

immunohistochemical staining (Fig. 2). Based on histological and immunohistochemistry findings, we made the final pathological diagnosis of PEComa in the duodenum. The patient was alive and well without evidence of recurrence or metastasis 6 months after tumour resection.

Gastrointestinal PEComas are very rare. To our knowledge, only 72 cases (including our current case) of gastrointestinal PEComa have been reported in the English literature.^{3,6–8} The most common location of gastrointestinal PEComas is the colon ($n = 32, 43.8\%$), followed by the small intestine ($n = 23, 31.5\%$), rectum ($n = 7, 9.6\%$), stomach ($n = 4, 5.5\%$), caecum ($n = 4, 5.5\%$), appendix ($n = 1$), gallbladder ($n = 1$), and omentum ($n = 1$) (Table 1). Of the patients, 43 were females and 29 were males, and the ratio of female to male was 1.5:1. The age at diagnosis ranged from 5.5 to 71 years. The median age was 54.5 years. The median tumour size was 4.5 cm, with a range of 0.8 to 22 cm. Mhanna *et al.* first reported PEComa of the duodenum in a 12-year-old boy with a history of neuroblastoma.⁸ To date, only five cases (including our case) of PEComa of the duodenum have been reported in the English literature.^{3,7,9} Recently, we reported one case of malignant PEComa of the kidney with rare pulmonary and ileum metastases.²

The diagnosis of PEComa depends on the distinctive pathological features and immunohistochemistry staining. Histologically, the tumour usually appears in a haphazard pattern around a vascular lumen. Tumour cells surrounding the vessels are typically epithelioid cells with abundant granular eosinophilic or clear cytoplasm, round nuclei with small nucleoli. Some PEComas are dominated by spindle tumour cells. The tumour is highly vascular with thin-walled vessels that blend with the neoplastic cells. Tumour cells are commonly positive for HMB45, Melan-A, SMA, and desmin by immunohistochemistry staining. About 10% of tumours show focal expression of S-100 protein and strong nuclear staining of TFE3. In our case, the tumour cells were positive for HMB45, Melan-A, SMA, desmin, and H-caldesmon, while S-100 protein and TFE3 were negative.

PEComa of the duodenum should differentiate from gastrointestinal stromal tumour (GIST), leiomyoma, leiomyosarcoma, malignant melanoma, and spindle cell carcinoma. GIST is the most common tumour of the gastrointestinal tract and has diverse morphological features, similar to PEComa. However, the tumour cells of GIST are positive for CD117, CD34, and DOG-1, but negative for HMB45 and Melan-A. PEComa is difficult to differentiate from epithelioid smooth muscle tumours, but the tumour cells of leiomyoma and leiomyosarcoma are short of HMB45 and Melan-A expression. Malignant melanoma presents HMB45, Melan-A, and S-100 protein positivity, which is similar to PEComa. However, tumour cells are atypical and negative for SMA and desmin in malignant melanoma. Spindle cell carcinoma of the duodenum is very rare. Carcinoma cells are usually arranged in nests and are positive for CK and CEA, but negative for HMB45, Melan-A, S-100 protein, SMA, and desmin by immunohistochemical staining.

PEComa is rare and may pose differential diagnostic difficulty for both clinicians and pathologists, particularly if the tumour is encountered from needle biopsy. Berger *et al.* analysed 85 cases of renal angiomyolipoma (AML) and found the diagnosis of AML was ignored pre-operatively in 62 patients (73%). The small size of the AML, low proportion of fat, and male sex were significantly associated with misdiagnosis of AML.⁹ Agaimy *et al.* studied six cases of hepatic AML and found the initial diagnosis or impression

either at frozen section or during permanent histological evaluation was misleading in five cases, which included vascular lesion, focal fatty change, myelolipoma, hepatocellular tumour, and rhabdoid neoplasm. They suggest that awareness of the diverse morphological spectrum of PEComa in liver is necessary to avoid misdiagnosis as hepatocellular carcinoma, metastatic melanoma or other malignant neoplasms.⁴ The main reason for misdiagnosis in our case was the very rare site of PEComa and the limited tissue of endoscopic needle biopsy.

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Giant cell tumour of the gallbladder can mimic undifferentiated/anaplastic carcinoma clinically and pathologically

Sir,

Tumours rich in giant cells are extremely rare in the gallbladder, biliary tree and pancreas, and most cases are in fact undifferentiated/anaplastic sarcomatoid carcinomas with infiltrative growth and an aggressive clinical course.^{1–3} These latter tumours are usually associated with poor survival and extensive

local and metastatic disease at the time of diagnosis. In contrast, giant cell tumours of the gallbladder (analogous to giant cell tumours of the bone) have been recently recognised and attributed to an indolent clinical course. These tumours have a unique immunophenotype that separates them from the potentially fatal undifferentiated/anaplastic carcinomas.⁴

