J Ped Surg Case Reports 4 (2016) 38–41

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Gastric plexiform fibromyxoma tumor in a child – Case report and review of the literature

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ARTICLE INFO

Article history:
Received 26 October 2015
Received in revised form
31 October 2015
Accepted 3 November 2015

Key words:
Plexiform fibromyxoma
Gastric
Immunohistochemistry

ABSTRACT

Plexiform fibromyxoma tumor (PFT) is an exceedingly rare tumor, particularly in children where only four cases have been reported to date. The patient reported herein presented with abdominal pain and vomiting related to gastric outlet obstruction caused by a large, polypoid PFT. We describe the clinical features, diagnostic evaluation, and surgical treatment of this rare tumor in our patient. Further, we review the literature of FPT to bring attention to this rare gastric tumor to the Pediatric Surgeon.

1. Case report

A 9 year old female presented with a several month history of intermittent abdominal pain described as periumbilical in location, without triggering or relieving factors, and of short duration. Two months prior to presentation she developed associated nausea and vomiting, resulting in a 4.5 kg weight loss. The vomitus was free of coffee-ground material, blood, and bile. She was treated with anti-emetics without benefit and was treated empirically for giardiasis without benefit. The symptoms were increasing in frequency and severity prompting referral to gastroenterology, whence she underwent esophagogastroduodenoscopy (EGD) that demonstrated a large polypoid antral mass; a biopsy showed prominent granulation tissue suggestive for a pyogenic granuloma. She was referred to our institution for further evaluation and treatment.

At initial assessment, the patient's physical exam was unremarkable except for tachycardia and mild epigastric tenderness. Medical and family histories were unremarkable. Admission laboratory studies, including complete blood count, liver panel, and serum electrolytes, were normal except for mild hypochloremia (Cl = 97 mmol/L, normal 98–120 mmol/L) and mild alkalosis (CO_2 = 31 mmol/L, normal 22–29 mmol/L) consistent with her history of vomiting. The patient was admitted, made NPO, and started on intravenous fluids for resuscitation.

Upper gastrointestinal series demonstrated a large mass with a broad base attachment to the posterior wall of the distal stomach and additional findings of high-grade partial gastric outlet obstruction (Fig. 1A). Computed tomography (CT) scan further defined the 4 cm heterogeneously enhancing, predominately intraluminal mass in the distal stomach without findings to suggest external invasion or metastatic disease (Fig. 1B and C).

Following rehydration and correction of electrolyte abnormalities, the patient was taken to the operating room where EGD was performed with surgery in attendance to better assess the tumor prior to surgical intervention and to determine if endoscopic removal was possible. A large, polypoid mass arising from the posterior antral wall was noted to obstruct the pyloric channel (Fig. 2A). The base of the tumor could not be visualized and snaring the mass was deemed too risky. Thus, a laparotomy was then performed providing exposure for anterior gastrotomy, reduction of the polyp from the duodenum, and subsequent resection at the base from the posterior gastric wall.

Pathology demonstrated a plexiform fibromyxoma with extensive surface ulceration (Fig. 2B). Immunohistochemical

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http://dx.doi.org/10.1016/j.epsc.2015.11.002

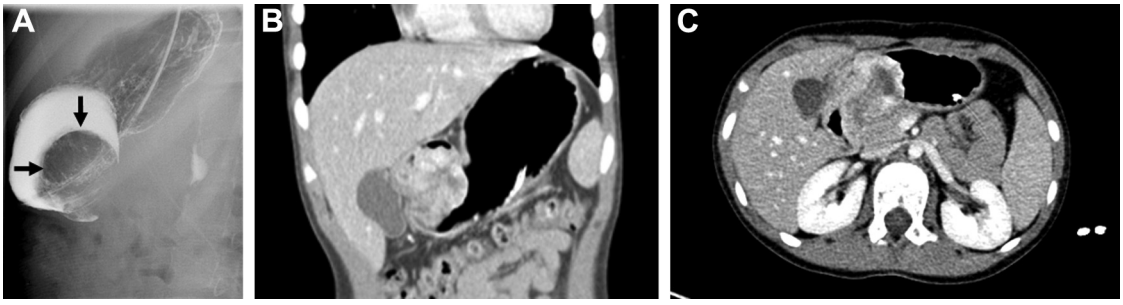

Fig. 1. Radiographic findings. UGI series demonstrated a large mass with a broad base attachment to the posterior wall of the distal stomach resulting in high-grade partial gastric outlet obstruction (A), the edge of the tumor is highlighted with arrows. The tumor was better delineated on CT scan (B, coronal view; C, axial view).

markers further supported the diagnosis with spindle cells staining positive for smooth muscle actin and calponin, and focal staining with desmin and CD10 (a marker for acute lymphoblastic leukemia, but also found in stromal tumors). The lesional cells were negative for antibodies found in GISTs, including DOG1 (discovered on GIST-1, a chloride channel protein), CD117 (C-kit, a tyrosine kinase receptor), and CD34 (a hematopoietic progenitor cell antigen). The tumor cells were also negative for antibodies found in neuronal tumors S-100 (identifies tumors of neuroectodermal origin) and GFAP (glial fibrillary acidic protein). Tumor cells were negative for cytokeratin and ALK1 (activin receptor-like kinase 1). In situ hybridization was negative for EBER (Epstein–Barr encoding region), excluding lymphoma. No acid fast bacilli were noted on special stains.

