A RARE CASE OF SMALL DUODENAL GIST

Sachin B Ingle, Chitra R Hinge (Ingle), Surabhi Chopra
Department of Pathology, Anaesthesia and ENT MIMSR Medical College, Latur
*Author for Correspondence

ABSTRACT
Gastrointestinal stromal tumors (GIST) are mainly located in the stomach and the small bowel, with the duodenum accounting for about 4%. We report the case of a 25 year-old male with a GIST of the duodenum that was treated by resection and end to end anastomosis. Since no definitive clinical criteria have been established to differentiate malignant from benign mesenchymal tumors, preoperative cytology was not available and surgical removal was mandatory.

Key Words: GIST and Duodenal Neoplasm

INTRODUCTION
Gastrointestinal stromal tumors (GIST) are mainly located in the stomach, small bowel and colon (Rudolph, 2002). Duodenal localization is rare, accounting for about 4% of all cases (Flinner, 1991). Since the clinical and pathological criteria to differentiate benign from malignant stromal neoplasms are not well defined, it can be troublesome to make the choice between local excision (when feasible) and radical resection. Here we present a rare case of a small duodenal GIST conservatively treated by resection and end to end anastomosis.

CASES
A 25-year-old male was admitted in YCRH Latur with a recent history of epigastralgia and mild anaemia (hemoglobin 13.2 g/dl, hematocrit 38.1%). Tumor markers including CEA, TPA, CA 19-9 and CA 72-4 were within normal limits. Upper abdomen ultrasonography showed a mass with a maximum diameter of 3.0 cm in third part of duodenum. Angio-CT revealed a uniformly enhanced hypervascularized tumor. Duodenal endoscopy showed an intact but friable and easily bleeding mucosa. Biopsy specimens were negative.

Figure 1: Showing characteristic fascicular pattern of GIST.

Then patient was submitted to excision of the mass. Since neither the pancreas nor the biliary duct was involved by the tumor it was decided to perform a local excision. The resected specimen consisted of an intestinal piece measuring 7 cm in length. On dissection oval encapsulated homogenous mass measuring 3.5 x 2 x 2 cm was noted just below the mucosa reaching up to the serosa, the cut surface was greywhite soft. No areas of hemorrhages and necrosis on gross examination. Microscopic examination showed an intramural proliferation of relatively uniform spindle cells
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with eosinophilic cytoplasm, arranged in short fascicles or whorls. The nuclei were oval shaped with fine vesicular chromatin. There were fewer than three mitoses per 50 high-power fields (HPF). No areas of necrosis or hemorrhage were seen (Figure 1).

Immunohistochemical characterization showed diffuse and strong cytoplasmic immunopositivity for CD117 (KIT), whereas immunoreactions for desmin and smooth muscle actin were negative. Less diffuse and faint positivity was also recorded for S-100 protein. The diagnosis was a Gastrointestinal stromal tumor of the duodenum. The postoperative course was uneventful and the patient was discharged on the seventh postoperative day and doing well till today.

DISCUSSION

Mesenchymal neoplasms are the largest group of primary nonepithelial tumors of the gastrointestinal tract. Within this group of neoplasms, GISTs are characterized by a high density of spindle-shaped cells that immuno- histochemically reveal differentiation toward interstitial cells of the Cajal-expressing c-kit oncoprotein (Rudolph, 2002). Their biological behavior can be malignant and malignancy is mainly associated with distal anatomical site (Emory, 1999), larger tumor size, nuclear pleomorphism and higher mitotic activity (mitotic index) (Miettinen, 2003). Radical tumor resection is the only cure available for these patients.

More recently, in a retrospective investigation of 53 patients with gastric stromal tumors it has been documented that radical resection with regional lymphadenectomy provides no benefit in terms of local recurrence and long-term survival compared to local excision with tumor free margins (Douglas, 1995; Lehnert, 1998; Grant, 1991). This is probably due to the tendency of the tumor to spread via the hematogenous rather than the lymphatic route (Miettinen, 2003). Since location in the upper gastrointestinal tract (stomach and duodenum), small size (<5 cm diameter) (Douglas, 1995) and low mitotic activity (mitotic index <5 mi- toses/50 HPF) are likely to identify very low risk tumors, it is reasonable to consider local excision of the 3.5 cm tumor of our patient as radical treatment in spite of the lack of preoperative immunohistochemical analysis. The great majority of duodenal mesenchymal tumors are GISTs, which show a variety of biological behaviors. Those arising from the gastrointestinal autonomic nerve tissue (GANT) (Meesters, 1998) or fibrous tissue (GIFT) (Rudolph, 2002) are more aggressive. However, in view of the differentiation potential of interstitial Cajal cells and of the shared genetic features with the conventional spindle cell and epithelioid cell forms of GIST, it appears no longer临床- useful to discriminate between these tumors and GISTs (Lee, 2001). A mutilating and more radical resection could therefore be recommended for neoplasms of larger size (>5 cm) and high mitotic index (Takahashi, 2001) especially in the case of younger patients. Consequently, all efforts should be made to obtain a preoperative histological diagnosis. Today, the availability of endoscopic ultrasound-guided fine-needle aspiration biopsy (Fu K, 2002) is helpful to indicate the correct surgical procedure, especially, as in the case reported here, in the case of small gastroduodenal submucosal mesenchymal tumors with negative endoscopic biopsy results.

REFERENCES


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