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A CASE REPORT

Malignant Gland Retroperitoneal Extra - Gastrointestinal Stromal Tumour

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Abstract

In this paper we introduced a rare pathology of a 69 years old patient (female), who underwent a surgery at the "Kavaja Hospital" in Prishtina, due to a huge tumour (extra gastrointestinal stromal tumour) spread upon 2/3 of her abdominal cavity. Tumour formation covered retroperitoneal space, from which it originated, and had penetrated up to the pelvic and intraperitoneal space, by covering subdiafragmal region of both parts. The tumour was in contact with the left lobus of the liver and with the caudal part of the pancreas, located behind the stomach (in omentalis stock) in curvatura maior where it had infiltrated a part of its wall. Neoplastic process had infiltrated spleen chyllus, as well. During the abdomen exploration left kidney with its over kidney gland were presented, structured not involved in the pathological process. Meanwhile, left ureter partially was compraised by causing hydronephroses of grade II, as it was prescriped in computerized tomography (CT) examination. Tumour size was incapsulated with thick walls, covered by inflamed omentum, while tumoural space had necrotic detritus, with hemorragic contents verified by punction while operation. Tumour had cystic and septs spaces in between, accompanied with many adhesions in intra-abdominal organs. Abdominal space was oppened with infa and supraumbilical as well and with subcostal incision from left side sec. Courvoisier.

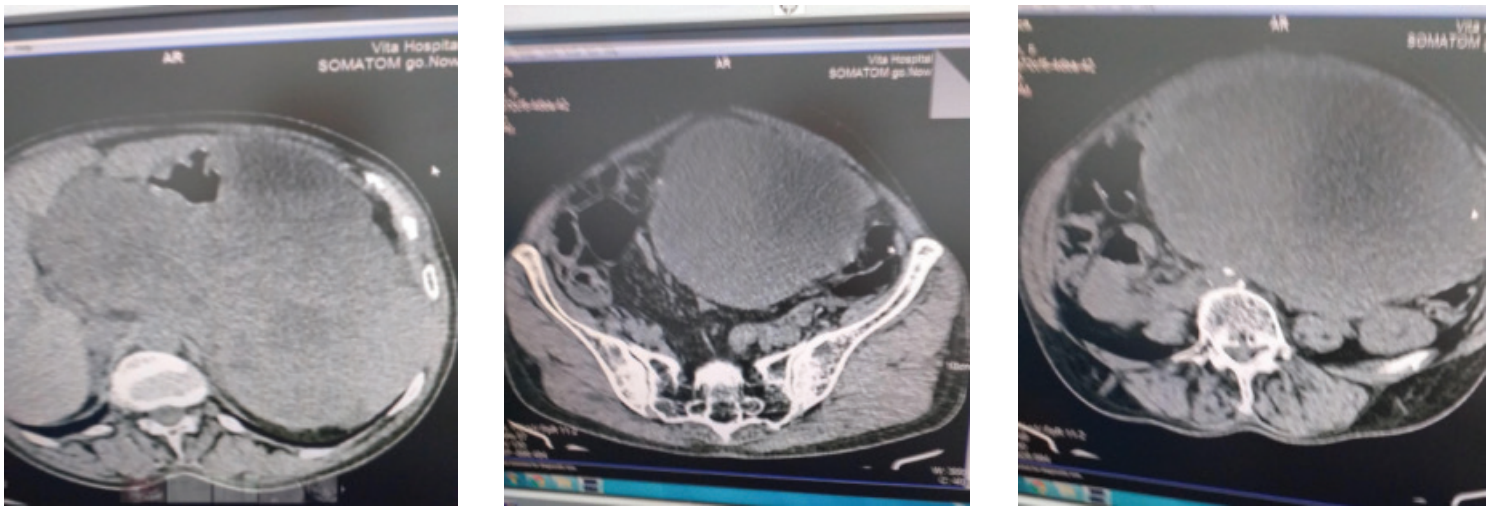
Tumour mass was fully removed via the excision of a part of stomach wall in height of curvature maior followed by a partial omentectomy. This was done after the tumour was released from many adhesions around organs. Microscopic preparatory removed by operation, was sent in hystopatologic examination, undergoing a series of tests: lab. analyses, tumour markers, physico-clinical examinations, ultrasound and CT of the abdomen. After pre-operative preparation, the patient was operated under general endotracheal anesthesia. The operative and post-operative phase went smoothly after the application of resuscitation measures where several doses of blood and plasma were prescribed along with other infusion solutions. We conclude that in these cases we should insist on removing the tumor as a whole. This procedure carries a better prognosis for the patient to ensure longer survival in the post-operative course, in addition to the relevant therapeutic measures according to the histopathological form of this neoplastic process.

