ABSTRACT

Aim: Gastric hemangiomas (GHs) are rare vascular tumors representing an extremely uncommon cause of gastrointestinal bleeding in children. Up to now, only few cases have been reported in pediatric literature. Methods: The Authors report the case of a 6-year-old-girl with GH presenting with hypovolemic shock due to acute gastro-intestinal bleeding. Focusing on diagnostic and therapeutic assessment, a detailed systematic review of pediatric literature on GH is provided. Results: In our patient a combination of abdominal ultrasound, endoscopy and CT scan were helpful in reaching a diagnosis. Gastric antrum resection and Billroth’s I gastro-duodenal anastomosis was curative. Conclusions: A not invasive diagnostic work-up, including ultrasound as first line examination should be performed in every previous healthy children presenting acute and/or chronic GI bleeding. Once diagnosis of GH is made the appropriate endoscopic and/or surgical treatment should be accomplished as early as possible to avoid severe and potentially life-threatening complications.

Key words: Gastric hemangioma, Children, Ultrasound scan (US), Gastrointestinal bleeding

INTRODUCTION

Gastric hemangiomas (GHs) are rare vascular tumors representing an extremely uncommon cause of gastrointestinal bleeding in children.1,2 Up to now, only 18 cases have been reported in pediatric literature.1-18 The Authors report the case of a 6-year-old-girl with GH presenting with hypovolemic shock due to acute gastro-intestinal bleeding. Diagnosis of GH was made and gastric antrum resection and Billroth’s I gastro-duodenal anastomosis were promptly performed. Focusing on diagnostic and therapeutic assessment, a detailed systematic review of pediatric literature on GH is provided.

CASE REPORT

A 6-year-old-girl was referred to our Department because of hypovolemic shock due to acute gastro-intestinal bleeding with hematemesis and melena. Her clinical history was unremarkable; no drug consumption was recorded. At admission abdominal tenderness was evident at palpation in epigastrium and left hypochondrium. No cutaneous hemangioma or other vascular skin lesions were detected. Hemoglobin was 7.5 g/dl, hematocrit 21.8%, platelet count was 354,000 per cubic millimeter. Prothrombin and partial-thromboplastin and routine blood chemistry were normal. At abdominal ultrasonography (US) a large, not homogeneous, intraluminal and hyperechoic mass with well delineated edges was detected at the gastric antrum (Figure 1 A). Liver, pancreas and spleen were normal. Upper GI tract X-ray showed an almost complete obstruction of the gastric outlet and gastric dilatation (Figure 1 B). Abdominal computer tomography (CT) scan showed a 4x4-cm round protruding mass arising from the posterior wall of the gastric antrum, with vascular enhancement. (Figure 1 C). Upper GI tract endoscopy was then performed which showed a large, polypoid protruding mass at the posterior wall of the gastric antrum. The mass was bluish even if partially covered with clots and fibrin, and some bleeding was coming from the medial and inferior surface of the tumor. Surgery was promptly done. The abdomen was entered through a midline incision and a short gastrostomy on the anterior wall of the gastric antrum where the tumor could be palpated was performed. Gastrointestinal continuity was accomplished by a gastrectomy and Billroth’s I gastro-duodenostomy. Careful examination of the other abdominal viscera showed no other vascular malformations. Gross examination of the resected specimen showed a polypoid, sessile and ulcerated gastric tumor measuring 3.5 cm in diameter (Figure 2 A). Resection edges were free from disease. Pathology showed a network of abnormal proliferating blood vessels endothelium, intermingled with loose connective tissue containing some miofibroblastic cells (Figure 2 B and C). Proliferating vessel were present within all layers of gastric wall...
and endothelium showed intense CD34 and factor VIII staining. Diagnosis of cavernous hemangioma of the stomach was proposed. The postoperative course was uneventful. At 12-year follow-up the patient is growing normally and is asymptomatic.

**Figure 1.** A: Ultrasound (US) examination showing a large hyperechoic mass in the gastric antrum protruding in the lumen and with no extension beyond the gastric wall. B: Upper contrast X-ray showing an almost complete obstruction at gastric outlet and gastric dilatation. C: CT scan showing a round mass protruding from the posterior wall of the gastric antrum. No calcifications or adenopathies are seen.

