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Case Report: A Rare Case of Tumor: A Metastatic GIST

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Abstract

Gastrointestinal Stromal Tumors "GIST" is a rare form of digestive tract cancer with an estimated incidence of 15 cases per million population with an average age of 60 years. The surgical methods remain the main form of treatment and the only curative one. This case study is about a 5-year-old patient who presented non-specific abdominal pain and impaired general state. The CT-scan showed a voluminous abdominal mass located in the stomach as well as two liver metastases. We decided to perform a surgical biopsy of the gastric mass in order to identify its type. The anatomopathological analysis concluded in a Gastrointestinal Stromal Tumor, probably of gastric origin. In this context, we decided to initiate an Imatinib treatment: 400mg a day per os in one take. The patient initially responded very well to the treatment with a decrease of the size of the gastric tumor but, after 4 years, the mass started to further increase in volume.

After multidisciplinary discussion and involvement of the patient, surgical resection was agreed upon. No lymph node resection was done since lymph node metastases are rare. We performed a total gastrectomy with complete monobloc surgical resection of the tumor as well as a metastasectomy of the two liver lesions, an ablation of lesion on the ligamentum teres hepatis and hepatic hilum and finally a preventive cholecystectomy

Keywords: GastroIntestinal Stromal Tumors (GIST); Imatinib, Surgery

Abbrevations: GIST: Gastro-Intestinal Stromal Tumor

Introduction

Although gastrointestinal stromal tumors (GIST) are the most frequent sarcomas, they have been very poorly studied up until twenty years ago [1].

These tumors are the most common mesenchymal tumors of the gastrointestinal tract but still represent less than 1% of all gastrointestinal tumors [2]. GIST is a group of mesenchymal tumors that derive from interstitial cells of Cajal and which develop from the Muscularis propria. They may be acquired or genetic [3].

Imatinib has revolutionized the management of locally advanced and metastatic GIST. However, the surgical methods remain the main form of treatment and the only curative one [4]. This case report presents a metastatic gastric gastrointestinal stromal tumor recently removed in a 65-year-old male patient, the outcome and a literature review of the pathological identification, sites of origin, prognosis and treatment.

Case

A 65-year-old male patient with no significant medical history other than type 2 diabetes consulting for non-specific abdominal pain associated with a 2kg weight loss and an impaired general state, which wasn't investigated. Family history was noncontributory.

The abdomen was soft and painless during physical examination.

A contrast-enhanced thoraco-andominal CT-scan showed a voluminous abdominal mass measuring 25 x 20cm located in the stomach as well as two liver lesions. No pulmonary lesions were found. After a multidisciplinary discussion, we decided to proceed to a surgical biopsy of the gastric mass in order to identify the etiology. The anatomopathological analysis revealed a Gastrointestinal Stromal Tumor, probably of gastric origin. We decided to start a treatment with Imatinib (Glivec) 400mg [5].

The medical treatment with Imatinib worked well at first but a further increase in volume of the mass, but not the metastases, was discovered after four years. After a new multidisciplinary discussion, given the resectability of the lesion, it was decided to perform a surgical "en bloc" resection of the tumor [6].

Discussion

Until 1998, gastrointestinal stromal tumors remained largely unknown. In the beginning of the 80's, the generalization of immunohistochemistry techniques allowed us to take enabled a big step in the diagnosis, but also in the treatment of these tumors since they are characterized by a very frequent expression of KIT (CD117), whose expression is specific to the interstitial cells of Cajal. GISTs are slightly more prevalent in male patients, with an average diagnosis age of 60 years [7,8].

Our patient suffered from abdominal discomfort and abdominal bloating. After further questioning, he confirmed a feeling of early satiety. He denied any other symptoms.

If surgery is the standard procedure for all GIST of small intestine or rectum it is not the case for gastric GIST. The treatment strategy in these cases is affected by several factors: the size of the tumor, its location, its adhesions to nearby structures and presence or absence of metastasis [1,9] (Figure 1-7).

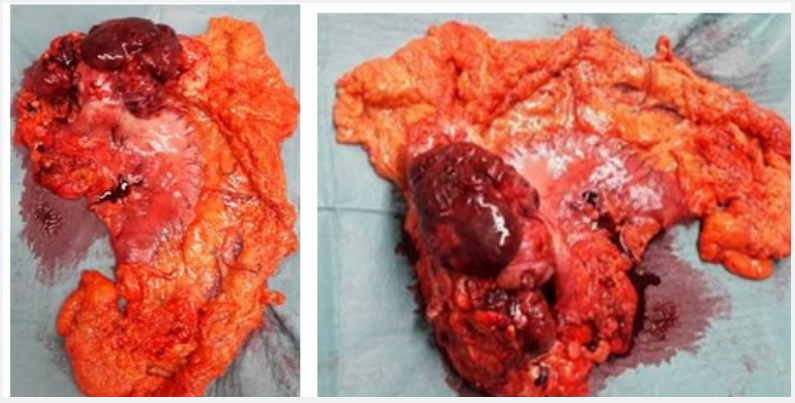

Figure 1: Macroscopic: Gastric GIST.

Figure 2: Voluminous heterogeneous mass exceeding 25cm of diameter.

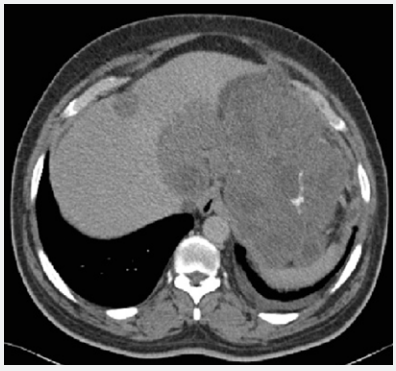

Figure 3: Hepatic lesion that correspond to a metastasis.

