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

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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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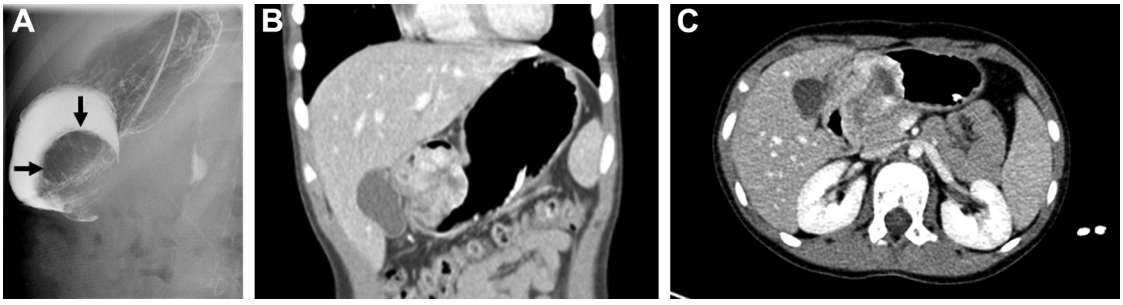

Fig. 1. Radiographic findings. UGI series demonstrated a large mass with a broad base attachment to the posterior wall of the distal stomach resulting in high-grade partial gastric outlet obstruction (A), the edge of the tumor is highlighted with arrows. The tumor was better delineated on CT scan (B,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

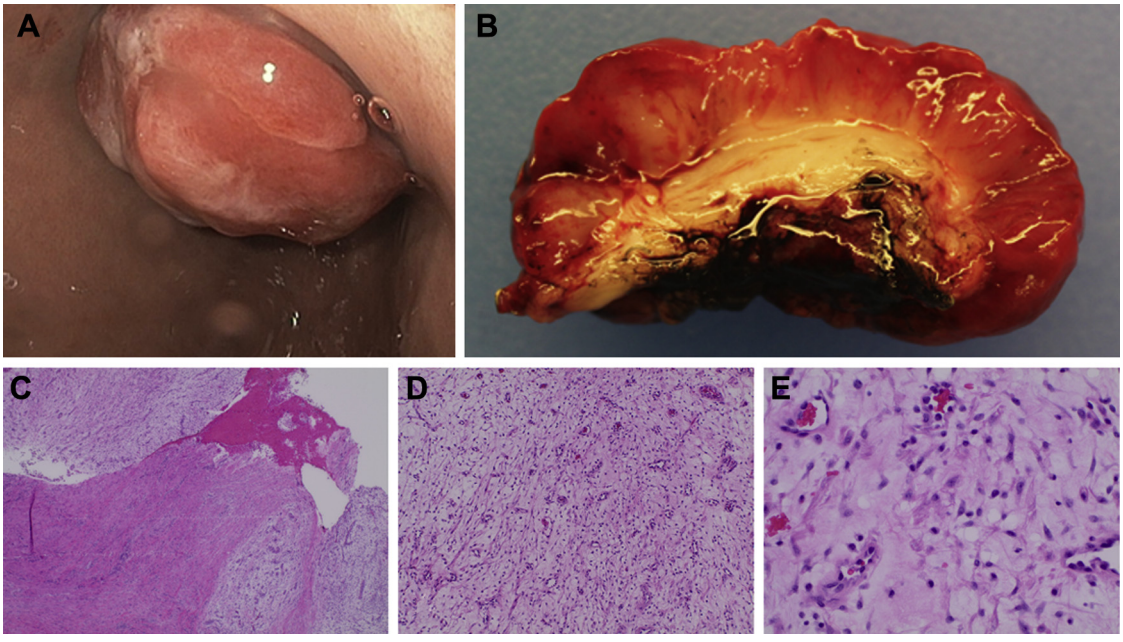

Fig. 2. Endoscopic and pathological features of the tumor. At endoscopy a large, friable, lobulated mass was seen filling the pyloric antrum and obstructing the pylorus (A). The tumor showed extensive surface ulceration (B). Hematoxylin and eosin stained sections of the mass demonstrate classic features 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).