Keywords: Extra-gastro-intestinal stromal tumour (GIST) retroperitoneal, diagnosis, the approach of surgery intervention.

Introduction

The tumor formation known as GIST was first described by Golden and Staut (1). The term "stromal tumor" is derived from Miettinen and Mazur Clark-1983 (2, 3). GISTs arise from mesenchymal tissue due to mutations

Photo 1. Patient CT (Z. N, 1951). The tumor formation is seen on a larger surface, which is compact.CT -Description:



in genes exon 9, 11, 13, and 17, influenced by the Thyroxine Kinase enzyme on the receptors of the Procto-oncogene or C-KIT-CD-117 gene (4). In the digestive system, it is formed by Kajli's cells, while in the retroperitoneal location, it is formed by connective tissue (5). Special forms of the GIST tumor are the Familial GIST and the pediatric one where the gene mutation in the latter is the PDGFRA (platelet-derived growth factor receptor alpha) gene, particularly exon 18, in 85% of cases (6). The familial form may be inherited as an autosomal dominant trait, though it is more common in patients with neurofibromatosis type I, occurring in 5% of cases. This type of tumor was previously called a leiomyoma or leiomyoblastoma.

The GIST tumor is more often located in the esophagus, duodenum and jejunum, while less often in the esophagus, rectum, appendix, omentum and in the retroperitoneal location. Its average size is 15-30 cm, but it can also be millimeter in size. The GIST tumor in the differential diagnosis must be distinguished from other tumor formations such as: leiomyomas, leiomyosarcomas, diverticulosis, lymphomas and from benign formations in the small intestine (7, 8).

The definitive diagnosis of the GIST tumor is determined in the histopathological examination accompanied by immuno-histochemical examinations with the application of certain CD-117 antibodies (c-KIT) (9, 10, 11).

Materials and Methods

In abdominal pathology diagnostics, various methods (including advanced ones) are used based on indications or when cases are unclear, in both elective and emergency scenarios. These methods include detailed patient history, clinical examination, laboratory analysis including tumor markers, abdominal ultrasound, native abdominal X-rays, and computed tomography (CT). Additional methods such as positron emission tomography (PET) and endoscopic ultrasound examination (EUS) may also be utilized.

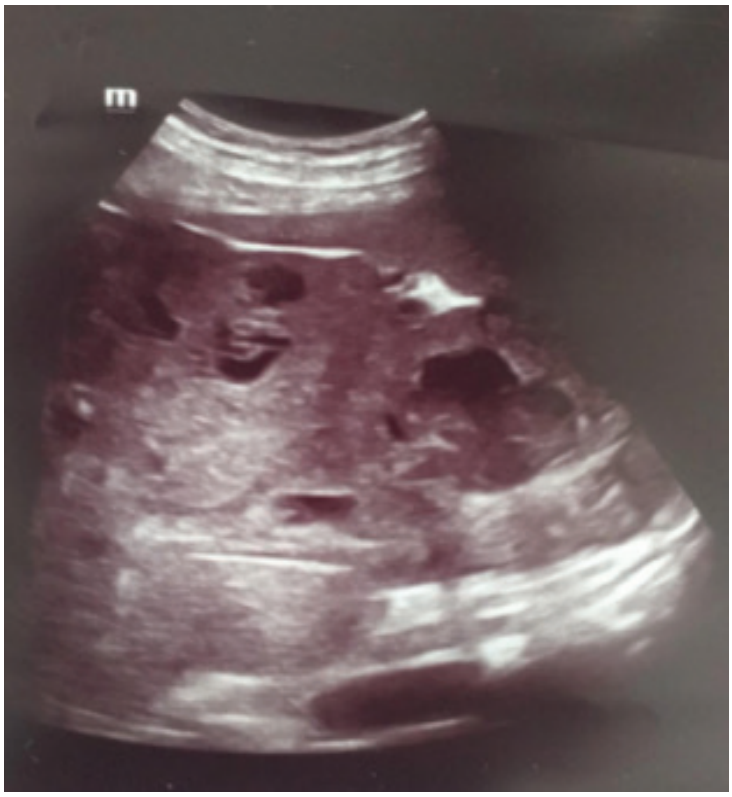
The definitive diagnosis in this case was achieved through histological and immuno-histological examination, employing antibodies such as CD117 (c-KIT), Vimentin, CD34, S-100 protein, HMB45, Melan SMA, Desmin, Chromogranin, CD 68, and EMA. The findings indicate a high and diffuse expansion.

