A huge gastric stromal tumor in a 13-year-old girl

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A 13-year-old girl presenting with severe anemia was diagnosed to have a large gastric tumor protruding toward the antrum with two central ulcerations. Partial gastrectomy including antrectomy and gastroduodenostomy were performed. Histologic and immunohistochemical studies revealed one of the most uncommon gastric tumors in children; a gastrointestinal stromal tumor. Close follow-up of the patient with endoscopy, abdominal ultrasonography and/or computed tomography in three to six month intervals revealed no recurrences or metastasis of the tumor following its complete excision.

Key words: gastrointestinal stromal tumor, stomach, immunohistochemistry, childhood.

Gastrointestinal stromal tumors (GIST) constitute the most uncommon category of primary nonepithelial tumors of the stomach and small bowel. They are supposed to arise from the cells located in the walls of the organs[1]. GIST have a wide variability in their clinical behavior, and malignant potential is often difficult to predict. Although there are no exact criteria for determining the malignant condition and prognosis of GIST, there are some variables that affect the prognosis[2].

To the best of our knowledge the presented case is the youngest patient with gastric stromal tumor[2]. Although the patient did not fulfill the criteria for a manifest malignancy, the tumor seemed to have had a malignant potential. Because of the rarity of GIST in childhood and limited information about the long-term survival and life expectancy, periodic endoscopy and radiologic investigations seem to be important in the follow-up of these patients.

Case Report

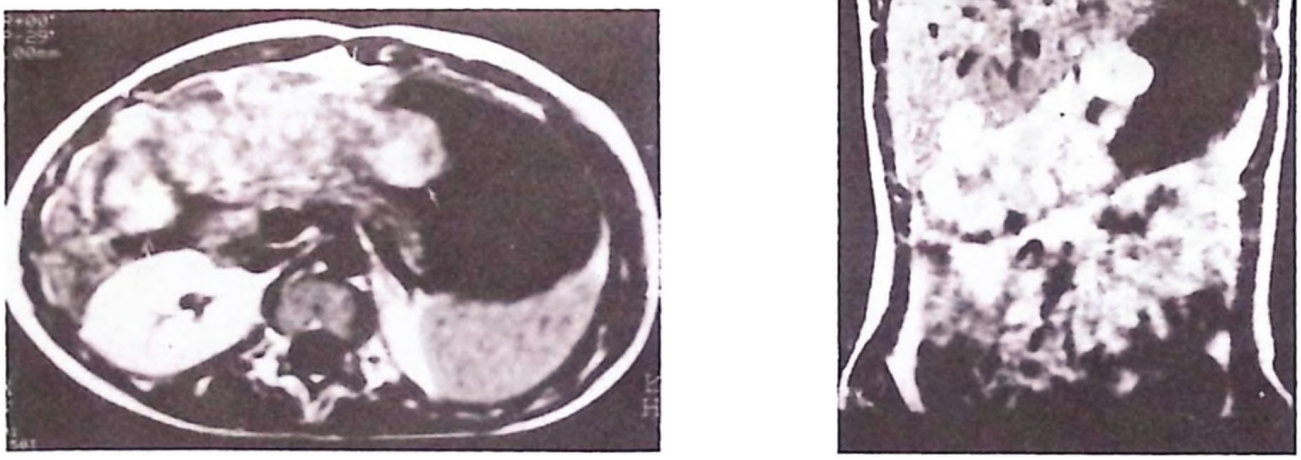
A 13-year-old girl presented with easy fatigability, paleness and anemia of two years' duration. Laboratory investigation revealed low Hb and Htc levels (6.0 g/dl and 19.9%, respectively), a moderate deficiency of serum iron (38 µg/dl) and ferritin (8.5 µg/dl) with normal WBC and platelet counts. The occult blood in stool was strongly positive. Abdominal ultrasonography (US) showed a tumor diffusely infiltrating the stomach from the lesser curvature through the antrum, extending into the gastric lumen and exophytically outside the gastric wall. No other metastatic lesions or lymph nodes were identified. Magnetic resonance imaging (MRI) confirmed the mass (Fig. 1a and b). Gastroscopy that was performed just before the operation revealed an ulcerated mass protruding into the gastric lumen which bled easily on touch. During laparotomy, a multilobulated exophytically developed large mass originating from the lesser curvature was found. The tumor mass extended through the antrum and protruded into the lumen but did not obstruct the first part of the duodenum (Fig. 2a). Two deep ulcerations were present on the mucosal surface (Fig. 2b). Frozen section performed from the serosal nodules revealed a benign tumor suggestive of leiomyoma. Partial gastrectomy including antrectomy with a safe margin was performed. The corpus was tubularized and gastroduodenostomy was done. Microscopic examination of the tumor with H&E staining showed fusiform cells with acidophilic cytoplasm and round-to-polygonal cells with central nucleus in most of the areas, resembling a leiomyoma (Fig. 3a). Mitotic activity was low, less than 5 per 50 high-pover fields. Some areas showed features of neural differentiation and were composed of palisading spindle cells (Fig. 3b). Immunohistochemical staining with antibodies to S-100 protein

(DAKO, antiserum), neuron specific enolase (DAKO, clone: BBS/NC/VI-H 14), and smooth muscle actin (DAKO, clone: 1A4) were all negative. These histopathological and immunohistochemical findings were consistent with GIST, lacking differentiation toward any cell type.

