CASE REPORTS

A Rare Case of Gastrointestinal Stromal Tumor of the Abdominal Wall

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Abstract

Gastrointestinal stromal tumors (GIST) are rarely encountered in medical practice, developing with predilection in people over 40 years old. Their starting point is found in the digestive tract, and can appear anywhere along it. Normally, in the human body cells go through the processes of development, division and then apoptosis. In the case of gastro-intestinal stromal tumors, the cells divide continuously and the process of apoptosis is no longer carried out, forming these tumors, which can be benign or malignant. The essential medical treatment used in the case of gastrointestinal stromal tumors is Imatinib, which has the role of preventing the growth or even regression in size of the tumor in most cases. The specificity marker for GIST is represented by c-KIT protein, and immunohistochemically, the majority of gastrointestinal stromal tumors show positivity for CD34 and c-KIT. There are also extragastro-intestinal stromal tumors, for example located in the liver, duodenum, pancreas, but they are extremely rare, being considered rather as metastases from the primary tumor.

Keywords: gist, stromal tumors, digestive neoplasia, abdominal wall.

Rezumat

Tumorile stromale gastro-intestinale(GIST) sunt rar întâlnite în practica medicală, dezvoltandu-se cu predilecție la persoanele peste 40 ani. Punctul lor de plecare se regăsește la nivelul tractului digestiv, putând apărea oriunde de-a lungul acestuia. În mod normal, în corpul uman celulele parcurg procesele de dezvoltare, diviziune și apoi apoptoză. În cazul tumorilor stromale gastro-intestinale, celulele se divid in mod continuu și nu se mai realizează procesul de apoptoză, formându-se aceste tumori, ce pot avea caracter benign sau malign. Tratamentul medical esențial folosit în cazul tumorilor stromale gastro-intestinale este Imatinibul-ul, ce deține rolul de a împiedica creșterea sau chiar regresia în dimensiuni a tumorii în majoritatea cazurilor. Markerul de specificitate pentu GIST este reprezentat de proteina c-KIT, iar imunohistochimic, marea parte a tumorilor stromale gastro-intestinale prezintă pozitivitate pentru CD34 și c-KIT. Există si tumori stromale extragastro-intestinale, de exemplu cu localizare în ficat, duoden, pancreas, însă sunt extrem de rare, ele fiind considerate mai degrabă metastaze date de tumora primară.

Cuvinte cheie: gist, tumori stromale, neoplazii digestive, perete abdominal.

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INTRODUCTION

Gastrointestinal stromal tumors have a low incidence in people under 40 years of age, they can appear spontaneously or in the context of family history. The predominant location is not a specific one, due to the fact that they can develop anywhere along the digestive tract.[1] At the same time, the sizes of gastrointestinal stromal tumors can vary greatly, from small tumors to giant tumors. The latter worsen the symptoms, especially due to the compression on the other neighboring organs. Normally in the human body, cells grow, divide and then the process of apoptosis takes place. In the case of gastrointestinal stromal tumors, cells divide continuously, and the process of apoptosis is absent, an aspect that leads to the appearance of these types of tumors, which can be both benign and malignant. In patients with small tumors, as a rule, no specific symptoms are found, but in those with large tumors, the clinic may involve abdominal pain, fatigue, loss of appetite, leading from the impairment of intestinal transit to the installation of intestinal occlusion, due to the compression achieved of tumor on the organs. Gastrointestinal stromal tumors have as their starting point the organs of the digestive tube.

CASE PRESENTATION

In the Surgery Clinic of the Dr Ion Cantacuzino Clinical Hospital, a 31-year-old patient presents herself for diffuse abdominal pain, continuous spontaneously and on palpation, alteration of intestinal transit, with a history of transverse colon GIST, for which she was operated on 4 years en bloc subtotal colectomy with partial resection of greater gastric curvature.The confirmation was brought by the histopathological examination, which supported the diagnosis of GIST, the patient following postoperative treatment with Imatinib. The clinical examination reveals a voluminous formation on the abdominal wall, with dimensions of approximately 210/108cm.

As an observation, we can note the fact that this formation presents a development at a very fast pace.

The laboratory analyzes described us leukopenia (2.33*1000/Ul) and a slight anemia (hemoglobin 11.1 g/dl), the rest of the laboratory analyzes being within normal limits.