CT of the Abdomen revealed a tumor measuring 19 x 11.5 x 25-30 cm located subdiaphragmatically on the left, in contact with the spleen and its hilum. It extends into the thorax from the major curve, contacting the left lobe of the liver, and further distally from the adrenal gland and left kidney to the pelvis. The tumor exhibits lobular contours with solid and cystic components, along with necrotic masses. Urinary stasis from the left side of the grade II with compression of the left ureter was observed. Additionally, small cysts of about 5 mm in both kidneys were noted, clinically insignificant.

Ultrasound Description from the radiologist:

The large mass extending from the left upper abdomen and epigastric region towards the pelvis and ileocecal area is covered in dense contents, distinctly bordered by the spleen, liver, and spleen's large curve above the pancreas, making visualization challenging.

Photo 2. The appearance of the tumour formation in the sonographyc examination of the abdomen.



Case Presentation

Patient Z.N. was admitted to the Surgical Department on July 2, 2020, presenting digestive issues including persistent moderate stomach pain, flatulence, constipation, and brown stool, and experiencing a sensation of heaviness in the stomach area, where palpation revealed a firm mass occupying nearly two-thirds of the abdominal space. These symptoms have been notably aggravated over the past 6 months. Previously, she had been using pain medications prescribed by her primary care physician. Upon admission, she brought along all medical documentation related to the diagnostic imaging methods previously performed.

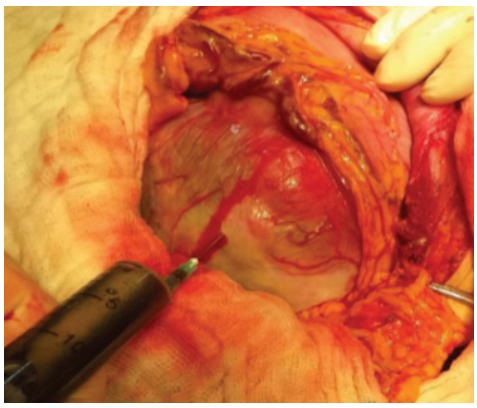
On the laboratory analysis we noted the following: glycemia 5,5 mmol/ L, Urea 4,9 mmol/ L, Creatinine: 68,9 µmol/L, Cholesteroli 5,0 mmol/ L, Fe 2,8 µmol/L). **Immune assays analysis: AFP** 3,9 ng/mL, (>9,5 ng/L), **CEA.** 1,7- (5,8 – 10,0 ng/L) , **CA.. 125** 113,8 (0-35 U/mL) , **CA 19-9**.. 11,5 (0-37 U/mL. **Urin sediment :** Leukocytes-(WBC) -8-10, enough erytrocites , some Amof. Urates, and 3-4 epitelial cells.

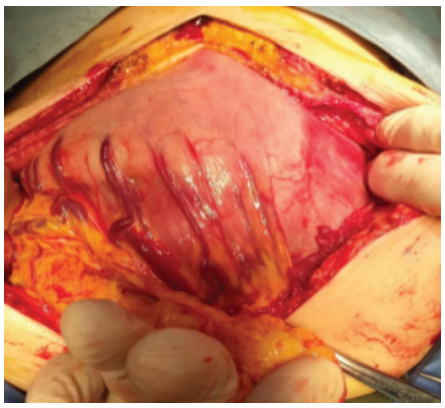
Surgical Techniques

With infra- and supraumbilical, as well as subcostal incisions from the left side sec. Courvoisier, the abdominal space is opened layer by layer. Upon exploration, a tumor formation with a retroperitoneal location is palpated, extending into the pelvis and occupying 2/3 of the abdominal cavity, with the inflamed omentum surrounding it. The tumor, with thick walls, is situated behind the greater curvature. The neoplastic process has infiltrated the spleen hilum. Further, a portion of the inflamed omentum covering the tumor is initially removed via partial omentectomy. The tumor extends bilaterally into the subdiaphragmatic region, exhibiting a lobular structure and firm consistency. Numerous adhesions to abdominal organs such as the liver, mesentery, small and large intestines are carefully dissected using “Ligasure” cutters. Following puncture of the tumor, aspiration and evacuation of hemorrhagic contents and necrotic detritus masses are performed.

