Gastric plexiform angiomyxoid myofibroblastic tumour

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Abstract

Plexiform angiomyxoid myofibroblastic tumour (PAMT) of the stomach is a rare mesenchymal tumour predominantly involving the gastric antral region. There has been continued dispute about its nomenclature with other terminologies used for this tumour as plexiform angiomyxoid tumour, plexiform fibromyxoma. Pathologically, this tumour exhibit distinct morphological and immunophentypic features which aid in differentiating it from a wide range of differential diagnoses list. Here, in this paper, we present a case of PAMT with detailed discussion about its clinical profile, radiologic, and pathologic features including its histogenesis and differential diagnoses.

Keywords gastric; GIST; mesenchymal tumour; myofibroblastic; plexiform

Plexiform angiomyxoid myofibroblastic tumour (PAMT) of the stomach is a rare mesenchymal tumour with 41 cases being described in the English literature so far.[1–3] This entity was first described by Takahashi et al.[4] in 2007. Since then a number of papers have been published describing the unique clinico-pathological features of this tumour. There has been a constant dispute about the terminology used for this tumour. Yoshida et al.[5] also reported two cases of similar tumors in 2008, and they described it as "plexiform angiomyxoid tumour" while Miettenen et al.[6] called it "Plexiform fibromyxoma", the same name adopted by WHO 2010, under the section "mesenchymal tumors of the stomach".[7] In this paper we would discuss the unique clinical profile, radiological, and pathological features of this tumour including its histogenesis and differential diagnoses with a little discussion about the nomenclature.

Case report

A 40 year old female on treatment for mood swings presented with dyspepsia. Upper GI endoscopy performed showed a submucosal polypoid mass in the gastric antrum. CT abdomen revealed an exophytic mass on the serosal aspect of the antral portion of stomach measuring 6.1 × 8.7 cm.

Distal gastrectomy was performed which on gross examination revealed an exophytic, non-encapsulated and multicystic tumour in the antral region along the posterior wall of greater curvature. The tumour measured 9.5 × 6.5 cm, with a glistening cut surface. The mucosa overlying the tumour appeared puckered and granular (Figure 1).

Microscopically, the tumour appeared multinodular, dissecting through the muscularis layer onto the serosa (Figure 2a and b). The mucosa was uninvolved. The tumour was composed of bland spindle shaped cells embedded in richly vascular myxoid background (Figure 2c and d). Numerous mast cells were seen dispersed in the stroma. Tumor necrosis, mitoses or cellular atypia was not observed. Lymphovascular permeation was not present.

Immunohistochemistry revealed positive staining for smooth muscle actin (SMA), (Figure 2e), CD10 and vimentin, and the tumour cells were negative for desmin and S100. CD117 highlighted the mast cells but was negative in the tumour cells. (Figure 2f) Ki67 proliferation index was very low at 2%. A diagnosis of Plexiform Fibromyxoma was made on the basis of morphological and phenotypical features.

Discussion

Clinical findings

Gastrointestinal stromal tumour (GIST) is the most common primary mesenchymal tumour of the stomach accounting for nearly 60% of the GIST cases of the entire gastrointestinal tract.[8] Plexiform angiomyxoid myofibroblastic tumour (PAMT) is a recently described mesenchymal tumour of the stomach with an estimated frequency of 1 in 150 compared to that of gastric GIST.[6]

PAMT classically occurs in the antro-pyloric region of the stomach with a wide range of age distribution (7–75 years) with a mean age group of 43 years.[9] Male and females are equally affected.[9] The presenting clinical features are related to the either the mass effect i.e., gastric outlet obstruction such as epigastric pain, nausea, emesis, weight loss or due to ulceration of the overlying mucosa. Such patients present with upper GI bleed and report black tarry stools. One of the cases reported presented

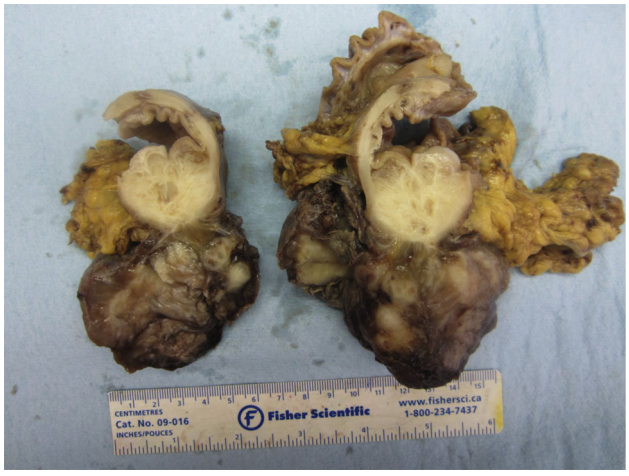

Figure 1 Gross photograph showing cut surface of tumor, with gelatinous areas admixed with foci of cystic degeneration.

