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Small bowel stromal tumor a case report

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Abstract
Gastrointestinal stromal tumors (GIST) are the most common digestive mesenchymal tumors. Characterized by over-expression of the tyrosine kinase receptor KIT. GISTS span a wide clinical spectrum from benign to highly malignant. Surgery is the only curative treatment for GIST. The development of tyrosine kinase inhibitors has changed the management of unresectable GIST. One of them, imatinib mesylate, has been proved to improve survival of metastatic GIST. We report a case of stromal tumor of small bowel localization brought in the surgery service of the 5th Military Hospital Guelmim in Morocco.

1. Introduction

Stromal digestive tumors are the most frequent common mesenchymal tumors of the digestive tract. These tumors were the object of numerous controversies in terms of histogenesis and classification. They become integrated now into a frame precise nosologic since the discovery of the expression by the tumoral cells of the protein c-kit.

2. Observation

45-year-old patient, without histories, admitted in the service for small rectorragies associated to an abdominal mass sitting at the level of the left side.

The beginning of the symptom went back to 1 month by the appearance of mœlena then rectorragies of low abundance, the whole evolving in a context of preservation of the general state.

The somatic exam in the admission was without peculiarities. The rectal touch did not show anomaly. The oeso gastro duodenal fiberscopy showed no anomaly.

The colonoscopy did not show anomaly at the level of all the colonic frame.

The abdominal scanner showed a tissular formation in depends of hail 10 cms in diameter (figure1).

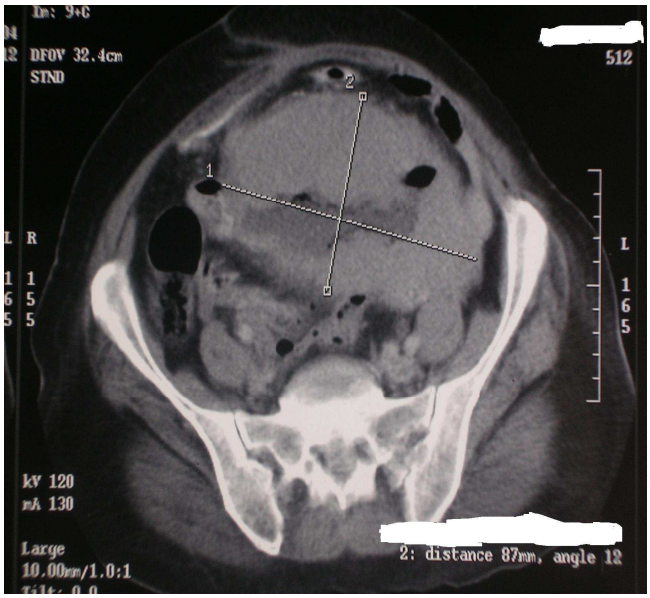

Figure 1. *Abdominal scan showing a tumor developed at the expense of small bowel*

He is operated by a median laparotomy, the exploration discovered a jejunal tumor in 1m 80 of the treitz angle(figure2).

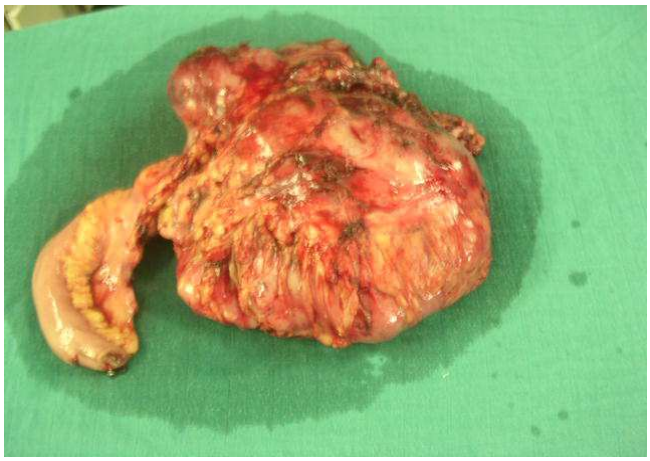

Figure 2. *Operating piece showing a small bowel location of stromal tumor*

We realized a jejunal resection taking the tumor with anastomosis jejuno jejunal . The post-operative suites were simple. The anatomopathologiqc and immunohistochimical study showed cells expressing intensely and vaguely the CD117 (C-Kit) and the CD34, and more irregularly the protein S100. This profile immunophenotypic conjugated, in the morphological aspect, corresponded to a malignant potential intestinal stromal tumor