Following this, the tumor “en bloc” is excised, along with adhesiolysis in the subdiaphragmatic region on both

Photo 3. Tumour removal progress the appearance of the tumoural with the opening of the abdomen. You can see the changed omentum above the tumoural formation. Punction and aspiration of contents with syringing where hemorrhagic contents are obtained. The beginning of the separation from the surrounding structures and many adhesions.





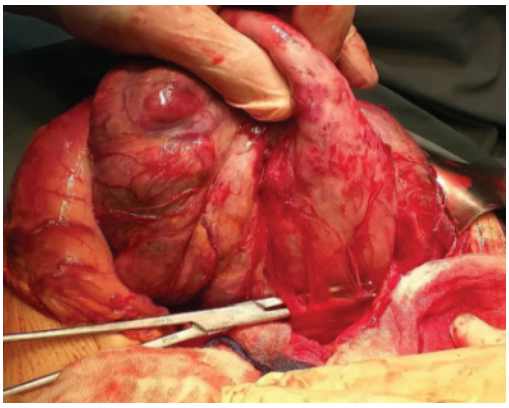


Photo 4. The procedure of removing the tumoural formation from the caudal part of the pancreas.

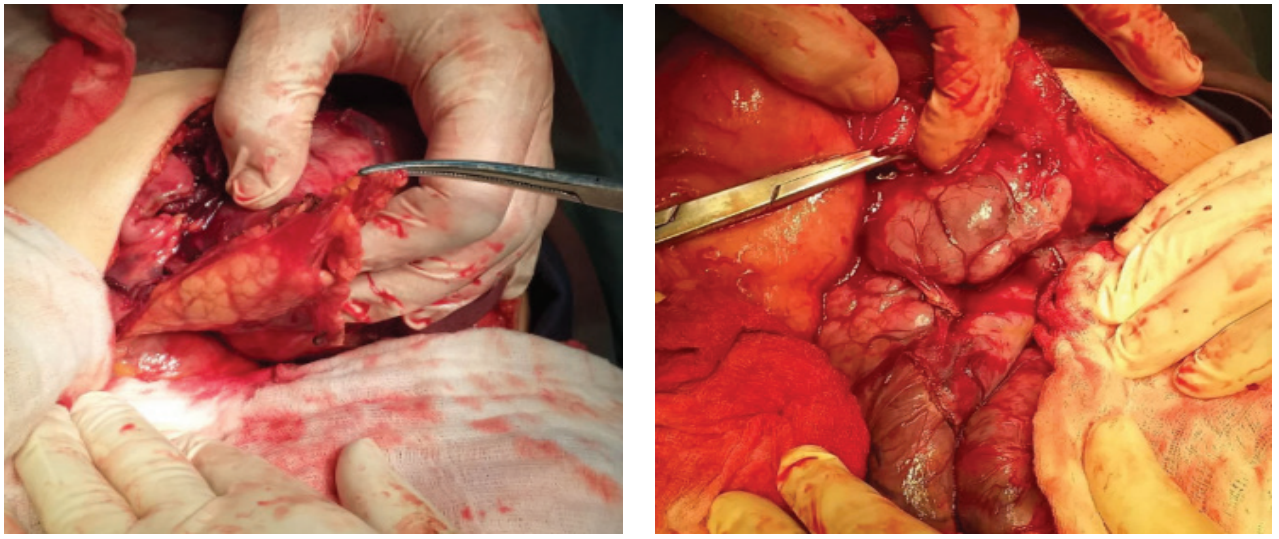


Photo 5. Total extirpation of the tumor "en bloc" with splenectomy.
DG: HP: Malignant Gland Retroperitoneal Extra-Gastro-intestinal stromal tumor.