**DISCUSSION**

**Epidemiology**

GH is very uncommon accounting for only 0.05% of GI tumors in all patients and 1.6% of all benign tumors of the stomach. Up to now, only 18 cases (13 females) are reported in childhood. Nevertheless, as suggested by Fishman, the magnitude of the disease could be underestimated, as there are many different and sometimes confusing terms used to identify the same vascular anomalies. Median age at symptom presentation was 3.4 years (range 7 days to 13 years); neonatal presentation is extremely uncommon (18%). Median age at treatment was 8.7 (range 0.4-24) years, with a long delay between the age at symptoms presentation and age at treatment. In 7 cases (47%), GH was associated with similar skin lesions and in 4 of these, GH being also part of diffuse hemangiomatosis of the GI tract.

**Figure 2.** A: Gross examination of the resected specimen showing a polypoid, sessile and ulcerated tumor measuring 3.5 cm in diameter. B: Intense CD34 staining endothelium. C: Histological examination: network of abnormal proliferating blood vessel with, intermingled with loose connective tissue containing some miofibroblastic cells.

**Pathological examination**

GH usually ranges from a few millimeters up to 2 cm in the greater diameter. Largest tumors may infiltrate most of the stomach walls or be polypoid and largely protruding into the gastric lumen, as observed in our case. Based on the capillary network size and GI tract involvement, Taylor classified GH into 4 types: a) cavernous (dilated vessels usually with thin walls), b) capillary (vessels are very small and in the early phase, lesions are lobulate and highly cellular), c) mixed capillary and cavernous type, d) multiple teleangectasia and diffuse hemangiomatosis. The cavernous type was the most
frequent, being present in 67% of cases in whom the anatomical diagnosis was reported.

**Clinical presentation**

The clinical presentation of GH is variable (Table 1). Single or recurrent episodes of hematemesis were the commonest symptom recorded in this series (60%), mostly associated with melena. In the remaining cases bleeding was chronic and insidious and often associated with unspecific complaints like mild epigastric pain, dyspepsia and/or microcytic, hypochromic anemia due to the chronic blood loss which was persisting for several years. Acute, severe and life-threatening GI bleeding due to ulceration of a giant hemangioma as presenting symptom has been reported only in 2 cases (13%), including our patient in whom it caused hypovolemic shock. In these cases, the poor tumor vitality related to the unpaired circulation inside the tumor itself, together with the gastric acid, are supposed to be the factors leading to the hemangioma erosion and torrential bleeding. Obstructive symptoms have been described only in adults.30

**Diagnostic assessment**

The diagnostic work-up following GI bleeding in children includes a variety of imaging techniques. Upper GI tract RX is almost completely abandoned, being replaced by other more sensitive radiological investigations. Although recent advances in US technology have improved the US diagnostic accuracy in GI disease, US has never been reported in the diagnostic work-up of GH.21 US can differentiate vascular parietal tumors from solid or cystic lesions and has a high sensitivy in detecting more common pediatric causes of GI bleeding, like obstruction of the portal vein (cavernoma). Moreover, US is not invasive and can be easily performed regardless of the patient’s clinical condition. Abdominal CT scan and MRI are the next appropriate, noninvasive investigations to confirm the diagnosis of GH.11,14,22 Unenhanced CT scan may demonstrate small soft tissue calcifications (fibrolititis) within the gastric wall,23 which are, since a long time, considered to be pathognomonic of GH.24 Contrast-enhanced CT scan shows areas with strong vascular enhancement with the “filling-in” pattern in some cases, which is consistent with cavernous hemangioma.11 Actually, MRI is considered the preferred diagnostic investigation for GH in children.22,25 MRI accurately defines the extent and nature of this vascular tumor without providing any radiation to the patient. We choose CT scan instead of MRI since at the time we treated our patient, MRI was not available in emergency setting. Angiography has been reported to be definitive in order to establish the diagnosis of GH and location,4,10,12,24,26 but now it can be replaced by the above mentioned examinations. In adults, angiography has been reported to successfully allow a GH transcatheater treatment by embolization,27 but in the only pediatric case in which left gastric artery embolization was performed, it failed to stop bleeding from GH.13 Furthermore arterial embolization is not well performing in the capillary type vascular malformations12 and finally angiography is quite invasive and potentially harmful in small children. Upper GI tract endoscopy, although has been reported not to be diagnostic in all cases in which it was done,11 can induce the suspicion of a vascular lesion. Endoscopic biopsy of the tumor is not advisable as it may results in massive hemorrhage.

