Endoscopic-assisted laparoscopic surgical removal of a gastric neurofibroma in a child

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A B S T R A C T  
An 11-year-old boy, who presented with abdominal pain and vomiting was noted to have a gastric submucosal mass at endoscopy. Endoscopic ultrasound showed it arising from the fourth ultrasound level of the gastric wall precluding endoscopic removal. Open surgery was avoided by use of endoscopic-assisted laparoscopic surgery (EALS) to remove the mass. The mass was found to be an isolated gastric neurofibroma, a rare tumor in children. We show that combined use of intraluminal endoscopy and laparoscopic surgery allows for safe and less-invasive surgery for removal of a submucosal mass in a child. Further, we review the rare finding of gastric neurofibromas.  

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1. Patient report  
An 11-year-old male presented with a several week history of abdominal pain and vomiting. The pain occurred daily without identifiable triggering or relieving factors, persisted for hours, and resolved spontaneously. The pain was localized to the epigastrium but was poorly characterized; it awakened him from sleep 5 days a week. He had associated vomiting, which was devoid of blood, coffee-ground material, and bile. He had no fever, diarrhea, rectal bleeding, jaundice, or weight loss. Past medical history was positive only for hospitalization for wheezing at 6 months of age, and incision and drainage of a suppurative lymph node at 2 years of age. Family history was significant for a hiatal hernia and gastroesophageal reflux in the father; there was no history of migraines, peptic ulcer disease, or inflammatory bowel disease. Examination revealed normal growth parameters, a normal abdominal examination, and a rectal examination negative for occult blood. Laboratory studies to include a complete blood count, erythrocytic sedimentation rate, and urinalysis were normal. He was placed on a weight-appropriate dose of omeprazole for 2 months, without improvement in the abdominal pain. An upper endoscopy was performed which showed erythema of the antrum without nodularity, erosion, or ulceration; a rapid urease test was positive within 15 min. A 1-cm submucosal mass with central ulceration was seen on the greater curvature of the body of the stomach (Fig. 1); it was non-compressible with closed biopsy forceps. Biopsies taken from the
antrum revealed moderate active chronic gastritis with *Helicobacter pylori* (Hp) organisms identified on toluidine blue-stained sections. The patient was treated with a 2-week course of amoxicillin, clarithromycin, and lansoprazole for Hp-induced gastritis and repeat endoscopy scheduled for endoscopic ultrasound and biopsy of the mass.

One month later the patient had resolution of his abdominal pain, and stool for Hp antigen was negative. At repeat endoscopy the mass was determined to arise from the fourth ultrasound level of the gastric wall suggesting a gastrointestinal stromal tumor (Fig. 2). Attempts at fine needle biopsy were unsuccessful so a needle knife was used to open the mucosa followed by pinch biopsies of the mass. The mucosal defect was closed with endoclips.

Immunohistochemical studies showed that the tumor cells were diffusely positive for S-100 and negative for desmin, myogenin, CD117 and CD34 confirming a diagnosis of neurofibroma (Fig. 4D and E).

After discussion with the family and Pediatric Surgery, it was decided to perform endoscopic-assisted laparoscopic removal of the gastric mass. Following informed consent, placement of appropriate monitoring lines, and induction of general anesthesia, a 5-mm trocar was placed. Carbon dioxide was infused into the abdominal cavity to a pressure of 15 mm Hg. A 30-degree laproscope was introduced into the abdominal cavity. Next, three 5-mm trocars were placed; one in the right upper quadrant and two in the left to mid upper quadrant. The stomach was examined laparoscopically, but the location of the mass could not be identified. The pylorus was then occluded with a grasper, and an upper endoscopy was performed showing a mass arising on the posterior wall of the stomach (Fig. 3). The stomach was desufflated, and the short gastric arteries were taken down using a harmonic scalpel. The stomach was then turned to expose the posterior wall. Once again, the pylorus was occluded with a grasper, and upper endoscopy was performed. Pressure was applied to the gastric mass from within the stomach with biopsy forceps, and the tented-out stomach was grasped on the serosal surface through the laproscope. The stomach was then desufflated, and an Endo GIA60 stapler™ (2.5-mm load) was placed across the stomach containing the gastric mass. Prior to deploying the stapler, the endoscope was advanced back into the stomach to confirm that the mass was successfully captured within the portion of the stomach to be removed. The stapler was then deployed to ligate and transect the portion of the stomach containing the mass. The stomach specimen was then removed through the umbilical port and submitted to pathology.

The tumor was a firm nodule (2.0 × 1.5 × 1.5 cm) with ulceration at the center (Fig. 4A), composed of interlacing bundles of bland spindle cells with wavy nuclei admixed with fibroblasts and characteristic dense bundles of collagen (Fig. 4B and C). Immunohistochemical studies again showed that the tumor cells were diffusely positive for S-100 and negative for desmin, myogenin, CD117 and CD34 confirming a diagnosis of neurofibroma (Fig. 4D and E).
obtained by fine needle aspiration and Tru-cut needle biopsy, the tissue sample makes up only a small portion of a tumor and may miss malignant cells. Excisional biopsy with adequate tumor margin is the preferred method for gastric submucosal tumors [3,4].

Minimally invasive surgery has revolutionized the approach to gastric tumors, highlighted by rapid advances in laparoscopic surgery (LS) and therapeutic endoscopy. Prior to LS, gastric tumors required open laparotomy. With the advent of LS and improvement in laparoscopic instruments a number of laparoscopic techniques have been employed for the removal of gastric wall tumors including enucleation, transgastric tumor-evertting resection, intragastric tumor wedge resection, and extraluminal wedge resection [3]. However, each of these techniques has drawbacks. Enucleation of GISTs is technically successful but there is a high rate of tumor recurrence [5]. Transgastric tumor-evertting resection can lead to intra-abdominal contamination with gastric juice, tumor seeding, and injury to the esophago-cardiac junction [3]. Intra-gastric wedge resection requires special equipment that may not be readily available at some centers and there remains a risk of tumor seeding and gastric mucosal injury [3]. A major drawback of extraluminal wedge resection is difficulty in determining the appropriate resection line as it is impossible to ascertain with certainty the margins when the tumor is intraluminal [6].

