UPPER GASTROINTESTINAL HEMORRHAGE DUE TO DUODENAL STROMAL TUMOR

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ABSTRACT – ***Background*** – Gastrointestinal stromal tumor represents a rare neoplasm that originates in the muscular wall of the hollow viscera. ***Aim*** – To report gastrointestinal stromal tumor as a source of upper gastrointestinal bleeding, which required urgent surgical control. ***Patient/Method*** – A man with 61 years old was admitted to the emergency service sustaining hematemesis and melena. Endoscopy showed active bleeding from a tumor in the second portion of the duodenum, which was controlled by heater probe cauterization. Surgery was performed through a median laparotomy. A local resection of a 4 cm tumor in the second portion of the duodenum was carried out, together with a primary end-to-end anastomosis and a duodenal diverticulization. No complications happened during the post-operative period. Morphologic examination showed gastrointestinal stromal tumor with no atypical mitosis and a preserved capsule. ***Conclusion***: Albeit not being common, gastrointestinal stromal tumors can represent a source of substantial gastrointestinal hemorrhage.

HEADINGS – Gastrointestinal, hemorrhage. Duodenal neoplasms. Smooth muscle tumor. Stromal cells.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are defined as mesenchymal neoplasms originating in the muscular wall of hollow viscera. Their first description as leiomyomas or leiomyosarcomas happened by reason of the histological appearance, resembling uterine tumors[7]. Nowadays there is compelling ultrastructural and immunohistochemical evidence demonstrating that these lesions can exhibit neural ganglionic and neural-myoid histological features[7].

Characteristically rare, these neoplasms account for nearly 0.01% of hospital admissions[3]. The peak incidence occurs in patients older than 50 years old, and stomach is the most frequent organ involved[5]. Benign lesions seldom present significant clinical signs, and gastrointestinal bleeding might be the only symptom[2]. Ten year survival ranges between 50% and 70%, depending on specific prognostic factors[5].

The authors report a duodenal stromal tumor as source of upper gastrointestinal bleeding, which required urgent surgical control.

CASE REPORT

A 61 years old man was admitted at Emergency Service of the "Santa Casa", São Paulo, SP, Brazil, transferred from another hospital, sustaining hematemesis and melena. He had been taking acetylsalicylic acid because of a previous deep venous thrombosis and chronic venous edema. There were no other associated diseases. He had already been admitted previously due to gastrointestinal hemorrhage, but there was no diagnostic investigation. Physical examination showed tachycardia, cutaneomucosal pallor and blood pressure reached 110/60 mm Hg.

Before the transference, 5 units of packed red blood cells had been transfused. At the admission, hemoglobin was 10 g/dL and coagulation tests confirmed no alterations. Endoscopy revealed a bleeding tumor in the second portion of duodenum, which was successfully controlled with heater probe cauterization (Figure 1). Computed tomography scan showed a 4 cm mass in the second portion of duodenum, without any signs of metastasis or carcinomatosis (Figure

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2). During diagnostic investigation, patient became hemodynamically unstable despite volume replacement and the operation was indicated.

Operative treatment was carried out through a midline longitudinal laparotomy. During the intraoperative assessment, there were no signs of advanced malignancy. An extended Kocher maneuver exposed the well limited, apparently encapsulated 4 cm tumor, exactly in the transition of the 2nd and 3rd duodenal portions (Figure 3). Considering that pancreas and major papilla were not involved, a local resection was performed, with a 1 cm disease free margin. Bearing in mind the identification and preservation of the major papilla, duodenum was primary anastomosed (Figure 4). In order to divert gastric contents away from the duodenum, a partial gastrectomy with Billroth II gastrojejunostomy was carried out. A small 0.5 cm nodule in the jejunal serosa, approximately 100 cm from Treitz's ligament, was also resected. Duodenal suture was widely drained. By the end of operation, nine units of packed red blood cells had been transfused, and patient sustained hemodynamic stability.