The patient did well postoperatively with resolution of symptoms and was discharged home on postoperative day 7. She has remained symptom free for 4 months of follow-up and has gained 4.7 kg. Although PFTs are benign and recurrence has not be reported, we plan on a follow-up endoscopy 6 months from the time of surgical removal.

2. Discussion

First describe in 2007 by Takahashi et al., plexiform fibromyxoma tumor (PFT) is a rare mesenchymal tumor with only 32 cases reported to date in the English literature (Table 1) [1–16]. Alternatively referred to as plexiform angiomyxoid tumor, gastric fibromyxoma, and plexiform angiomyxoid myofibroblastic, PFT is the preferred nomenclature by the World Health Organization classification of tumors of the digestive system [17]. The tumors are characterized by a plexiform growth pattern, a myxoid stroma rich in small vessels, and the myofibroblastic nature of the tumor cells [1]. While some tumors may contain cells with fibroblastic or smooth muscle characteristics, myofibroblastic cells are the predominant cell type in the majority of reported tumors [6].

Plexiform fibromyxomas have been reported both in adults and children with an age range from 7 years to 75 years (mean age of 40.3 ± 19.0 years; median age of 42.5 years). Tumors are more commonly reported in adults then in children, with an adult-to-child ratio of approximately 5:1. There is no gender predisposition, with a male-to-female ratio of approximately 1:1 [1–16].

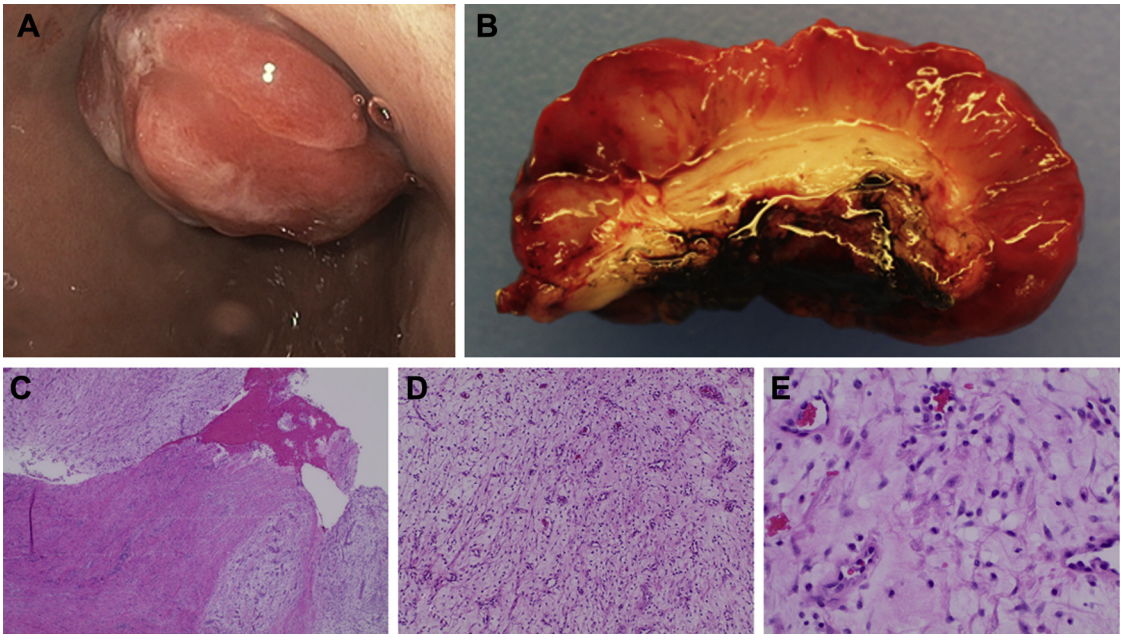

Fig. 2. Endoscopic and pathological features of the tumor. At endoscopy a large, friable, lobulated mass was seen filling the pyloric antrum and obstructing the pylorus (A). The tumor showed extensive surface ulceration (B). Hematoxylin and eosin stained sections of the mass demonstrate classic features of plexiform fibromyxoma. The lesion is arranged in a plexiform growth pattern extending into the gastric wall (C, 4× magnification). Loose stroma separates numerous small vessels (D, 10× magnification) and bland spindled cells (E, 20× magnification).