The CT examination of the chest-abdomen and pelvis with contrast material describes a tumor formation with maximum axial diameters of approximately 190/115mm and cranio-caudate of 221 mm, well demarcated with inhomogeneous iodophilia, which shows invasion in the rectus abdominis muscles on the right side and comes in contact with loops of small intestine and large intestine, against which it does not present a cleavage plane. Superiorly, the tumor formation extends to the level of the gallbladder and the visceral face of the left liver lobe, and inferiorly it reaches the vicinity of the right antero-superior slope of the segmental urinary bladder without a cleavage plane in relation to it, with the related vasculature from the

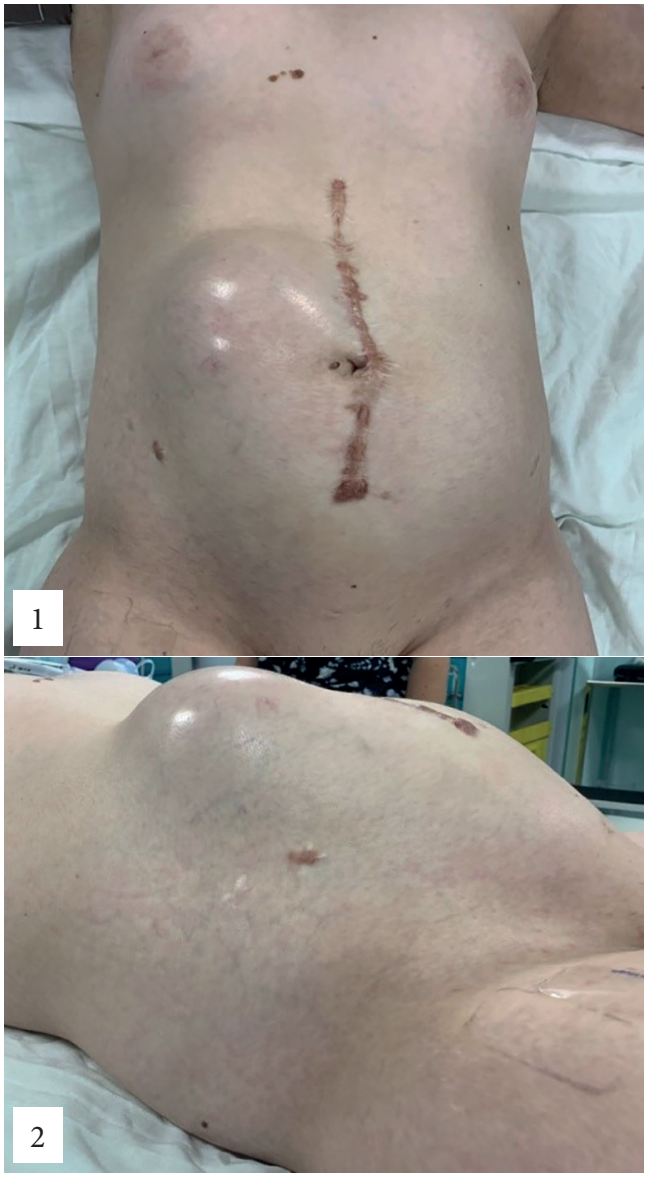

Figure 1,2. Preoperative images

inferior epigastric artery and several bladder branches predominantly on the right side. No abdominal-pelvic adenomegaly with pathological significance, no ascites. The patient's case was submitted to the oncology commission, which concluded the need for surgical intervention, both for the purpose of abdominal decompression and as a curative treatment.