The clinical evolution was good with a backward movement of 12 months without any sign of second recurrence.

3. Discussion

Stromal digestive Tumors are rare, which could develop along the digestive tract or sometimes from the epiploon and the mesentery.

These tumors were the object of multiple controversies, they were confused for a long time with the other conjunctival tumors, worth namely mainly schwanomes and leiomysarcomas. But thanks to the progress of the immunohistochimy, they establish an entity recent and precise nosologic, grouping all the digestive conjunctival tumors which express the protein C-Kit, and the potential of wickedness of which is often difficult to estimate.

Stromal tumors are particularly on the agenda, since the discovery of a treatment of the malignant forms by an inhibitive molecule of tyrosine kinase (STI571 or GLIVEC)[1].

Stromal digestive Tumors or GIST (Gastroenteritis Intestinal Stromal Tumor) are not epithelial tumors developed at the expense of the connective tissue of the wall of the organs of the digestive tract. Recently, several studies revealed of immunohistochimical and ultrastructural similarities between the cells which compose stromal digestive tumors and the interstitial cells of Cajal [2]. The cells of Cajal, which constitute the autonomous nervous system of the digestive tract are located in the normal state in a diffuse way between and in the circular and longitudinal coats of muscular digestive. These cells play an important role in the motricity of the digestive tract said activity of pacemaker. The tumoral cell are characterized by the expression of the marker CD34, common to the cells of Cajal, and by the expression of the receiver tyrosine kinase tc kit, (CD117) under a moved and/or activated shape. These transformations are of arisen premature and constitute even maybe the event initial of the disease.

The incidence is estimated unless 5000 cases / year in the United States and in 1000 cases / year in France [3] . Represent approximately 10 % of sarcomas, less than 1 % of the malignant tumors of the digestive tract and 20 % of the malignant tumors of the small intestine, if we exclude lymphomas [1, 2].

The leiomyosarcoma represents 10 % of the malignant tumors of the small intestine [3].

Schwannosarcoma is rarer, representing only 0,2 to 0,6 % of the malignant tumors of grele [1,4].

The age of arisen is generally situated between 45 and 60 years [2]. The male genital organ is the most affected in the literature, as it was the case in our series.

Leiomyosarcoma is frequently localized in the ileum and constitute the most frequent tumor of the diverticule of Meckel [3]. Schwannosarcoma sits in order of frequency decreasing in the jejuno-ileon, the duodenum and, finally, the stomach and the rectum. The clinical diagnosis is often late, what explains that most of these tumors are discovered

at a late stage by abdominal mass. The digestive bleeding is the clinical sign most frequently found [2, 4]. The discovery of an abdominal mass is found in 40 % of the cases [1,3].

In a macroscopic manner, the GIST is called encapsulate tumors , even in case of malignant GIST, often containing foyers of bleeding and necrosis . They are often associated with ulcerations of the mucous membrane recovering them explaining their mode of presentation under the shape of digestive bleedings. Microscopically, it is about uniform proliferations of mesenchymental cell which have the peculiarity to be generally strongly positive immunohistochimy for the c-kit (CD 117). For their histological diagnosis and immunohistochimic, other markers are used and necessary such as the CD34.

Stromales tumors of hail determine most of the time to the echography a voluminous tissular mass of polylobed outlines, homogeneous or heterogeneous [2,5].