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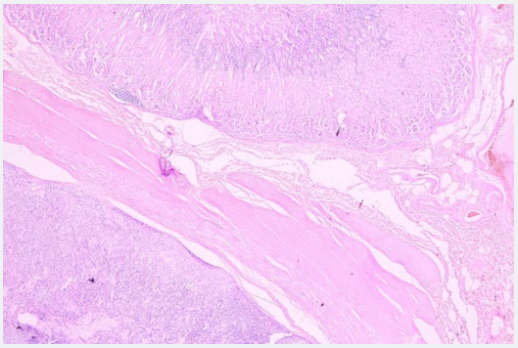

Figure 4: Localization in the depth of the gastric wall at the level of the muscularis propria.

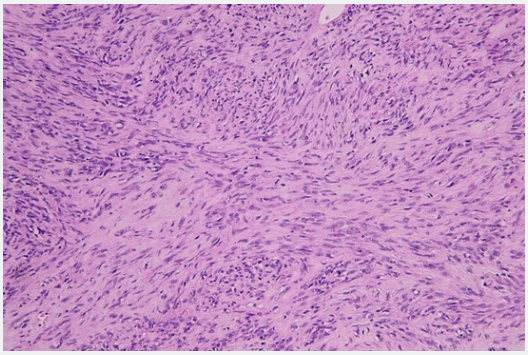

Figure 5: Fascicular structure with elongated fusiform cells. This is a feature of GISTs. Presence of some cellular atypia.

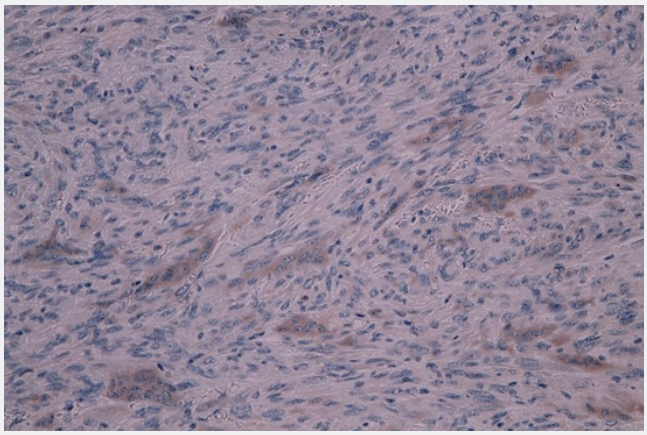

Figure 6: Immunolabelling (this analysis is essential to confirm the diagnostic of GIST): CKIT -CD117: positive with presence of brownish

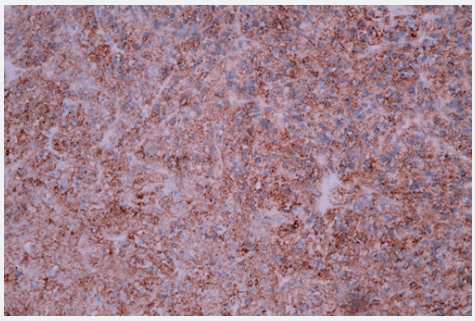

Figure 7: Immunolabeling of DOG 1: The expression of the protein “DOG1” is more specific than the expression “KIT” which can be observed in many other tumors.

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Some studies have suggested the high frequency of small gastric GIST (<10mm in diameter) in adults after 50 years of age. The evolution of those tumors is not certain, and they could even regress in the future. Other studies seem to show that the risk of malignancy in GIST of the stomach is very low or inexistent when their diameter is smaller than 2cm [10-12].

Thus, for gastric GISTs of less than 2cm, the choice between clinical surveillance and surgery will be based on symptoms (minor at this size), patients' general condition and location of the lesion in the stomach, facilitating resection or not. Endoscopic excision may be a treatment option for gastric GISTs of this size. Some Asian teams use this technique, which remains less invasive. It can also be combined with laparoscopy, but its efficiency remain uncertain. For non-metastatic gastric GIST greater than 2cm, complete surgical excision is the standard procedure with or without prior Imatinib treatment [13,14].

Our case is different from all these situations since it is a metastatic gastric GIST. The studies show that in responding or stable patients using Imatinib and potentially accessible for an R0 resection, the role of surgery remains to be clarified. Its feasibility has been shown, but its impact on survival hasn't yet been established. Two randomized trials (continuation of Imatinib versus surgery and continuation of Imatinib) were suspended due to slow recruitment. The Chinese trial, which included only 41 patients, did however show a non-significant trend in favor of the "surgery" group [15].

Our institution believes surgery remains the best solution and the only curative one if R0 is possible, even in metastatic cases [13]. We therefore proceeded to a total gastrectomy with complete monobloc surgical resection of the tumor as well as a metastasectomy of the two liver lesions, a removal of the ligamentum teres hepatis and hepatic hilum metastases and, finally, a preventive cholecystectomy. No lymphadenectomy was performed since lymph node metastases are rare, even in advances cases, and all the forty-eight lymph nodes associated to the surgical piece showed no anatomopathological signs of malignancy. At six months postoperatively, an abdominal contrast-enhanced CT scan showed no sign of resurgence.

Conclusion

Even though this is an isolated case, it might be worth thinking about the management of stable metastatic GIST under Imatinib. A new randomized study could, of course, allow a big step in the management of these tumors. Unfortunately, recruitment is a main limitation since these cases are rare.

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