Endoscopic submucosal dissection (ESD) has become an acceptable modality for removal of benign and malignant gastric tumors with excellent success rates and safety. Initially, it was thought that endoscopic removal of gastric tumors arising from the muscularis propria was contraindicated due to a high risk of perforation and bleeding [1,7]. However, subsequent studies in small numbers of adults have shown that, in experienced hands, gastric tumors arising from the muscularis propria layer can be successfully removed (75%–90%) with low rates of bleeding (0%–10%) and perforation (0%–10%) [8,9]. An important limitation of ESD is that complete resection relies solely on endoscopic observation; it is difficult to achieve complete histological resection of tumors related to an inability to obtain sufficient margins during endoscopy [8]. In a study assessing endoscopic mucosal resection of early gastric cancers nearly 17% had incomplete resection [10].

EALs for gastric tumors was first introduced in 1999 by Aogi et al. [11]. Initially, endoscopy was used primarily as an “extra set of eyes” allowing better localizing of the tumor [11–21]; recent studies have combined ESD with LS with excellent results [22,23]. Benefits of a combined approach include better localization of the tumor, determining the best laparoscopic approach, verifying complete resection, ensuring adequate margins, and assuring leuk-proof suture lines [22,23]. In the present case, endoscopy confirmed that the tumor was localized to the gastric body in a location accessible for laparoscopic removal. Biopsy forceps were then used to show the surgeons the location by pressing on the tumor and “tenting” the gastric wall for laparoscopic visualization. Endoscopic visualization during placement of the gastric stapler confirmed that the entire tumor was captured and that the gastric lumen would not be impeded once the tumor was removed. Finally, after resection the stomach was gently insufflated to ensure that the suture line was leak-proof.

The finding of a gastric neurofibroma is exceedingly rare, particularly in children. Neurofibromas account for 0.1% of gastric tumors found at autopsy [24] and 0.06% of surgically removed gastric tumors [25]. Of individuals with gastrointestinal neurofibromas, 15%–17% have neurofibromatosis (von Recklinghausen disease), while the remainder (83%–85%) have an isolated neurofibroma [26,27]. It has been estimated that 25% of individuals with neurofibromatosis develop gastrointestinal neurofibromas [28]. The distribution of neurofibromas differs between individuals with neurofibromatosis and those with isolated neurofibromas. In

2. Discussion

Gastric submucosal tumors in children are a rare occurrence. Studies in adults have shown that these tumors are most commonly gastrointestinal stromal tumors (GIST) or leiomyomas; more rare tumors include leiomyosarcomas, schwannomas, and neurofibromas. Differentiation between these tumors cannot be made solely by endoscopic visualization. Endoscopic ultrasound (EUS) has been used with fair success (85% sensitivity, 86% specificity, 80% overall accuracy) in differentiating GISTs from leiomyomas [1,2]. However, the malignant potential of submucosal tumors cannot be made with certainty by EUS, thus a definitive diagnosis requires pathological confirmation [1]. Standard forceps biopsy is inadequate for obtaining tissue from submucosal tumors as the biopsies typically do not reach beyond the mucosa. While deeper biopsies can be
neurofibromatosis, the most commonly involved site is the jejunum, followed by stomach, ileum, duodenum, and colon [27]. For isolated neurofibromas the ileum is the most commonly involved site, followed by the jejunum, duodenum, and stomach [29]. Within the stomach, the antrum is the most commonly involved site (39%), followed by the posterior wall (16%), lesser curvature (13%), fundus (10%), body (10%), greater curvature (6%), and anterior wall (6%) [26]. Presentation of gastrointestinal neurofibromas depends on where the tumors arise. Most arise from the submucosal myogenic (Auerbach’s) plexus, leading to suberosal lesions, which tend to present with obstruction due to external compression or volvulus [30]. Neurofibromas can also arise from the submucosal (Meissner’s) plexus, which protrude into the lumen. These lesions may be found incidentally at endoscopy, or present with abdominal pain, anemia, gastrointestinal bleeding, or vomiting [26,30]. The definitive diagnosis of neurofibroma is dependent on microscopic examination. Neurofibromas of the gastrointestinal tract are histologically identical to those that occur in extraintestinal locations, showing bundles of proliferating wavy, hyperchromatic spindle cells admixed with collagen, scattered neuritis, and variable degrees of myxoid matrix [31]. The main differential diagnosis is schwannoma, another benign peripheral nerve sheath tumor. Both neurofibromas and schwannomas show some $100$ immunohistochemical reactivity. However, schwannomas are typically encapsulated with a distinctive combination of Antoni A and B growth patterns. Neurofibromas, on the other hand, are usually not encapsulated and show a range of cell types, including Schwann cells and fibroblasts [32]. Other lesions in the differential diagnosis include leiomyoma (a benign smooth muscle tumor typically positive for desmin and other smooth muscle markers) and GIST (a spindle cell tumor recapitulating the interstitial cells of Cajal, typically positive for CD117 and CD34).

3. Conclusion

We report the successful use of EALS for the removal of a submucosal gastric neurofibroma in a child. We present this case to highlight the feasibility of a combined surgical-endoscopic approach to submucosal gastric tumors in children. Also, we provide a literature review of gastric neurofibromas, a rare finding in children.

Conflict of interest
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