**Therapeutic options**

The natural history of asymptomatic GH is unknown. On the contrary, symptomatic lesions need an aggressive treatment since they don’t supersede over years, but potentially life-threatening complications have been reported. In children, up to now, wedge excision has been the treatment of choice for small GH (< 2 cm) (Table 1). More recently, the growing experience in the endoscopic treatment of different and life threatening bleeding lesions of the stomach should result in exploration of the feasibility of the endoscopic treatment for small GH, as well as reported for hemangioma located in the large bowel. A major advantage might come from the endoscopic ultrasonography. Up to now endoscopic ultrasonography, has been successfully employed only in adult patients.28 Endoscopic ultrasonography has been reported allowing a better definition of gastric wall layers involved with the tumor, that in case of GH are confined mainly to the submucosal layer of the gastric wall,29 helping in a safer endoscopic resection. A more liberal use of this technique might be advised, above all in older children in whom larger endoscopes can be used. The therapeutic decision is more difficult in case of extended or giant GH. In these patients an individualized and interdisciplinary approach based on pharmacological treatment, endoscopy, interventional radiology and surgery is often required to avoid extended gastric resections. In GH, Pharmacological angiogenesis inhibition (PAI) has been used in few patients (Table 1), but it was unsuccessful in all except one infant treated with interpheron alpha-2B, after failure of steroid therapy (Table 1). Furthermore, in that patient interferon administration was preceded and subsequently associated with repeated sessions of endoscopic argon plasma coagulation. In case of giant or extended GH, medical treatment should be advised only in patients in whom the tumor is in the proliferative phase, and mostly in case of capillary type hemangioma, which usually well responds to treatment with steroids or interferon.29 Patients with an established lesion more probably would not benefit from antiangiogenic therapy. Propranol in the management of visceral hemangioma has been used with promising results.30 In large, unifocal lesions, sclerocant therapy and endoscopic laser carries a high risk of mucosa and submucosa necrosis, as well as gastric perforation and should be avoided. Location of the tumor is another crucial issue. Alcohol injection in tumor located at or near the gastric antrum could cause a so severe gastric wall edema to determine a complete obstruction to gastric emptying. A good alternative might be the use of adhesive substances, like isobutyl-2-cyanoacrylate (Histoacryl), which has been reported very effective in controlling huge hemorrhages from varices of the gastric fundus.31

**Surgery**

Surgery still represents the gold standard of the treatment of extended or giant GH (Table 1). Whenever possible, wedge excision should be the preferred technique, however tumor
location and in some cases its extension dictate the choice of the technique. In our patient, due to the tumor localization at the gastric antrum and to her young age, we found Billroth’s I intervention was the safer operation to perform, as confirmed also by the long-term patient follow-up.

In conclusion, we reported an uncommon case of GH in childhood. Our case deserves interest both for its presentation, which makes mandatory a differential diagnosis with other more frequent disease causing severe GI bleeding, and for its size and location. Based on our experience in GI bleeding of different etiologies, we believe that a not invasive diagnostic work-up, including ultrasound as first line examination should be performed in every previous healthy children presenting acute and/or chronic GI bleeding. Once diagnosis of GH is made the appropriate endoscopical and/or surgical treatment should be accomplished as early as possible to avoid severe and potentially life-threatening complications.