Postoperative course of the patient was uneventful. Three months after the operation the patient had a normal Hb level and had no complaints with a normal gastroscopic examination. The abdominal US and computed tomography findings did not reveal any mass one year after the operation.

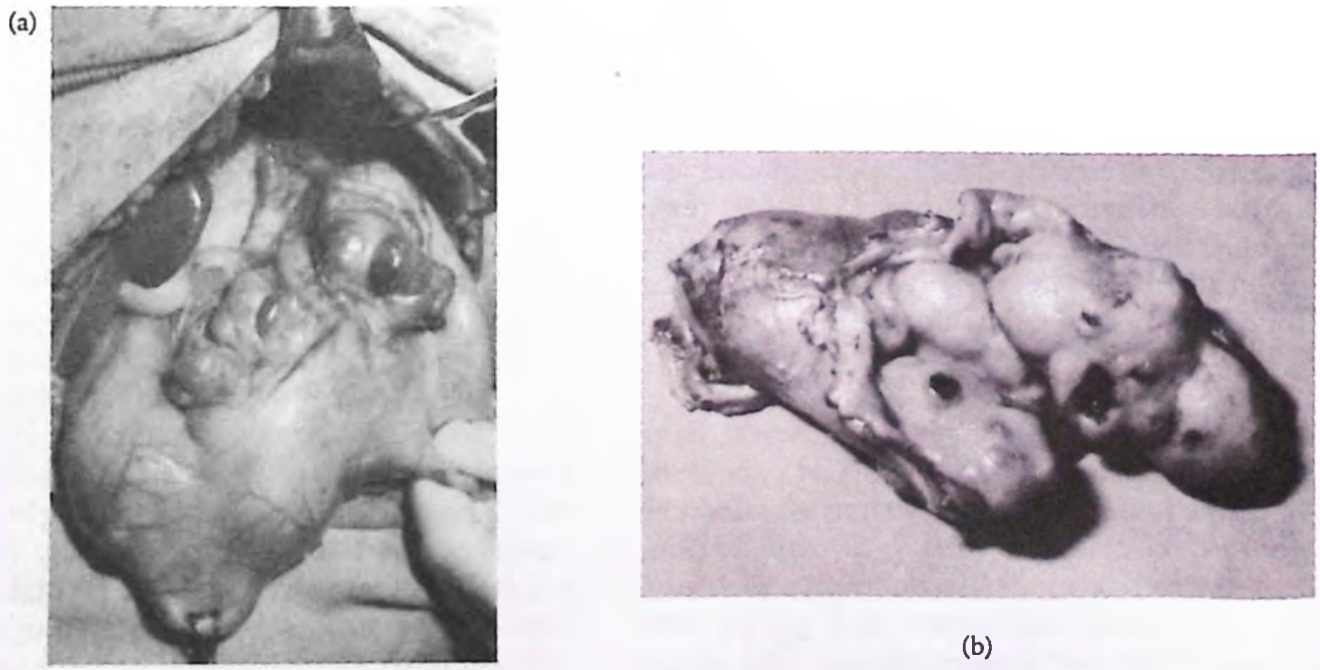
Discussion

Mesenchymal tumors of the gastrointestinal system arise from the cells located in the wall of the stomach and small bowel[1]. They are a heterogeneous group of tumors with unclear cell lineage[3,4]. However, immunohistochemical studies using S-100 protein, desmin, vimentin and smooth muscle actin show that gastrointestinal



