

A GIST or not a GIST? That is the question

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Several reports have recently focused on stromal tumors and their acute or chronic complications [1,2]. Gastro-Intestinal Stromal Tumors (GIST) are mesenchymal neoplasms that represent a small percentage of gastric cancers (1-3%) and arise more often from the stomach (60%) [3]. Expression of c-KIT (CD 117) in the tumor cells define their character in a proportion of 95% with the remaining 5% representing c-KIT-negative GISTs associated with platelet-derived growth factor receptor-amutations [4-6]. Imatinib, an inhibitor of the oncoprotein BCR-ABL is used as an adjuvant therapy in metastatic c-KIT metastatic GISTs and recently in primary resected GISTs and has changed their prognosis.

Desmoid tumors are well differentiated, locally aggressive fibrous neoplasms that occur in patients with familial adenomatous polyposis at an approximately rate of 10%. Histologically their character is benign but could lead to obstruction of vital structures and organs. They tend to recur and their therapeutic management is often very difficult.

An 18-year-old male with free medical history was admitted in our department due to acute abdominal distension. Standard blood tests were normal. Abdominal ultrasound revealed an epigastric mass of 120.8x107.3 mm that repelled spleen, without a central fusion and cystic components with rich neo-vascularization. CT could not establish the origin organ of the lesion. MRI showed that the origin of the mass was from the layers of the stomach (Fig. 1). Upper GI endoscopy and colonoscopy were normal. Endoscopic ultrasound (EUS) revealed in the major arc of the stomach a sizeable hypoechogenic and in homogeneous arrangement

of 122.2x108 mm in contact with the fourth sonographic layer (muscularis propria). Due to the size and pressure of the mass EUS could not determine that it originated from the stomach. The patient underwent a surgical resection of a 25x20 cm mass of 4 kg (Fig. 2). The macroscopic diagnosis was as of a GIST. However, the histology revealed a tumor inherent in the stomach with few mitoses, 1-3/50 mitoses per field, asteroid or elongated cells with fusiform nuclei with histological and immunohistochemical characters compatible with intraabdominal fibromatosis. Tumor cells penetrated the main focal muscular tunic but the supernatant submucosa and mucosa of the stomach were intact. The surgical margins as well as the omentum, mesentery lymph nodes and three omentum lymph nodes were free of tumor infiltration. Immunohistochemistry showed positivity for Vimentin. S100 protein was positive in scattered cells and β -catenin positive in isolated plasmatic and nuclear staining of cell damage. SMAActin, MSActin and desmin were negative. Finally CD117 and CD34 were also negative. Because of the size of the mass EUS could not establish the origin of the tumor. On the other hand, histology revealed a giant intra-abdominal fibromatosis based on positivity of β -catenin and negativity of CD117, CD34 and although the mass was inherent with muscularis propria of the stomach the origin of the tumor could not be established.

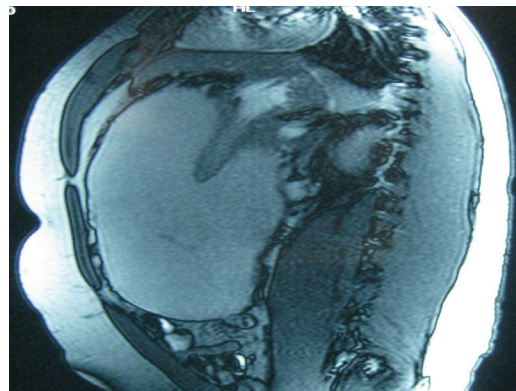


Figure 1 MRI showing the limits between the stomach and mass

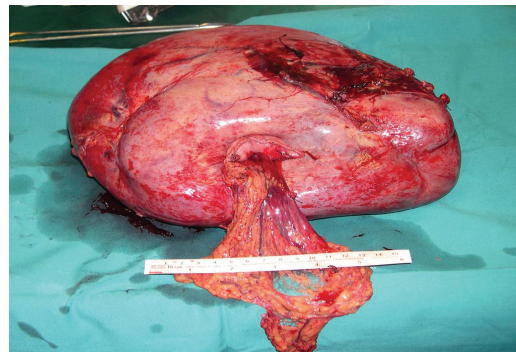


Figure 2 Postsurgical image of the abdominal mass

In vivo results such as paraclinical tests or intraoperative findings do not always correlate with *in vitro* results such as histological findings as in our case, which makes the paraphrase of Hamlet's question opportune.

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