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Case Report

Duodenal Leiomyosarcoma Presenting with Gastrointestinal Bleeding: A Case Report

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Abstract

Duodenal leiomyosarcomas are exceptional. Their preoperative diagnosis is difficult. The association of 2 "mass-haemorrhage" symptoms should suggest the presence of leiomyosarcoma. The means of radiological, endoscopic and above all anatomopathological study make it possible to make a positive diagnosis.

We report the case of a 52-year-old man who presented with gastrointestinal bleeding and obstruction who was diagnosed with duodenal leiomyosarcoma after surgical resection.

Keywords: Leiomyosarcoma; Duodenum; Gastrointestinal Bleeding; Immunohistochemistry

Introduction

Since the discovery of the particular phenotype of gastrointestinal stromal tumors with the frequent expression of the CD34 receptor and the almost constant expression of the c-Kit protein, these tumors are truly individualized and currently clearly distinguished from other digestive mesenchymal tumors such as leiomyomas, leiomyosarcomas, neurofibromas or schwannomas. Gastric leiomyosarcomas are tumors developed at the expense of the smooth muscle fibers of the gastric wall, currently becoming extremely rare with the development of the concept of gastrointestinal stromal tumors. According to the literature, they represent only 1-3% of all malignant tumors of the stomach [1,2]. They usually affect adults in their fifties without any gender predominance.

Medical Observation

We report the case of a 52-year-old man with no particular history admitted for upper digestive hemorrhage of a type of hematemesis of low abundance accompanied by intermittent vomiting and unquantified weight loss. The clinical examination found a hemodynamically stable patient with slight tenderness of the left hypochondrium without a palpable mass, of. There was no hepatomegaly and the lymph node areas were free. Complete blood count showed microcytic hypochromic anemia at 10 g/100ml.

Esogastroduodenal fibroscopy with biopsies did not show any remarkable abnormalities

we completed with an abdominal scan which showed a voluminous mass of 20 cm long axis of tissue appearance adhering to the spleen and the greater omentum and driving back the posterior face of the gastric antrum without invading it. It enhances heterogeneously after injection of the contrast product, revealing some areas of central necrosis. An exploratory laparotomy was thus indicated making it possible to perform, after release of the adhesions, a one-piece resection removing the mass. Exploration of the abdominal cavity had found no liver lesions or peritoneal carcinomatosis. The postoperative course was simple. Histological examination of the surgical specimen revealed a tumor measuring 20 x 12 x 12 cm.

At the microscopic study, the samples taken from the tumor correspond to a sarcomatous proliferation. It is made up of long bundles intersecting at right angles, made up of fusiform cells, with moderately abundant eosinophilic cytoplasm, with elongated nuclei with moderately anisokaryotic rounded ends, sometimes pleomorphic, showing atypical mitoses estimated at 15 mitoses/10 fields at high magnification. This proliferation is 10% necrotic and contains some cystic areas with hyalinized fibrosis.

The sarcomatous proliferation comes into contact with the duodenal submucosa without infiltrating it. Elsewhere, the duodenal intestinal mucosa is unremarkable.

An immunohistochemical study was carried out which shows the positivity of the tumor cells for the following antibodies: Anti AML; Anti Desmin; and Anti Hcaldesmon

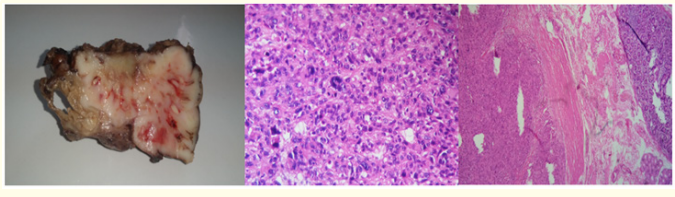

Figure a

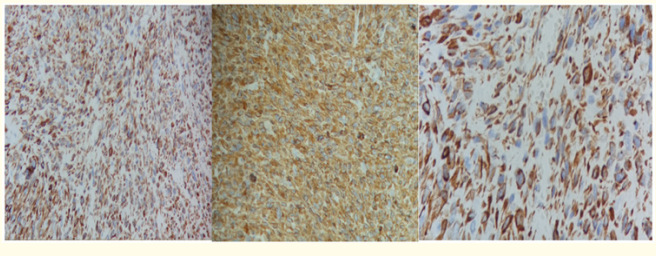

Figure b

Discussion

Leiomyosarcoma of the duodenum was first reported by von Salis in 1920, and since then there have been a number of reviews and reports of isolated cases in the literature.

Abdominal pain was the most common symptom. Gastrointestinal bleeding was also frequent. These ranged from massive hemorrhage requiring emergency surgical control to slow bleeding with associated anemia, weakness, and pallor. Less common symptoms were vomiting, diarrhea [3-5].

Apart from the discovery of an abdominal mass, the physical examination is most often normal. Gastric endoscopy contributes little to the diagnosis because these submucosal lesions are often predominantly exophytic. Fibroscopy may show a normal aspect or simply an endoluminal curvature caused by an extrinsic expansive process. Perendoscopic biopsies are generally superficial and negative [6]. Echo-endoscopy is the reference examination which confirms the intraparietal nature of the lesion or, on the contrary, its extrinsic origin, with a sensitivity of around 97% [7]. It also makes it possible to study the relationship of the tumor with neighboring organs, to judge the degree of invasion and to specify the existence, although rare, of loco-regional lymph nodes [8].

Abdominal CT without then with injection of contrast product and ingestion of water or Gastrografin on the examination table is an interesting tool for analyzing these tumors, especially lesions larger than 10 cm, and for looking for metastases, especially hepatic [7,9]. The images without injection make it possible to visualize the intra-tumoral hemorrhages and the rare endo-lesional calcifications. In the arterial phase, these tumors enhance heterogeneously and transiently due to their hypervascularization with the appearance of central hypodense images of tumor necrosis. Gastric leiomyosarcomas with exoluminal development push back intra and/or retro peritoneal organs and can lead to confusion with lesions of neighboring organs.

The diagnosis of a gastric leiomyosarcoma is difficult preoperatively, it is generally made on the pathological examination of a surgical resection piece. Histologically, leiomyosarcomas are made up of spindle-shaped cells with eosinophilic cytoplasm, coiled central nuclei and remarkably high mitotic activity. However, they remain largely confused with gastrointestinal stromal tumours, at least on endoscopy and imaging. The diagnosis of certainty is made thanks to the immunohistochemical study which shows a diffuse expression of the muscular markers mainly smooth muscle actin associated with the absence of the expression of CD117 and CD34 [6,9]. This histological profile, which corresponds perfectly to our observation, may pose the problem of differential diagnosis with other spindle cell mesenchymal tumors such as leiomyoma, leiomyoblastoma and schwannomas. Complete surgical excision of the tumor with healthy margins remains the standard treatment for gastric leiomoysarcomas. Atypical gastrectomy (wedge resection) with minimum safety margins of 2 cm may be sufficient, but subtotal gastrectomy is necessary in the case of a large tumor

[10]. Sometimes a resection of a neighboring organ is necessary to ensure complete excision of the tumor tissue. Moreover, lymph node dissection is not systematic because it does not significantly influence patient survival [7].

Conclusion

Duodenal leiomyosarcoma is a rare and its diagnosis is difficult. Clinical, endoscopic, scannographic data are not sufficient to confirm the diagnosis . the advances in immunohistochemistry and molecular biology which make it possible to establish the diagnosis.

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