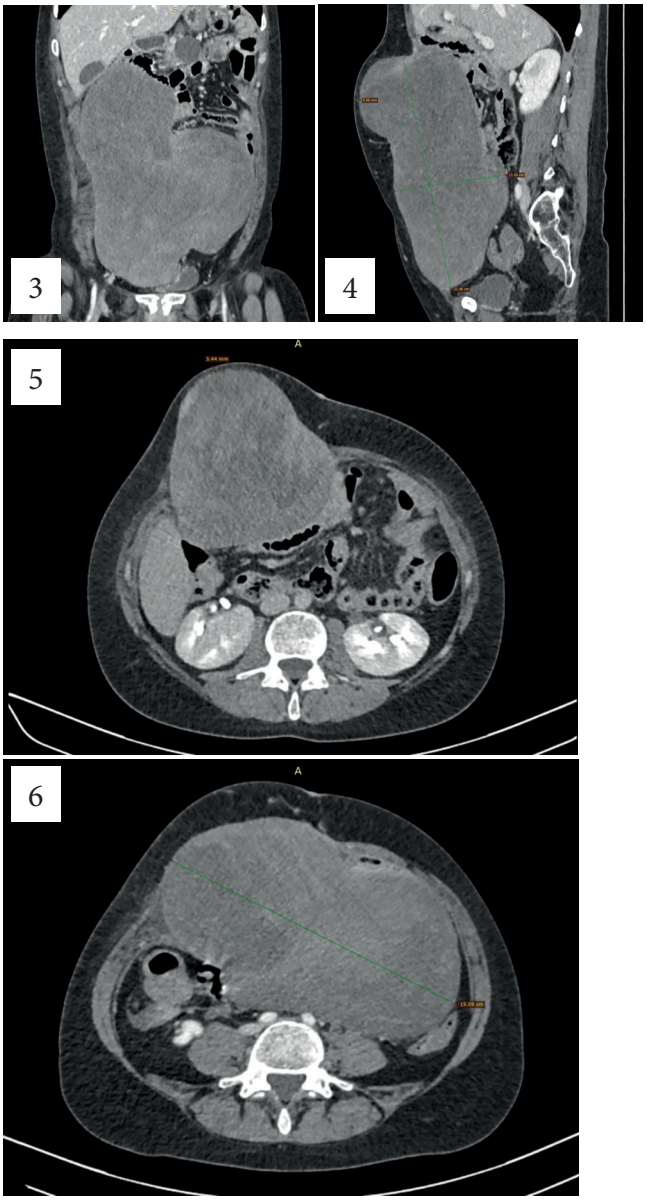

Figure 3,4,5,6. Computer Tomograph images

During the surgical intervention, the skin-mucosal flaps were removed bilaterally and at a distance from the edges of the tumor formation and the peritoneal cavity was penetrated through a breach located at the level of the left hypochondrium. The tumor formation was excised in one block, without any damage to the abdominal, vascular or bladder organs.

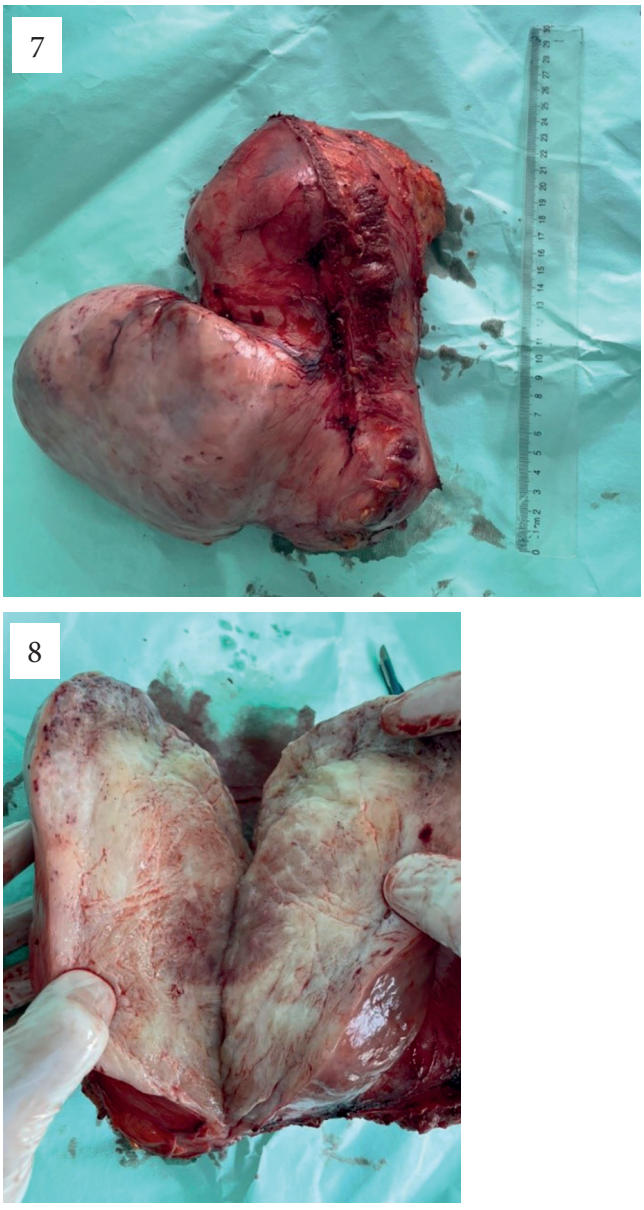

Figure 7,8. Tumor images

The excision of the tumor formation, which weighed 3.8 kg, no longer allowed the closure of the abdominal wall in anatomical planes, which is why the closure of the parietal defect was achieved by applying a Dual-mesh 30x30 cm abdominal wall substitution table, sutured with continuous thread.

The postoperative evolution was favorable, with the gradual remission of the pain syndrome and the re-

sumption of intestinal transit, the patient was discharged 14 days postoperatively.