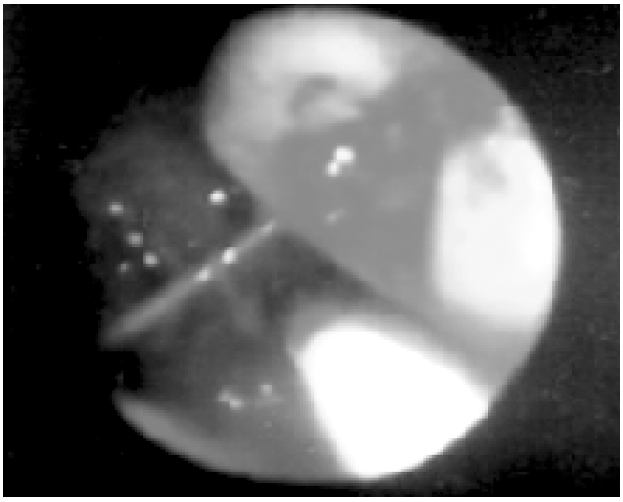

FIGURE 1 – Endoscopy view of the duodenal tumor and active bleeding

FIGURE 3 – Intraoperative view: duodenal tumor

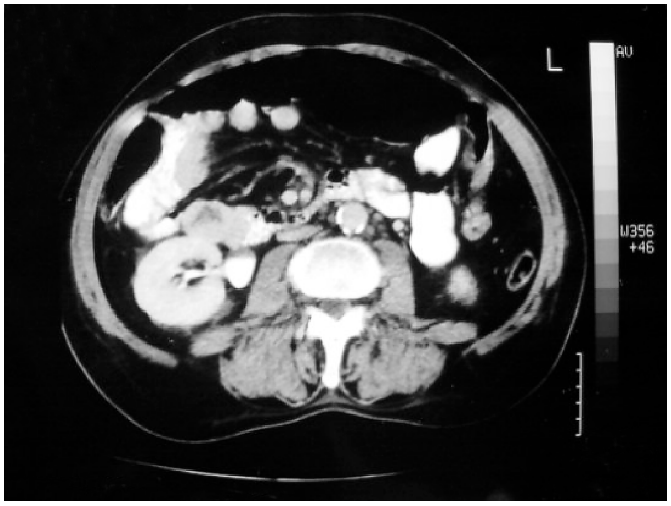

FIGURE 2 – Computed tomography showing 4 cm duodenal tumor anterior to the right kidney

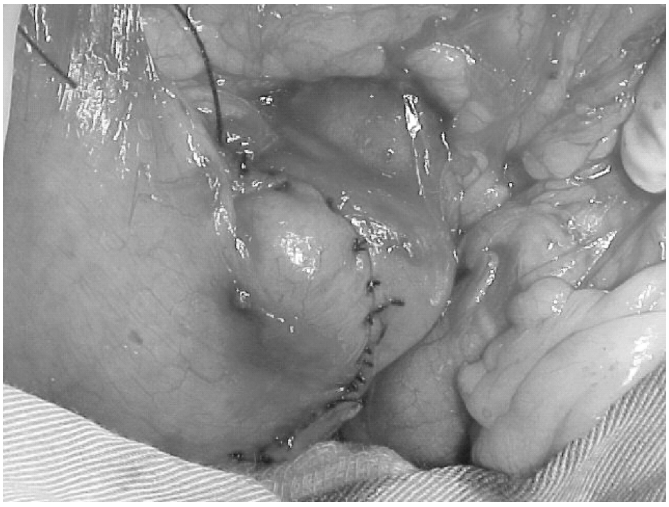

FIGURE 4 – Intraoperative view: final aspect of the duodenal anastomosis

An uneventful recovery followed through postoperative period. Drains were retrieved on the 6th postoperative day (POD), oral feeding started on the 7th POD, and the patient was discharged home on the 9th POD. Histological exam confirmed a 4.0 *vs.* 3.5 *vs.* 2 cm GIST, free from atypical mitosis figures and rounded by an intact fibrous capsule. Immunohistochemical assessment was positive for vimentine, actin 1 and 4 and S100 protein, while negative for ceratin and desmin. This suggests a smooth muscle and neural differentiation. Curiously, the small jejunal nodule was characterized as a synchronous GIST.

DISCUSSION

Normally, gastrointestinal bleeding secondary to small bowel tumors arises like a progressive process, determining chronic anemia and bringing up diagnostic difficulties[12]. Only 44% to 50% of leiomyomas present symptoms, and another considerable number are identified at autopsy or unrelated surgery[2, 10, 11]. However, specifically in this case, endoscopy could locate the source of bleeding and diagnosis was easily established. The severity of the hemorrhage, which required urgent surgical control, might also be considered unusual for this sort of tumor.

Surgery remains the standard of care for GISTs. Regarding periampullary duodenal tumors, the treatment of choice for malignant lesions consists of pancreaticoduodenectomy, which should be performed only if physiologic conditions permit[2]. Segmental resection is generally preferred for benign neoplasms sparing major papilla. In critical circumstances, morbidity and mortality increase, and major surgical procedures should be avoided[2]. Fortunately, in our case the neoplasm was deemed to be benign and spared major papilla, which allowed a local resection.

In this setting, suture dehiscence and duodenal fistula may be a serious complication. Sometimes, this sort of fistula is difficult to manage and reoperation might be necessary, which would definitely worsen prognosis. In order to divert gastric contents away from duodenal anastomosis, "diverticulization" of the duodenum through gastrectomy and gastrojejunostomy (Billroth II reconstruction) was performed.