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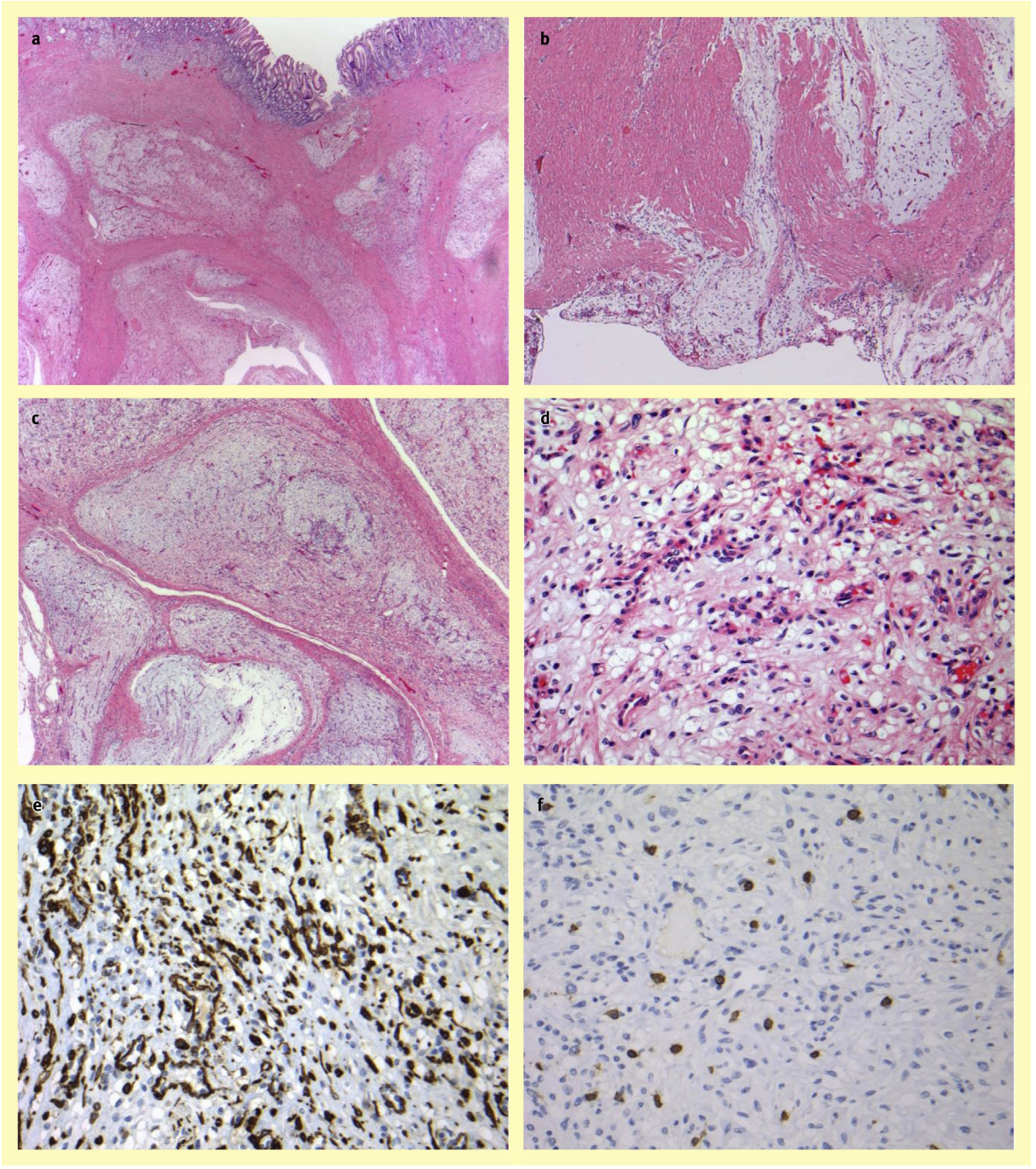

Figure 2 Microscopic images of Plexiform fibromyxoma. (**a** and **b**). Low power (×2.5) magnification: Tumor nodules dissecting through the muscularis layer onto the serosa. (**c**). Tumor nodule with thin vasculature and myxoid areas. (**d**). High power magnification (20×) Bland tumor cells in a richly vascular myxoid stroma. (**e**). Positive staining for SMA in tumor cells and interspersed capillaries. (**f**). CD117 highlighting the mast cells and negative in tumor cells.

with acute abdominal pain due to perforation[4] and one was detected incidentally during cholecystectomy.[4]

Gastroscopy shows a sub-mucosal elevated polypoid lesion in the antrum often associated with overlying central ulceration. CT findings may suggest the hypervascular nature of this tumour with localisation and any additional associated features. One of the cases described by Lee et al.[10] demonstrated contrast enhancing mass in the antrum with fistula formation.

Pathologic features

Grossly, the tumour appears as an elevated or polypoidal mass lesion involving the antral region of the stomach, however, cases has been reported involving the fundus,[11] and body of stomach as well as esophagus.[1] Involvement of the pylorus or the duodenal bulb can be seen in approximately one third of the cases.[9] The tumour size ranges from 1.9 cm to 15.0 cm, with a mean value of 6.3 cm[9]. The cut surface shows an unencapsulated tumour mass involving the sub-mucosal and muscularis layer