Histopathological aspects support our diagnosis of gastrointestinal stromal tumor (GIST).

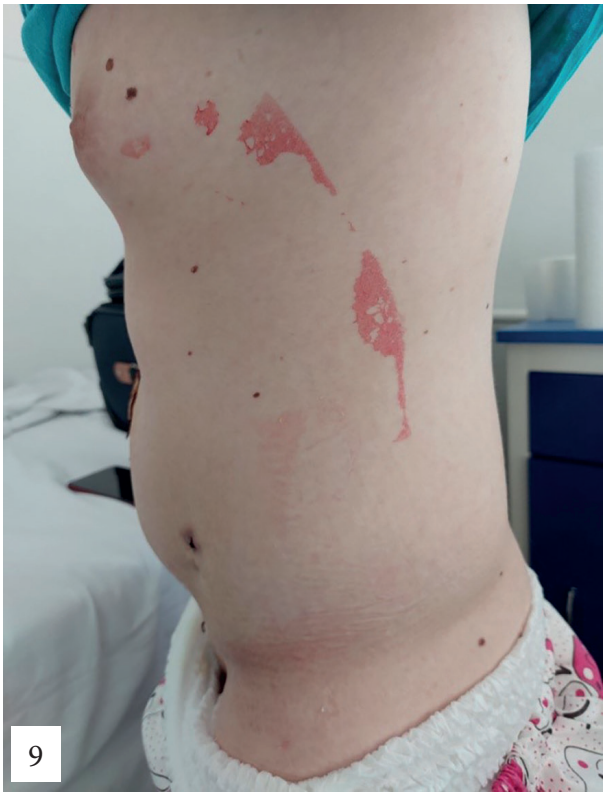

Figure 9. Postoperative imege

DISCUSSIONS

The medical treatment most used in the case of gastrointestinal stromal tumors is Imatinib, which has the role of preventing the growth or can even lead to regression in the size of the tumor in most cases. But in the case of our patient, despite the administration of Imatinib, the tumor continued to grow, which indicates that some patients may not respond or even develop resistance to treatment.

There are also extragastro-intestinal stromal tumors, for example located in the liver, duodenum, pancreas, but they are extremely rare, being considered rather as metastases from the primary tumor.

Rarely, GIST-type tumors have been reported that can develop in the adjacent mesentery, omentum, retroperitoneum and, extremely rarely, in the lungs and female genital organs, these being called extra-gastrointestinal GIST (EGIST)[5,6]. GISTs discovered at the level of parenchymal organs - liver, pancreas, prostate - are considered metastatic by direct extension from gastric and duodenal or rectal tumors.

The tyrosine-kinase and KIT inhibitor is represented by Imatinib, the latter being considered the most selective and not, last but not least, the preoperative and postoperative adjuvant treatment in GIST cases.

In cases of gastrointestinal tumors (GIST) refractory to Imatinib treatment, data from the literature have described Nilotinib as an alternative, used together with Imatinib or as monotherapy.

The cause of resistance to Imatibib treatment is not fully explained, but it is thought to be due to the emergence of new mutations, the acquisition of resistance during treatment or even toxicity.

CONCLUSIONS

Gastrointestinal stromal tumors can also be malignant, sometimes the symptoms have a major impact on the quality of life, and sometimes the symptoms are absent.

The vast majority of gastrointestinal stromal tumors occur in the stomach, followed by those in the small intestine, but in particular they can also occur in the peritoneum, of course in a much smaller percentage.

The specificity marker for GIST is represented by the c-KIT protein, and immunohistochemically, the majority of gastrointestinal stromal tumors show positivity for CD34 and c-KIT. The CD34 cell has the role of coordinating intestinal motility, being called a pacemaker cell.

The standard treatment in GIST cases remains complete surgical resection alongside treatment with Imatibib and/or Nilotinib in refractory cases, with surgical cytoreduction having a remission rate of approximately 50% of cases.

Radiotherapy and chemotherapy in GIST cases is not a method of choice, as it has not brought therapeutic benefit in the vast majority of cases.

If chemotherapy is still preferred, a preliminary preparation of the body is indicated, so-called nutritional therapy, which will help the body to cope with the oncological treatment, which is usually quite aggressive.[7]

Statistical data support a 5-year survival rate of approximately 90% in cases of localized GIST, and in disseminated and distant GIST a rate of approximately 50%.

Compliance with ethics requirements: For the publication of this article the authors declare that there is no conflict of interest and the patient's informed consent was obtained for the publication of images and personal data, preserving the patient's anonymity.

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