Table 1
Clinical features of reported patients with PMAT.

Ref	Gender	Age (yr)	Symptoms	Location	Ulcerated	Size (cm)*	Treatment	Outcome
1	M	50	Abdominal pain	Antrum	Y	4	DG	nd
	M	68	Incidental	Antrum		4.5	DG	Alive no tumor (12 mo)
2	F	50	Nausea	Antrum	Y	1.9	WR	Alive no tumor (3 mo)
3	M	61	Hematemesis	Antrum	Y	3.7	DG	Alive no tumor (3 mo)
4	F	19	Mass	Antrum		4.5	DG	Alive no tumor (9 mo)
	M	46	UGIB	Antrum	Y	3.5	DG	Alive no tumor (4 mo)
5	F	38	UGIB	Antrum	Y	3	DG	nd
	M	62	Weight loss	Antrum		4	PG	nd
	F	75	Unknown	Antrum	Y	5	STG	Died unknown cause (2 mo)
	F	65	Weight loss, ulcer	Antrum, DB	Y	5	PG	Died unknown cause (14.5 yr)
	M	33	Anemia	Antrum	Y	5.5	DG	Alive no tumor (19.7 yr)
	M	43	UGIB	Antrum	Y	5.5	PG	Alive no tumor (18.4 yr)
	F	56	Unknown	Antrum, DB		5.5	PG	Alive no tumor (19.9 yr)
	M	50	Gastric outlet obstruction, anemia	Antrum, DB		7	DG	Alive no tumor (25.5 yr)
	M	21	Anemia	Antrum, DB	Y	9	Antrectomy	Alive tumor status unknown (22 yr)
	F	16	Hematemesis	Antrum	Y	10	DG	Alive tumor status unknown (3 yr)
	F	30	Gastric ulcer	Antrum	Y	10	DG	Alive tumor status unknown (24 yr)
	F	7	Vomiting, mass	Antrum, DB		15	WR	nd
6	M	23	Abdominal pain, melena	Antrum, DB		14	PG	Alive no tumor (12 mo)
7	F	23	UGIB	Antrum	Y	8	DG	nd
8	F	54	Dyspepsia, vomiting	Fundus	Y	1.5	ER	Alive no tumor (6 mo)
9	M	34	Abdominal pain, mass	Antrum		3.5	DG	nd
10	F	35	Incidental [CT]	Antrum	Y	4	surgery	Alive no tumor (12 mo)
11	M	52	Dyspepsia	Antrum	Y	3.5	WR	Alive no tumor (5 mo)
12	M	47	Asymptomatic	Body	Y	3	WR	Alive no tumor (6 yr)
	F	63	Asymptomatic	Body	Y	2.2	ER	Alive no tumor (1 mo)
13	F	42	Abdominal pain anemia	Antrum	Y	12.9	DG	Alive no tumor (3 wk)
14	M	60	Epigastric pain	Antrum		2	PG	Alive no tumor (12 mo)
15	M	32	Asymptomatic	Antrum		3.4	PG	Alive no tumor (3 yr)
16	F	16	Incidental	Esophagus		nd	surgery	Alive no tumor (14 mo)
	F	11	Anemia	Antrum, DB	Y	3.5	DG	Alive no tumor (15 mo)
Our case	F	9	Gastric outlet obstruction	Antrum	Y	5	PG	Alive no tumor (6 mo)

Size (*) is expressed as the length of the longest axis of the tumor; DB, duodenal bulb; DG, distal gastrectomy; ER, endoscopic resection; nd, no data; PG, partial gastrectomy; STG, sub-total gastrectomy; UGIB, is upper gastrointestinal bleeding (including hematemesis and ulcers leading to melena); WR, wedge resection.

