CASE REPORT

Plexiform angiomyxoid myofibroblastic tumour of the duodenum: a rare entity

Niladri Banerjee, Shahana Gupta, Suvashis Dash, Shibajyoti Ghosh

Department of Surgery,
Medical College, Kolkata,
Kolkata, West Bengal, India

Correspondence to
Dr Shahana Gupta,
shahanagupta@yahoo.co.in

Accepted 8 July 2015

SUMMARY

Plexiform angiomyxoid myofibroblastic tumour (PAMT) has recently emerged as a new entity among gastrointestinal mesenchymal tumours. All of the 27 cases reported until now originated from the stomach. We report the first case of a duodenal PAMT arising from the first part of the duodenum in a 19-year-old woman presenting with upper abdominal pain and an abdominal lump.

BACKGROUND

Although gastrointestinal stromal tumour (GIST) is the most commonly encountered gastrointestinal (GI) neoplasm of mesenchymal origin,¹ plexiform angiomyxoid myofibroblastic tumour (PAMT) has emerged as a new entity among GI mesenchymal tumours in recent years. The first report was from Takahashi *et al* in 2007.² Histologically, the tumour reveals bland spindle cells arranged in a plexiform nodular growth pattern in a myxoid or fibroblastic stroma.² The PAMT cases reported until now have been located in the gastric antrum.^{2–10} We report a case of PAMT arising from the duodenum (D1), the first such report in the English literature.

CASE PRESENTATION

A 19-year-old woman presented to the Outpatients' Department of Surgery, Medical College, Kolkata, India, with dull upper abdominal pain for 1 month, along with a lump in the right hypochondrium. Abdominal examination revealed a 10 cm×7 cm mobile, firm, non-tender, intraperitoneal mass in the right hypochondrium extending to the epigastrium, with a smooth surface and rounded margins, the upper margin being indistinct.

INVESTIGATIONS

Ultrasonography of the abdomen revealed a 13.5 cm×8.5 cm well-defined solid-cystic lesion below the liver, pushing the pancreas posteriorly, suggestive of GIST. On upper GI endoscopy, there was evidence of a bulge in the posterior wall of the duodenum (D1), with surface ulceration, suggestive of duodenal GIST. Endoscopic biopsy suggested a benign mesenchymal tumour. Contrast-enhanced CT scan suggested a large solid/cystic SOL (13.8 cm×8.6 cm) related to the first part of the duodenum (figure 1).

TREATMENT

On exploratory laparotomy using a Chevron incision, a glistening, well-defined 13.5 cm×8.5 cm tumour was found arising from the posterior wall of the duodenum (D1), free from the stomach and pancreas (figure 2). Distal gastrectomy, proximal

Figure 1 Contrast-enhanced CT of the abdomen showing 13.8 cm×8.6 cm solid/cystic SOL related to the duodenum.

duodenectomy and Billroth II gastrojejunostomy were performed. On gross examination, the specimen consisted of a well-circumscribed tumour measuring 13.8 cm×8.6 cm adherent to the posterior wall of the duodenum. The cut surface was greyish and fleshy, with cystic and solid components (figure 3).

Figure 2 Intraoperative picture.

Figure 3 Cut section of the tumour.

To cite: Banerjee N, Gupta S, Dash S, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-210004

Figure 4 Histopathology showing nodular plexiform growth pattern (x4) (inset showing the interlacing fascicles of bland spindle cells in fibromyxoid stroma (x10)).

Figure 5 Immunohistochemistry showing PgR (prostaglandin receptor) positivity.

Histopathological examination showed the tumour to be sub-mucosal, with a multinodular, plexiform growth pattern. The nodules were composed of interlacing fascicles of plump spindle cells with bland nuclei separated by a fibromyxoid stroma. Extensive areas of myxoid degeneration and a plexiform capillary

pattern were compatible with a gastrointestinal mesenchymal tumour (figure 4). The tumour was confined to the duodenum, not extending to the stomach, with no lymph nodal metastasis. Immunohistochemistry showed tumour positivity for prostaglandin receptor (PgR; figure 5) and immunonegativity for SMA (smooth muscle actin), MSA, CD10, Desmin, Calponin, c-kit, DOG1, S-100 protein, CD34, HMB45 and Melan A (figure 6).

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative recovery. There was no evidence of recurrence or metastasis on follow-up imaging for 6 months after surgery.

DISCUSSION

PAMT is a rare tumour of mesenchymal origin, previously known as angiomyxoid tumour, plexiform angiomyxoma or myxofibroma.¹¹ The PAMT:GIST frequency ratio is less than 1:150.³ The age of presentation varies from 7 to 75 years, with a slight female predilection (male:female=13:15). All 27 cases reported in the literature until now originated from the gastric antrum.^{2-14 15} Clinical presentation ranged from upper gastrointestinal haemorrhage (29%) and epigastric pain (18%), to anaemia, abdominal mass and weight loss (11%). Five cases were diagnosed incidentally.^{11 15} The case described in this report is the only one arising from the duodenum (D1), presenting with abdominal pain and lump. Endoscopically, mucosal ulcerations were found in all but 2 of the 27 reported cases. The case we report also had mucosal ulceration on endoscopy. The tumour sizes reported until now varied widely from 1.5 to 14 cm (mean: 6.5 cm); that of the case we report is near the higher end of this range (13.8 cm×8.6 cm). Histologically, PAMT has been described as a proliferation of bland spindle cells along with myofibroblastic differentiation.² Immunonegativity to c-kit and CD34, and absence of c-kit and PDGFA gene mutations, characteristically differentiate PAMT from GIST. Differential diagnosis such as myxoid variant of GIST, inflammatory fibroid polyp and solitary fibrous tumour can be excluded if there is absence of the distinct plexiform growth pattern seen in PAMT.¹¹ Plexiform growth pattern is also seen in other mesenchymal tumours such as plexiform leiomyoma or nerve sheath tumours (schwannoma, neurofibroma and perineuroma). Plexiform leiomyoma has a characteristic differentiation of smooth muscle, such as diffuse expression of desmin and actin. Negativity to neural and perineural markers, such as S-100 and

Figure 6 Immunohistochemistry showing immunonegativity for CD10, Desmin, Calponin, c-kit, DOG1 and CD34.

EMA/Claudin-1, help differentiate PAMT from nerve sheath tumours. Low-grade endometrial stromal sarcoma (ESS) is another differential diagnosis, particularly in female patients.^{6,9} In the case we report, though the PAMT cells expressed PgR, hormone receptors such as those of oestrogen, were absent, so were CD10 and CD34. This ruled out a diagnosis of ESS. PAMT is a relatively new entity with limited follow-up data on recurrence. The short-term prognosis seems to be good with no recurrence or metastasis reported until now. All these, along with histological features such as bland nuclear morphology and low mitotic index, point towards a benign diagnosis. However, because of a single reported case of vascular invasion,⁴ one cannot exclude the possibility of a metastatic PAMT.

Learning points

- ▶ Plexiform angiofibromatous tumour (PAMT) can occur in the stomach as well as in the duodenum.
- ▶ PAMT should be differentiated from gastrointestinal stromal tumour (GIST), as the management strategies of these two conditions differ in respect to adjuvant therapy (imatinib in c-kit positive GIST, not in PAMT).
- ▶ The prognosis seems to be good with the limited follow-up data.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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