Gastric outlet obstruction caused by a pyloric submucosal fibroepithelial tumour

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ABSTRACT
A 57 year old male patient presented to our hospital with vomiting, epigastric discomfort, and loss of appetite over a 24hr period. Subsequent investigations demonstrated a submucosal lesion causing gastric outlet obstruction. The patients deteriorating condition mandated laparotomy and a pathologic diagnosis was made of a pyloric submucosal fibroepithelial lesion from the resected distal stomach and adherent transverse colon; no malignant features were found. This is to our knowledge the first instance of a fibroepithelial neoplasm occurring within the gastrointestinal tract.

INTRODUCTION
We present a case of gastric outlet obstruction (GOO) caused by a fibroepithelial tumour. This is the first instance of such a tumour being identified in the gastrointestinal tract. Interestingly also, our patient experienced no prodromal symptoms relating to gradual occlusion of the gastric outlet, and presented with an acute picture leading to diagnostic confusion initially.

CASE PRESENTATION
A previously well 57-year-old man presented to our emergency department with a 24hr history of epigastric fullness and discomfort, delayed non-bilious vomiting after oral intake, and anorexia. The patient took no regular medications and consumed 12-14 Units of alcohol per week. Family history was significant for paternal death from gastric adenocarcinoma. Examination revealed normal vital signs and epigastric tenderness. Initial impressions were of gastritis, biliary colic, or peptic ulcer disease (PUD). Intravenous fluids were commenced as the patient was unable to tolerate sips of water. Blood investigations demonstrated a white cell count of $16.1 \times 10^9 \text{L}^{-1}$, C-reactive protein of 247 mg L$^{-1}$ and normal amylase, liver function, and electrolytes. Endoscopy performed the following day revealed a pre-pyloric submucosal lesion with normal overlying mucosa (biopsy proven), with complete obstruction of the pyloric valve – a diagnosis of GOO secondary to a submucosal lesion was made. Ultrasonography that same day showed a heterogeneous mass, again obliterating the gastric outlet. A subsequent computed tomography (CT) scan demonstrated solid and cystic components within the lesion and no associated significant lymphadenopathy (Figure 1). Initial impressions included adenocarcinoma, gastrointestinal stromal tumour, and gastric lymphoma.
An endoscopic ultrasound examination (EUS), with a view to simultaneous fine needle aspiration cytology (FNA) was arranged, to further evaluate the lesion. Our patient’s symptom continued unabated and despite total parenteral nutrition was failing to thrive. Therefore a laparotomy was performed in which a gastric antral mass was identified; with the transverse colon adherent to the stomach on the serosal aspect of the mass. There was no palpable lymphadenopathy. An en-bloc distal gastrectomy and right hemicolecction with primary anastomosis was performed. Macroscopic examination of the specimen revealed a 9 x 8cm section of the stomach stapled along two margins, with a 30cm section of colon, including the caecum and appendix, attached. At the site of adhesion a 7cm mass was felt. On opening the lumen of both the transverse colon and stomach no lesion or ulceration was seen; although there was significant bulging of the gastric mucosa overlying the mass. Upon slicing a solid white tumour was seen to arise from the wall of the stomach, approximately 3cm from the nearest resection margin, with a well defined edge (Figure 2).

Microscopic analysis of sections from the stomach revealed a predominantly unremarkable mucosa, with some areas of intestinal metaplasia. Just beneath the muscularis mucosa, a nodular, plexiform tumour was seen. This consisted predominantly of epithelioid, spindle shaped cells set in a collagenous stroma containing thin walled vessels (Figure 3).

In places this stroma was rather hyalinised. The tumour...
also contained cleft-like spaces lined by gastric and intestinal type epithelium, seen more
towards the gastric side of the tumour nodules (Figure 4). This epithelium was in at least one
place continuous, with the normal gastric surface mucosa which was seen to dip down into
muscularis mucosa.

Immunohistochemical analysis revealed the stromal
cOMPONENT to be strongly smooth muscle actin (SMA) positive, with patchy, weak positivity
for vimentin and caldesmon. The tumour cells were negative for cluster of differentiation (CD)
117, CD34, CD99, CD68, S100, Activin receptor-like kinase-1, MNF116, and Human
Melanoma Black -45. The epithelial component showed variable positivity, in different areas,
for cytokeratin (CK)-7 and CK-20. There was no cytological atypia nor any features of
malignancy in either the epithelial or spindle cell elements. Sections from the adherent colon
showed unremarkable mucosa. The final diagnosis in this case, was that of a fibroepithelial
neoplasm. Our patient experienced a protracted recovery due to a post-operative pneumonia
and episode of haemorrhagic gastritis, but was eventually discharged home and remains well
to this date.

DISCUSSION

Considered separately, the epithelial component could not be explained by a developmental
cyst or reduplication as an associated smooth muscle component was not seen; and the
stromal elements did not fit with any of the recognised gastrointestinal mesenchymal
tumours. The only logical way to explain the findings in this case was to regard both the
stromal and epithelial components as part of the same process, and make a diagnosis of a
fibroepithelial lesion of the gastrointestinal tract. Fibroepithelial neoplasias are biphasic
lesions, consisting of both a stromal and epithelial components from two separate germ lines
– endoderm and mesoderm. They are most commonly seen in the breast as the Phyllodes
tumour (1). Their behaviour ranges from benign to malignant, and quantitative morphologic
criteria – including mitotic rate per 10 high-power fields, stromal cellularity, nuclear size,
stromal overgrowth, and the largest and smallest stromal-epithelial surface area ratios - exist
to stratify lesions accordingly (2). In our case there was no pathologic evidence of
malignancy. Gastric submucosal lesions are usually evident after endoscopic investigation,
which is in turn regularly performed due to the symptom profile of these lesions. In the case of
a submucosal gastric lesion, EUS with FNA of the lesion is the definitive investigation. This
has been shown to be the most reliable in determining whether a lesion is benign or
malignant, assessing the suitability of surgical versus endoscopic resection, and can also be
used to survey uncertain lesions (3). The addition of FNA to EUS is reported at achieving an
accurate tissue diagnosis of submucosal lesions in 89% of cases (4). CT scanning and
conventional transabdominal ultrasound can provide supporting information. Definitive management should be planned in accordance with the patient’s symptoms and tumour in mind. Resection can be carried out successfully laparoscopically, as shown for GIST’s (5), or by laparotomy in difficult or uncertain cases.

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