, 5 mm in diameter. Within the small bowel mesentery, 190 cm from Treitz’s flexure, a solid, chondroid tumor was observed, gray and cream in color, 15x8x8 cm in diameter causing chronic gastrointestinal obstruction. The remaining abdominal cavity organs showed no pathology. The small myomatous uterus with normal adnexa did not require surgical intervention. The procedure was taken over by the surgical team.

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The tumor along with a healthy mesentery tissue margin was excised together with a segment of the small bowel, 26 cm in length. Gastrointestinal continuity was restored by means of entero-enterostomy, end-to-end, using PDS 4/0 interrupted sutures.

Based on the histopathological examination of numerous tumor samples performed at The Department of Pathomorphology (number 281100) and immunohistochemical examinations, such as: CD34 (focal+), S-100 (-), SMA (focal+), CKAE 1/AE3(-), B-catenin (-), CD117 (-), ALK1 (-), desmin (-), as well as histochemical staining, according to Masson, Gomoriego and negative staining for amyloid, calcifying fibrous pseudotumor was diagnosed.

The postoperative period proved uneventful. The patient was discharged from the hospital on the fifth postoperative day in good general condition with a normally healing wound. The control examination showed proper postoperative recovery.

DISCUSSION

Calcifying fibrous pseudotumor (CFPT) is a soft tissue, benign tumor derived from fibrous connective tissue. In children and adolescents it is most often localized in the subcutaneous tissue of the trunk and limbs. In young adults CFPT cases were described to be present in the pleural cavity, mediastinum, retroperitoneal space, adrenal glands, and liver (3-6).

In general the tumor is usually diagnosed as a single lesion, although it may also be multifocal.

The location within the abdominal cavity, described in literature data is varied. Publications available in databases such as Medline (PubMed, Ovid), Scopus, ScienceDirect (Elsevier) and SpringerLink were analysed using the following key words: "calcifying fibrous pseudotumor" and "calcifying fibrous tumor".

By 2012, 49 cases of calcifying fibrous pseudotumor within the abdominal cavity were described. Analysing available literature data one can come to the conclusion that in most cases the lesion is single, its incidence observed in the stomach (12 cases), small bowel (11 cases), and retroperitoneal space (4 cases). The single, small bowel mesentery tumor was diagnosed in two patients (7, 8). Multifocal lesions involving the

serosa, mesentery, greater omentum, parietal peritoneum, and in selected cases the mesorectum and liver were described in 12 patients.

Most of the lesions are accidentally diagnosed during laparotomy, due to other indications, or draw attention because of local tumor growth.

Small tumors located in the abdominal cavity are usually asymptomatic and accidentally diagnosed. Clinical symptoms, if present, are usually unspecific and dependent of the location and size of the lesion. The most common symptoms include chronic abdominal pain, bloating, abdominal distention, and a palpable pathological mass.

Often, gradual tumor growth is the basis for emergency surgical intervention, due to developing complications. Liang et al. (9), and Emanuel et al. (10) observed patients with gastrointestinal obstruction symptoms, due to small bowel intussusception. Ben-Ihzak et al. (7) described small bowel mesentery torsion of CFPT located in the root of the mesentery. Jaundice may also appear as the first symptom – Zhang and Yan (11) presented a case of a 60-year old male patient with increasing parameters of cholestasis, due to CFPT growth in the lumen of the extrahepatic bile ducts.

Preoperative imaging diagnostics (ultrasonography, abdominal CT, endoscopy) demonstrating abdominal tumor presence is not sufficient to establish proper diagnosis, and the clinical diagnosis of calcifying fibrous pseudotumor is based on histopathological and immunohistochemical examinations.

Macroscopically, CFPT is a well-delimited, non-encapsulated tumor, 1 to 15 cm in diameter.

The microscopic image is dominated by a low-cellular count of collagenized fibrous connective tissue with the presence of spindle cells and psammoma bodies. There may also be small areas of higher cellularity of fibro- and miofibroblasts. Within the tumor one may observe a lympho-plasmocytic inflammatory infiltration of varying severity.

Considering immunohistochemical examinations CFPT show a positive reaction for the presence of vimentin and factor XIIIa. Reactions for the presence of actin, desmin, factor VII, protein S100, cytokeratine are negative, while in case of antigens CD34, SMA and CD31 they might prove focally positive (13). The

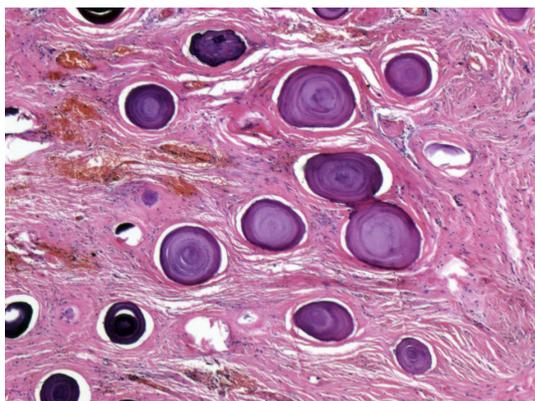


Fig. 1. Tumor area with the accumulation of psammoma bodies. Collagenized stroma in the background (H&E x40)

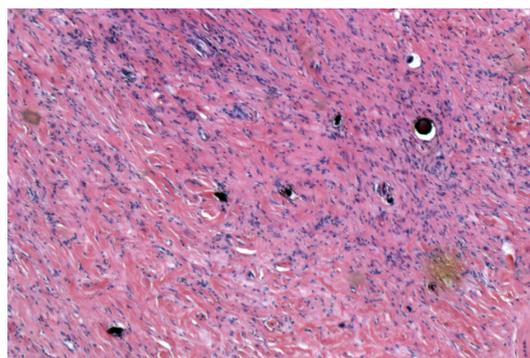
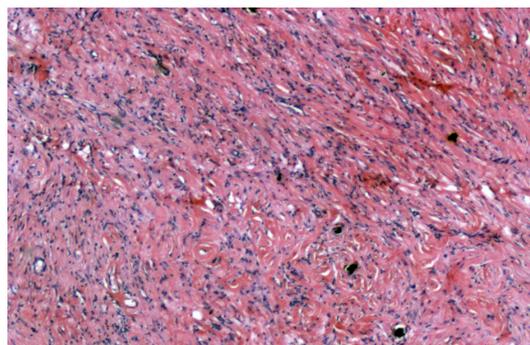


Fig. 2 and 3. Tumor area with predominance of collagenized stroma and a population of spindle cells and psammoma bodies

mytotic and proliferative index is very low or equal to zero (fig.1, 2, 3).

Histopathological differential diagnostics of CFPT includes GIST (gastrointestinal stromal tumors) tumors, desmoid tumor, inflammatory myofibroblastic tumor, and solitary fibrous tumor. It is necessary to perform immunophenotyping of the tumor in confrontation with the morphology of the lesion.

CFPT does not give metastases, malignant transformation was not observed. After total surgical excision of the tumor prognosis is good, although local recurrence was observed in isolated cases (12).

The presented case of a 48-year old female patient with accidentally diagnosed CFPT of the small bowel mesentery underline the rarity of its occurrence within the abdominal cavity.

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