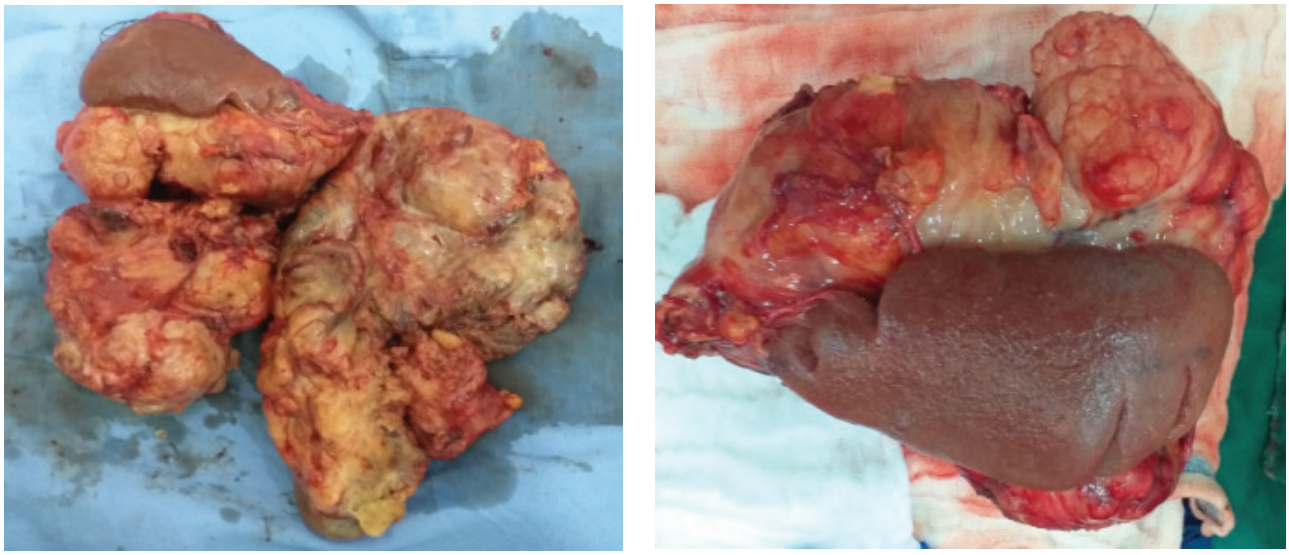
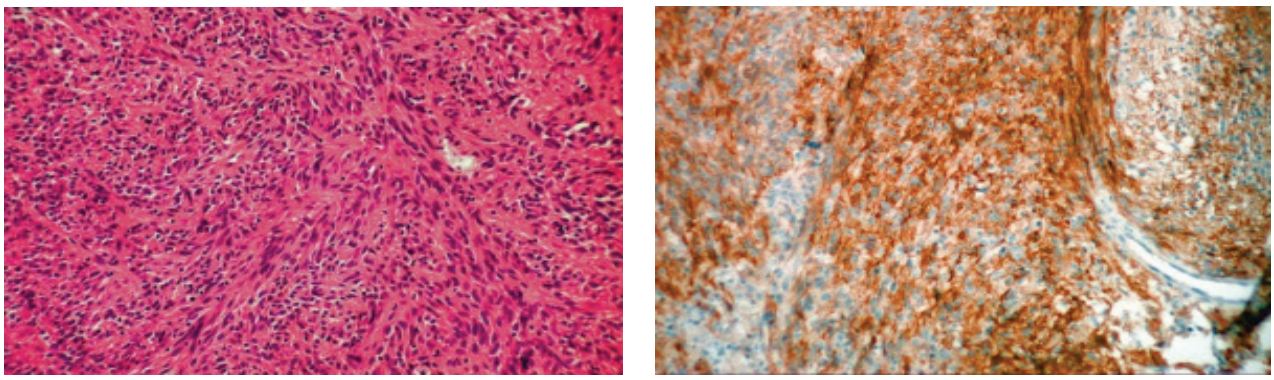


Photo 7. Histopathological description of the preparation / CD 117(c KIT). The tumor formation has cystic areas and gray-white to yellow solid areas and hemorrhagic areas. The lobar construction of the tumor is seen, with hypercellular areas with fusiform stromal cells, eosinophilic cytoplasm, with enlarged nuclei and prominent nucleoli. Areas of epithelioid stromal cells with vacuolated cytoplasm with atypical mitoses are also observed



sides, and splenectomy is performed. A portion of the tumor from the greater curvature of ventriculli is excised, along with a part of the tumour mass included in it, using a linear stapler using PDS 3/0 sutures. Finally, the abdominal cavity is rinsed with several liters of physiological solution, and two drains are placed, one in the Douglas space and the other in the subdiaphragmatic space on the left side.