A 56-year-old woman presented to the emergency department with a relatively short history of upper abdominal pain, abnormal liver function tests and raised C-reactive protein (CRP). The patient had a past medical history of multinodular

goitre. An ultrasound scan demonstrated a 52 mm polypoid mass with increased vascularity in the gallbladder, almost entirely occluding the lumen. Computed tomography (CT) scan and magnetic resonance imaging (MRI) confirmed a mass, 38 mm in diameter, which was considered to be highly suspicious of a gallbladder carcinoma and suggested possible infiltration through the gallbladder wall into segment 5 of the liver (Fig. 1A). There was no locoregional lymphadenopathy or metastatic disease on CT or positron emission tomography (PET) scan (Fig. 1B). The patient underwent an uneventful

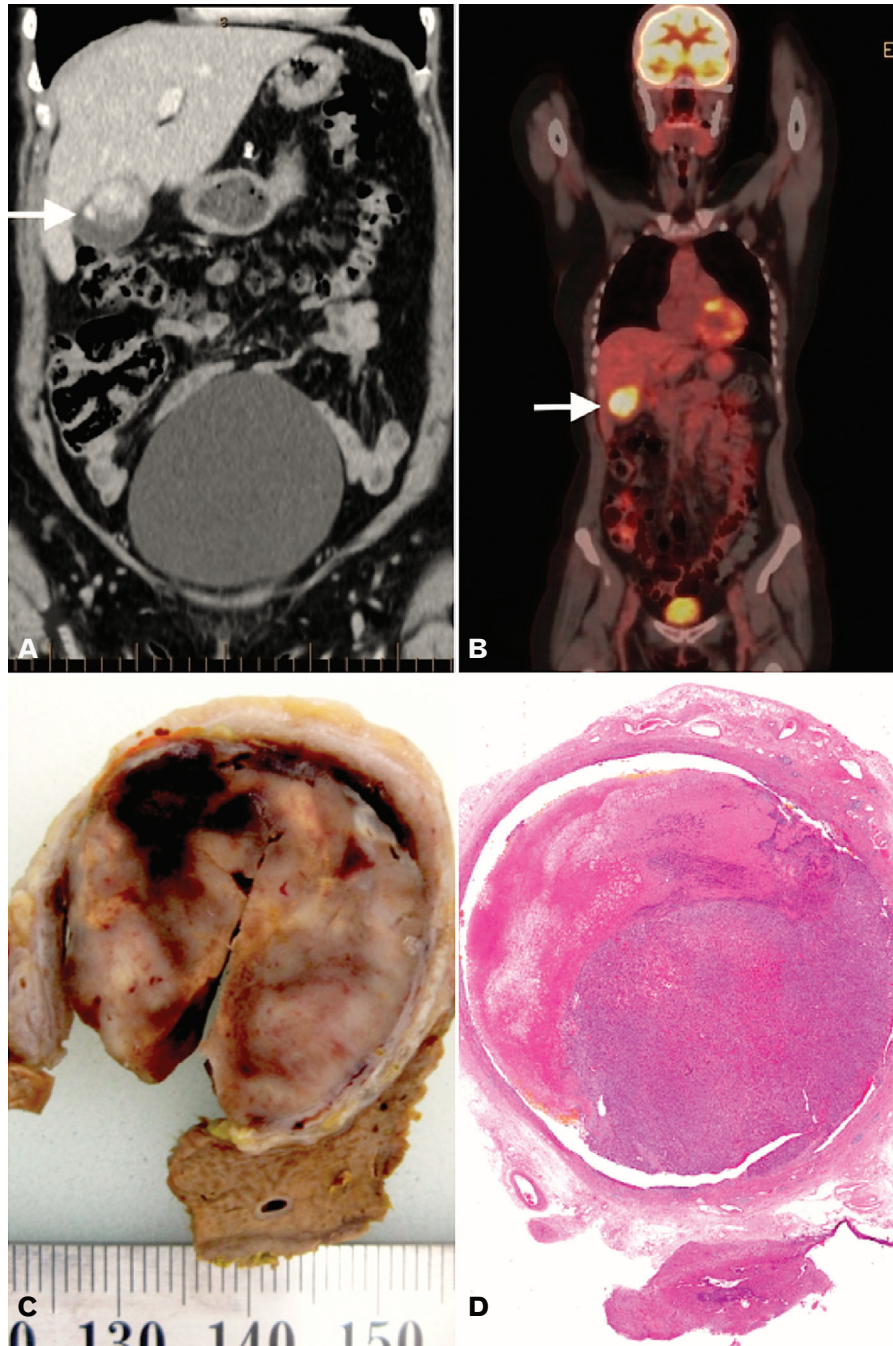


Fig. 1 (A) Computed tomography (CT) scan showing a hypervascular lobulated mass in the gallbladder (white arrow) with no locoregional abdominal lymphadenopathy. (B) PET scan revealing high metabolic nature (FDG-avidity) of the mass in the gallbladder with no metabolic activity in the adjacent liver or regional/distant lymph nodes. (C) On macroscopic examination, the mass occupied the entire lumen with focal attachment to the mucosal surface. The cut surface of the mass was heterogeneous. (D) The corresponding microscopic section at scanning magnification confirmed the mucosal confinement of the mass. Notice the intact gallbladder wall and attached portion of the liver (H&E).

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