Table 1. Systematic review of gastric hemangiomas in children

<table>
<thead>
<tr>
<th>Pt</th>
<th>Author</th>
<th>Sex, age</th>
<th>Clinical findings</th>
<th>Associated cutaneous hemangioma</th>
<th>Investigation</th>
<th>Treatment (age)</th>
<th>Pathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>McIntosh</td>
<td>F, 6 y</td>
<td>Melena, chronic anemia</td>
<td>Skin, small bowel, colon</td>
<td>UGIS</td>
<td>Wedge resection (36 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>2</td>
<td>Holden</td>
<td>F, 2 w</td>
<td>Neurological symptoms</td>
<td>Skin, small bowel, colon, brain</td>
<td>Necropsy</td>
<td>Died before treatment</td>
<td>Capillary</td>
</tr>
<tr>
<td>3</td>
<td>Singeton</td>
<td>F</td>
<td>Hematemesis</td>
<td>Skin</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>4</td>
<td>Mellish</td>
<td>F, 1 y</td>
<td>Severe anemia</td>
<td>Skin, small bowel, colon</td>
<td>UGIS, barium enema, angioscopy</td>
<td>Wedge resection (9 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>5</td>
<td>Mellish</td>
<td>F, 5 y</td>
<td>Melena, severe anemia</td>
<td>Skin, small bowel, colon</td>
<td>UGIS, arteriography</td>
<td>Wedge resection (6 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>6</td>
<td>Samarý</td>
<td>F, 2 y</td>
<td>Melena, hematemesis</td>
<td>Face, limbs</td>
<td>UGIS, EGDS, angiography</td>
<td>Pyloric resection (7 y); Total gastrectomy (11 y)</td>
<td>Diffuse angiomatosis</td>
</tr>
<tr>
<td>7</td>
<td>Taylor</td>
<td>F, 2 y</td>
<td>Hematemesis, severe anemia</td>
<td>Face, head, ears, leg</td>
<td>UGIS, EGDS, angioscopy</td>
<td>Total gastrectomy (4 y)</td>
<td>NA</td>
</tr>
<tr>
<td>8</td>
<td>Liebert</td>
<td>F</td>
<td>hematemesis</td>
<td>Isolated</td>
<td>NA</td>
<td>Partial gastrectomy (4 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>9</td>
<td>Oswalt</td>
<td>F, 13 y</td>
<td>Hematemesis, hemorrhagic shock</td>
<td>Isolated</td>
<td>NA</td>
<td>Wedge excision (15 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>10</td>
<td>Torricelli</td>
<td>F</td>
<td>Isolated</td>
<td>NA</td>
<td>NA</td>
<td>Partial gastrectomy (14 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>11</td>
<td>Nagaya</td>
<td>M, 7 d</td>
<td>Hematemesis, melena</td>
<td>Isolated</td>
<td>UGIS, EGDS</td>
<td>Subtotal gastrectomy (7 m)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>12</td>
<td>Schectl</td>
<td>F, 10 y</td>
<td>hematemesis</td>
<td>Isolated</td>
<td>EGDS, CT, MRI, angiography</td>
<td>Partial gastrectomy (10 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>13</td>
<td>Hardy</td>
<td>F, 1 m</td>
<td>Hematemesis, melena</td>
<td>Isolated</td>
<td>UGIS, CT, MRI, EGDS</td>
<td>Debulking (5 m)</td>
<td>Capillary</td>
</tr>
<tr>
<td>14</td>
<td>Hahn</td>
<td>F, 9 m</td>
<td>Melena, thrombocytopenia</td>
<td>Isolated</td>
<td>EGDS, CT, angiography</td>
<td>Corticosteroid, electrocoagulation, argon plasma</td>
<td>NA</td>
</tr>
<tr>
<td>15</td>
<td>Schettini</td>
<td>M, 5 y</td>
<td>Hematemesis, melena</td>
<td>Isolated</td>
<td>EGDS, MRI, US</td>
<td>Gastrectomy (5 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>16</td>
<td>Menon</td>
<td>M, 8 y</td>
<td>Hematemesis, melena</td>
<td>Isolated</td>
<td>EGDS, US, CT</td>
<td>Partial gastrectomy (8y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>17</td>
<td>Attash</td>
<td>M, 3 y</td>
<td>Hematemesis, melena, severe anemia</td>
<td>Isolated</td>
<td>US, EGDS, CT</td>
<td>Sleeve gastrectomy, splenectomy (3 y)</td>
<td>Cavernous</td>
</tr>
<tr>
<td>18</td>
<td>Garcia Hernandez</td>
<td>M, 7 y</td>
<td>Hematemesis, Hemorrhagic shock</td>
<td>Isolated</td>
<td>EGDS, MRI</td>
<td>Laparoscopic wedge excision</td>
<td>NA</td>
</tr>
<tr>
<td>19</td>
<td>Our case</td>
<td>F, 6 y</td>
<td>Hematemesis, melena, Hemorrhagic shock</td>
<td>Isolated</td>
<td>US, EGDS, CT, UGIS</td>
<td>Partial gastrectomy (6 y)</td>
<td>Capillary</td>
</tr>
</tbody>
</table>

Legend: M; male; F; female; y; year; m: month; d: day; EGDS: esophagogastroduodenoscopy; UGIS: upper gastrointestinal contrast study; US: ultrasound scan; CT: computed tomography, MRI: magnetic resonance imaging; NA: not available data.

REFERENCES