Clinical presentation of PFT varies from an incidental finding to bowel perforation requiring emergency surgical intervention [1,10,12,15,16]. Presenting clinical features were described in 31 of the 32 reported cases. The most commonly reported findings included upper gastrointestinal bleeding (melena and hematemesis) and abdominal pain and dyspepsia, both seen in 23% of patients. Other reported symptoms include anemia, nausea and vomiting, gastric ulcer, and weight loss. Vomiting due to gastric outlet obstruction by the tumor was reported in two patients. PFT occurred as an asymptomatic incidental finding in 20% of patients, including three during screening endoscopy, two by computed tomography for unrelated complaints, and one during cholecystectomy [1,10,12,15,16].

The vast majority of PFT reported have been located in the gastric antrum (88%), at times also involving the duodenal bulb (22%). Case reports have documented involvement of the gastric body (2, 6%) and fundus (1, 3%) [8,12]; the only report of PFT found outside the stomach was an incidental PFT of the esophagus (3%) identified by imaging for an unrelated complaint [16]. Tumors can involve the stomach from the serosa to the submucosa [5]. When mentioned, most PFTs present as a submucosal mass (75%); polypoid lesions account for 20% and isolated serosal lesions account for 5%. Extra-gastric serosal extension is reported in 22% of submucosal tumors highlighting their ability to be transmural; gastric perforation due to tumor extension through the serosa has been reported [1]. The tumors are not encapsulated and may appear as a nodular or granular mass when they extend through the mucosa or serosa. Fully two-thirds of the tumors are ulcerated explaining the high rate of bleeding associated with these lesions. The size of the tumors range from 1.5-cm to 15-cm in longest axis (mean of 5.6 ± 3.5-cm and median of 4.5-cm) [1–16].

The differential diagnosis of PFT contains other tumors that present as a submucosal gastric mass including gastrointestinal stromal tumors (GIST), neuronal tumors (schwannoma and neurofibroma), smooth muscle tumors (leiomyoma and leiomyomsarcoma), and fibroblastic tumors (solitary fibrous tumor, desmoid fibromatosis, and inflammatory fibroid polyp). Microscopic criteria for diagnosis of PFT includes a plexiform growth pattern, proliferation of bland-appearing spindle cells separated by an abundant intracellular myxoid matrix, and a stroma rich in Alcian blue positive, small vessels [1]. Immunohistochemistry is essential for establishing the diagnosis of PFT. Summary of the reported cases shows that PFT are positive for markers of myofibroblastic differentiation (smooth muscle actin [90%] and vimentin [100%]) and negative for markers of GIST tumors (C-kit, CD34, and DOG1) and neuronal tumors (s100). A number of other markers that have been investigated in small numbers, positive results were found for caldesmon and calponin; negative results were found for ALK1, catenin, cytokeratin, epithelial membrane antigen, neuron-specific enolase, and neurofilament. Mixed results were found for desmin (positive in 56%) and CD10 (positive in 20%). When studied, these tumors have low mitotic activity (0–4 mitoses per 50 high power fields) and low Ki-67 index (most <2%, one case had 6%) [1,2,4,6,8,9,12,13,15].

Treatment primarily consists of surgical excision (94%), whereas endoscopic resection has be performed in two cases [8,12]. Surgical technique is dictated by location, size, and depth of the mass. Distal gastrectomy (44%) is the most commonly reported surgery, followed by partial gastrectomy (22%), wedge resection (13%), antrectomy (3%), and subtotal gastrectomy (3%); surgery type was not specified in 9%.

Outcome data were available for 26 patients, 6 were lost to follow-up. Overall time of follow-up ranged from 1 month to 25.5 years (mean 4.8 years, median 1 year). Three patients (12%) died of causes unrelated to the tumor from 2 months to 25.5 years following resection [5], 3 patients (12%) were alive with unknown tumor status from 3 years to 24 years following resection [5], and 20 patients (76%) were alive without tumor recurrence 1 month to 20 years following resection [1–6,8,10–16]. To date there have been no reported cases of metastatic disease resulting from PFT.

3. Conclusion

We report only the fifth plexiform fibromyxoma in a child. Symptoms and signs of PFT are non-specific, the tumor is usually identified with radiologic imaging, and diagnosis based on histological findings. Treatment is surgical; metastasis and recurrence of tumor have not been reported with long-term follow-up.

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