Discussion

The GIST tumor in 70% of cases presents with obscure clinical signs, while in 20% of cases there are asymptomatic clinical signs. Clinical signs include pain in the abdomen, flatulence, intraluminal bleeding that appears as hematemesis, or melena and intra-abdominal bleeding in case of rupture of the tumor process.

Gastro-intestinal stromal tumor of mesenchymal origin can appear in different locations of the digestive system. It is a rare tumor and its terminology is relatively new. Previously, it was called a leiomyoma or leiomyoblastoma, which rarely appears as a retroperitoneal tumor, as is the case in the presented patient. Until now, not a large number of this formation in the retroperitoneal region have been described in the world literature, while compared to other tumors in the retroperitoneum, less than 1% appear. Namely, 7 patients per 1 million inhabitants. While, on the other hand, these formations include a total of 1-3% of cases from all other tumors of the digestive system. (12).

The tumor formation most often appears in the colon in 60% of cases, in the small intestine in 30%, the rectum in 5%, and the colon in 2% of cases and is accompanied by complications such as; hemorrhage, which indicates urgent surgical intervention, anemia, obstruction in the intestinal passage, etc. It is thought that these tumors can metastasize even 30 years after their extirpation. In the diagnosis of tumor formations in the abdomen, several modern diagnostic methods are applied, such as: CT of the abdomen, magnetic resonance, PET (Positron emission tomography) and endoscopic ultrasound examination with aspiration, as a pre-operative diagnosis (13, 14).

Percutaneous biopsy is not preferred due to the possibility of rupture of the tumor process and its dissemination in the abdominal space.

Abdominal CT and ultrasound were performed on our patient, and based on these examinations and clinical examination, the indication for open surgical intervention was determined. While laparoscopic operations are reserved for certain locations of tumors and those of smaller size.

In the conservative treatment of this pathology, the authors have used Imatinib Mesulat as adjuvant and non-adjuvant therapy, which can also be applied in the preoperative stage with the aim of reducing the tumor mass. The preparation also has an effect on the presence of tumor metastases at altitude applied in doses of 400-800 mg (15, 16). This type of treatment is based on the effect of Tyrosine Kinase inhibitors on membrane receptors C-KIT (procto-oncogene)- (17). The post-operative treatment consists in the application of the preparation Imatinib Mesulat 400-800 mg as a daily dose, 1-3 years as adjuvant therapy.

There are several methods of surgical treatment, ranging from open operative methods, laparoscopic methods and endoscopic ultrasound surgical interventions (EUS), to submucosal tumor location as a minimally invasive and combined method (18).

Conclusion

Neoplastic formations Extra Gastro-intestinal stromal tumor are rare formations, in particular those located in the retroperitoneum. While, according to statistics, these processes appear more often in the stomach (60%), duodenum and jejunum (30%), rectum 5%, while in the colon only in 2-%. of cases. The average age of appearance of these tumors is 50-60 years old.

In our material, this was the first case of this formation with a retroperitoneal location, which included most of the intraperitoneal space (2/3 of it). Tumors in 50% of cases undergo ulcerative changes and bleeding, but also obstructions in the digestive system and weight loss.

In our material, inside the tumorous mass, we encountered hemorrhagic areas, which was confirmed by puncture of the tumor. It is thought that the changes in this process appear as a consequence of gene mutation in Tyrosine-Kinase (KIT) receptors.

In the immuno-histological and histological examination we encounter the hyperplasia of Cajl cells in the muscularis propria of the intestine. However, its origin can also come from mixed cells (10%), epithelioid cells, and “spindle cell” cells in 70% of cases, as well as from the retroperitobneal connective tissue. Tumors of this nature, depending on the location, give metastases in the liver and lungs, sometimes even 30 years after the extirpation of the primary tumor.

Stromal tumors appear in three histological forms:

with finger-shaped cells (70%), epithelioid cells (20%) and in their mixed form (10%). In the histological aspect, in the differential diagnosis, leiomyoma, leiomyosarcoma, desmoid tumors, histiocytoma and malignant schwannoma should be distinguished.

Tumors of this nature appear in men in over 50% of cases, while the presented patient was female. The tumor is usually of small size, while in our material this was an exception as the tumor had enormous dimensions. In addition, tumors of this nature are usually accompanied by complications during their development, which did not happen to the patient in question. In the presented case, the operation was accompanied by splenectomy, since the process also infiltrated the splenic cell.

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