The ultrasound aspect of the mass is variable according to the existence or not of a communication with the digestive light. It allows to make an assessment of extension local and regional in search of metastasis and to guide the biopsies to confirm the diagnosis.

The transit of grele is the fundamental exam in the diagnosis of the tumors of grele. However, false negatives were reported in the literature because of the development under serous of the tumor [5].The sensibility of the examination is improved by the double contrast. The radiological aspect varies according to the tumoral setting-up in the wall and to the degree of invasion of the neighboring tunics from the coat of origin [2,5]. It can show an incomplete image, an ulceration or an extrinsic imprint.

The abdominal scan can show a voluminous tumor with exoluminal preferential development, of polylobed outlines, heterogeneous density with zones of necrosis [5]. The scanning also allows assessing extension local and regional.

The main differential diagnosis of a conjunctival malignant tumor of hail is the lymphoma, which is translated by the existence of a cavity image from which the characteristics postpone conjunctival tumors [1,4].

The treatment is essentially surgical, the ganglionic cleaning out is not systematic, because the ganglionic metastasiss are rare (less than 10 %) and the risk of ganglionic second recurrence is limited (less than 5 %). The forecast depends essentially on the mitotic index and on the tumoral size[3,5].

The imatinib (in the form of mesilate, formerly STI 571) is a selective inhibitor of proteins tyrosine kinases, in particular of c-kit and the receiver of the PDGF, at the level of their site of fixation of ATP [2,5]. This led to use it first of all in the treatment of the myeloid chronic leukemia with high rates of answers.

The justification of the use of the imatinib in stromal digestive tumors is connected to its inhibitive action of the protein c-kit activated independently of its ligand.[5,6].

The imatinib is administered by oral way (96 % bioavailability) in a daily taking because of a long half-life (18 hours). Capsules are measured in 100 mg. Its metabolism is hepatic and its essentially biliary elimination.

The Imatinib mesylate is a well-tolerated drug, and all undesirable effects could be ameliorated easily. The most frequent adverse effects related to the drug were nausea (78%), diarrhea (70%), dermatitis (62%), facial edema (61%), edema of the lower limbs (58%), vomiting (54%), and eyelid edema (51%). The most common hematological and non-hematological side effects were anemia and fluid retention, respectively

Since August, 2000, the use of the imatinib revolutionizes the care of stromal tumors to there, particularly chimio resistances and whose only potentially effective treatment was the surgery. The low rates of resecability of the second recurrences the medians of survival of which, even in complete resection, are 18-month, and the fact that the purely local second recurrences concern only the third of the patients, direct to a treatment by Glivec ® in first intention in these situations [2,4,6].

The radio chemotherapy is generally recommended in the palliative situations, but did not make the proof of its efficiency.

For the not metastatic localized tumors, the treatment of choice it is the surgical resection.

At the patients presenting a locally evolved tumor not resecable , it is necessary to establish a treatment by Glivec ® and to discuss a secondary surgical gesture as soon as the maximal answer is obtained. For tumors with peritoneal extension and/or hepatic, and considering the secondary risk of resistances and the rarity of the complete answers to the imatinib, we discuss secondarily, at the time of the maximal answer, the exerese of the remainders peritoneum and hepatic tumors.

4. Conclusion

Small bowel stromal tumors are rare. Their symptom is can specific, dominated by the digestive bleeding and the abdominal pain.

The expression of the C-KIT, a receiver membrane in tyrosine-kinase activity is considered as specific characteristic. The appeal to the immuno histochimy is necessary to obtain a definitive diagnosis. The scanning remains the examination the most used in our context, as well in a diagnostic purpose, that for the follow-up therapeutic comment.

The surgical resection is the treatment of choice of the localized tumors. The imatinib, as therapeutic novelty, is an example of therapy targeted for not resecables and metastatic tumors. The survival is connected to the complete character of the surgery. The prolonged surveillance is necessary because of the risk of the second recurrence.

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