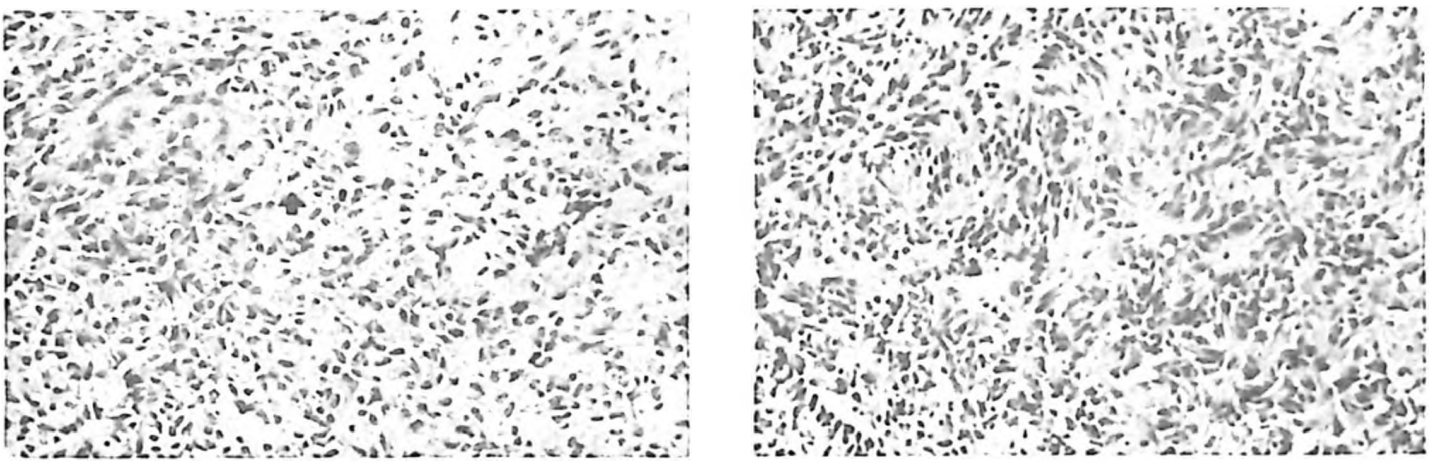
(a) (b)

Fig. 1. a) Transverse section of magnetic resonance image showing the huge tumor (arrows) originating from the lesser curvature of the stomach b) The contour of the mass (arrows) extending exophytically outside the stomach wall and protruding into the gastric lumen in the coronal section.



(a) (b)

Fg. 2. a) Gross appearance of the large tumor mass. b) The mucosal surface of the mass showing deep ulcerations (arrows) protruding into the gastric lumen.



(a) (b)

Fig. 3. a) Tumor area resembling leiomyoma with round-to-polygonal cells and some fusiform cells with scanty mitoses (arrow) (H&E stain, original magnification x 66). b) Tumor area with palisading spindle cells resembling neural differentiation (H&E stain, original magnification x 66).

mesenchymal tumors may be composed of cells showing differentiation toward smooth muscle cells and neural elements, dual differentiation toward both elements or showing no differentiation toward any cell type[1,3]. Tumors in the last category are undifferentiated tumors and are referred to as GIST[3].

Gastric stromal tumors (GST) may be asymptomatic for a long period of time. Symptomatic stromal tumors usually present with occult bleeding or severe anemia[5]. Neither malignant nor benign GST cause obstructive symptoms or symptomatic metastasis causing pain or weight loss[5]. Although endoscopy is indicated and demonstrate[5] the tumor mass, diagnostic yield of endoscopic biopsy is low because the mucosa may only show nonspecific pathologic changes[6]. Upper gastrointestinal series, abdominal US or computed tomography indicates the gastric tumor[5]. A spherical gastric filling defect with rounded regular edges and a smooth overlying mucosa suggest GST[5]. These tumors most frequently metastasize to omentum, liver or peritoneum. The above-mentioned features of GST differentiate them from other gastric lesions such as lymphoma and adrenocarcinoma[5].

Tumors smaller than 5 cm in diameter confined to the stomach have been reported to have a favorable outcome[3,6,7]. A high mitotic rate (over 5 mitoses per 50 high power field), cellularity and nuclear atypia are classifed as malignant but a low mitotic index does not indicate a benign course[5,6]. It is extremely difficult to distinguish benign lesions from malignant ones either by frozen section at the time of surgery or by postoperative routine histopathologic and immunohistochemical analyses[5,6]. Other factors in determining malignancy are resectability and/or presence of metastasis[6]. Tumor necrosis and mucosal ulcerations have also been stated to have a worse prognosis[3]. The tumor should be completely resected with negative tumor margins. These tumors have not been proven radiosensitive and no efficacious chemotherapy has been documented[5,8].

Although microscopic examination of the tumor with H&E revealed a leiomyoma-like pattern, some areas showing features of neuronal differentiation necessitated immunohistochemical staining with antibodies to S-100 protein, neuron specific enolase and smooth muscle actin, which were all negative, and the tumor was diagnosed as GIST of the stomach. In our case although the huge tumor size, serosal invasion and mucosal ulcerations were factors indicating poor prognosis, absence of high mitotic rate and nuclear atypia, total resection of the tumor with safe margins and absence of metastasis were more promising for a better outcome.

Gastric stromal tumors are uncommon in children. Because of the rarity of the disease, particularly in childhood, the evaluation of malignant potential, surgical treatment, adjuvant therapies and prognosis of the disease depend on the experiences of adult series. An upper abdominal mass with severe anemia in an otherwise healthy individual should suggest a gastric stromal tumor. An aggressive initial surgical approach seems to be the best treatment of these lesions. The unpredictable clinical and

histological behavior of the tumor necessitates a long-term follow-up of these patients with endoscopy and radiologic investigations.

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