Duodenal diverticulization was first reported by DONOVAN and HAGEN[4], in 1966, and subsequently updated by BERNE et al.[1], in 1968. This technique has been utilized for the treatment of complex pancreatoduodenal injuries. Nowadays, this procedure includes the suture of duodenal wound, together with a gastric resection and reconstruction through a gastrojejunostomy (Billroth II). As a consequence, gastric contents would be diverted away from the duodenum, decreasing pancreatic secretions. Therefore, it would protect an eventual anastomotic fistula, permitting oral intake and contributing to an earlier healing[3]. Nevertheless, no consensus exists about its use for non-traumatic purposes. Certainly, a duodenal wound with high chance of dehiscence represents an indication for duodenal diverticulization. Under our judgment, this is the exact situation in the case discussed. This old patient experienced a period of arterial hypotension, underwent an operation in a critical circumstance, received nine units of packed red blood cells and sustained a wound of great magnitude in the proximity of the major papilla. Thus, considering the high possibility of a subsequent fistula, duodenal diverticulization was carried out. However, the benefits of this surgical procedure must be weighed against the well known undesirable consequences of a gastric resection. Complications as anastomotic dehiscence, bleeding, and dumping syndrome have been reported[3].

A lacking of correlation between histological exam and prognosis has already been described for GISTs[5]. Sometimes, the identification of malign features is troublesome and might result in misleading conclusions. Concerning duodenal GIST, LEWIN et al.[8] suggest that lesions bigger than 4 cm or presenting atypical mitosis figures should be considered malignant. EVANS'[6] criteria for malignancy might be employed, which are: increased cell size, increased irregularity of cell size and shape, lack of complete cell differentiation, presence of plump cells with oval nuclei, and presence of cells with hyperchromic and multiple nuclei with variable staining reactions. EMORY et al.[5] analyzed 1,004 cases of stromal tumors, stating that site in gastrointestinal system, patient's age, tumor diameter, and presence of atypical mitosis remain the most important prognostic factors. Some investigators believe that the presence of more than two mitotic figures in 10 high power fields correlates to malignancy and worse prognosis[2].

Immunohistochemical methods have emerged to help in the differential diagnosis. Although GISTs encompasses most tumors previously classified as smooth muscle neoplasms, some express markers of neural origin. These kinds of lesions typically express CD 117, often are positive for CD 34, and occasionally carry alpha smooth muscle actin[9]. Nonetheless, these markers commonly have irregular behavior, varying in occurrence depending on the position in the gastrointestinal tract[9].

In the case reported, histological analysis confirmed GIST with smooth muscle and neural differentiation. Neither histological nor clinical evidences of malignancy were found. Surgical margins showed no signs of neoplasm. Therefore, we considered it as a benign lesion and suggested no other therapy.

Finally, in spite of representing an unusual neoplasm, GIST should be remembered as a cause of gastrointestinal hemorrhage. During operation, surgeon must be prepared to recognize malignant lesions with the purpose of managing them correctly. The appropriate operation depends on factors as general conditions of the patient, urgent situation, size and location of the tumor as well as suspicion of malignancy.

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Parreira JG, Freitas W, Rasslan S. Hemorragia digestiva alta por tumor estromal do duodeno. Arq Gastroenterol 2003;40(3):188-191.

RESUMO – ***Racional*** – Tumores estromais gastrointestinais são neoplasias raras que se originam na túnica muscular das vísceras ocas. ***Objetivo*** – Descrição de um caso de hemorragia digestiva alta causada por um tumor estromal duodenal que necessitou tratamento cirúrgico de urgência. ***Paciente/Método*** – Paciente com 61 anos de idade foi admitido no Serviço de Emergência da Santa Casa de São Paulo com hematêmese e melena. A endoscopia digestiva alta demonstrou um tumor na segunda porção duodenal com sangramento ativo, que foi controlado com cauterização por "heater probe". A operação foi indicada por instabilidade hemodinâmica persistente, sendo realizada por laparotomia mediana supra-umbilical. O tumor da segunda porção duodenal de aproximadamente 4 cm foi ressecado com margens de 1 cm. A anastomose primária duodenal término-terminal foi possível, sendo também indicada diverticulização duodenal através de gastrectomia parcial com reconstrução a Billroth II. O período pós-operatório transcorreu sem problemas. Exame anatomopatológico demonstrou um tumor estromal sem atipias e envolto por cápsula fibrosa íntegra. ***Conclusão*** – Mesmo não sendo sua apresentação mais comum, tumores estromais gastrointestinais podem ser causa de grave hemorragia digestiva alta.

DESCRITORES – Hemorragia gastrointestinal. Neoplasias duodenais. Tumor de músculo liso. Células estromais.

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