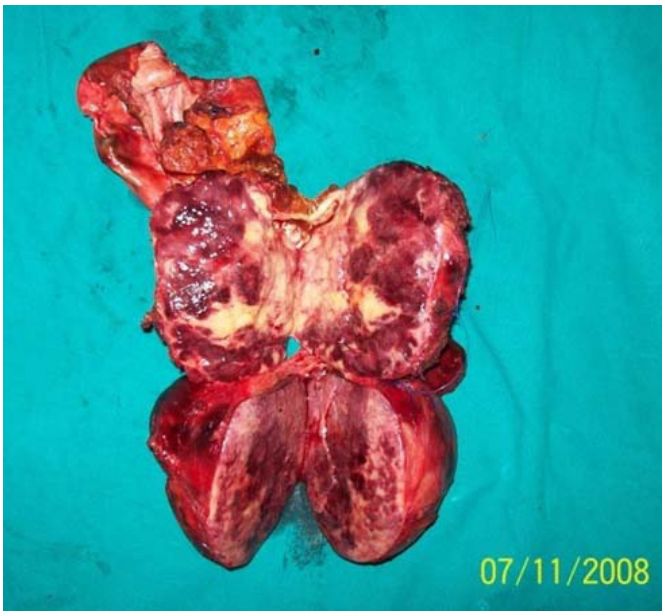

Figure 3: Cut-open view of the dumb-bell-shaped excised mass with portion of the dilated common hepatic duct.

aorta, in the organ of Zuckerkandl, and in the pelvis.[3] So far paragangliomas have been reported in a variety of unusual locations, including the liver, gallbladder, oesophagus, periampullary area, pelvis and urinary bladder. Biliary system paragangliomas are predominantly seen in women and so far only three cases of paraganglioma arising from the hepatic ducts have been reported.[3]

Paraganglioma has unique pathological features wherein the tumour has well-defined nests of round or polygonal epitheloid cells arranged in a typical Zellballen pattern and surrounded by sustentacular cells in a delicate, fibrovascular stroma.[3,4] The tumour in our patient measured 15 cm × 7 cm and exhibited the above classic histological appearance.

The clinical manifestation depends on the functionality of the tumour. Generally, functional tumours are chromaffin-positive and arise mostly from the adrenal medulla or organ of Zuckerhandl and along the peripheral sympathetic system.[2] However, most chromaffin-positive, retroperitoneal tumours appear to be functional as well. Symptoms are related to excess catecholamine production by the tumours, such as norepinephrine, epinephrine and dopamine. Non-functioning tumours are chromaffin-negative and usually arise from the head and neck or mediastinum. As a rule, biliary tract paragangliomas are non-functional.[1,3] All the gallbladder and hepatic duct paragangliomas reported so far were non-functional and presented with symptoms referred to biliary tract such as abdominal pain or obstructive jaundice.[3] Our patient did not have symptoms of catecholamine excess and presented only with obstructive jaundice.

Among the available tests, plasma-free metanephrine is considered the most sensitive to confirm excess catecholamine production.[3] Preoperative suspicion of paragangliomas mandates biochemical screening and prevention for perioperative hypertensive crisis. Imaging characteristics are not unique. They are usually large tumours with areas of haemorrhage or necrosis and heterogeneous enhancement after contrast injection.[3]

PRAKASH KURUMBOOR[1],
KAMALESH NP[1]
MATHEW PHILIP[2]

Correspondence: Dr Prakash K.
Department of GI Surgery[1] and Gastroenterology[2] PVS Institute of Digestive Diseases, Kochi, India. 682017.
Email: drkprakash@vsnl.com

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Perivascular epithelioid cell tumour of the duodenum

Introduction

Perivascular epithelioid cell tumour (PEComa) is a rare neoplasm. It is composed chiefly of HMB-45-positive epithelioid cells and is rich in vascular organoid architecture.

Case report

A 25-year-old man presented with melaena for 4 days. There was no history of abdominal pain, vomiting, jaundice or altered bowel habit. He was pale. Other physical examinations were normal. Laboratory investigations included packed cell volume 23%, marginally high C-reactive protein (30.9 mg/L), normal coagulation profile and liver function tests (LFTs).

Oesophagogastroduodenoscopy showed nodular, ulcerated and friable mucosa in the proximal second part of the

duodenum, suggestive of infiltration. Duodenal mucosal biopsy was reported as spindle cell proliferation, with the possibility of an underlying smooth muscle tumour. The tumour cells had displayed positive staining for smooth muscle actin (SMA), and occasional cells were positive for CD 117, S100 and CD34. Contrast-enhanced CT (CECT) scan showed a 7 cm × 5 cm, heterogeneously enhancing mass in the pancreatico-duodenal groove with thickening of the duodenal wall (**Figure 1**). The mass displaced the pancreatic head without obvious infiltration. The patient underwent pancreaticoduodenectomy with radiological access loop creation for hepaticojejunostomy. A solid cystic mass was seen in the region of the pancreatic head during the operation.

Grossly, it was a 5 cm circumscribed tumour in the wall of the duodenum, at the level of the papilla. There was no invasion of the adjacent pancreas. The lesion had a creamy white, partly necrotic cut surface (**Figure 2**). Microscopically, the lesion was found to originate in the submucosal layer of the duodenal wall with areas of mucosal ulceration and necrosis. Tumour cells were monomorphic, arranged as lobules and sheets of large polygonal cells with abundant granular eosinophilic to focally clear cytoplasm and large centrally placed vesicular nuclei (**Figure 3**). Prominent central eosinophilic nucleoli were present in the foci. The lobules were separated by sinusoidal vessels. The tumour was demarcated from the pancreas by fibrous capsule and there were foci of calcification. Lymph nodes were reactive. Mitotic activity was 1 in 50 high power fields. The phenotype of tumour cell was verified by immunohistochemistry. Cells were diffusely positive for HMB-45 antibody (melanocytic marker) (**Figure 3 inset**), and stained equivocally for neuron-specific enolase (NSE). Scattered

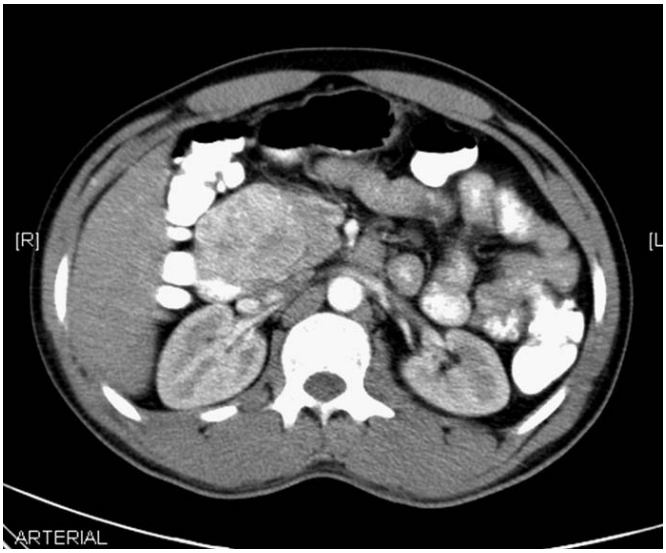

Figure 1: CECT abdomen showing a circumscribed, heterogeneously enhancing mass in the pancreaticoduodenal groove.

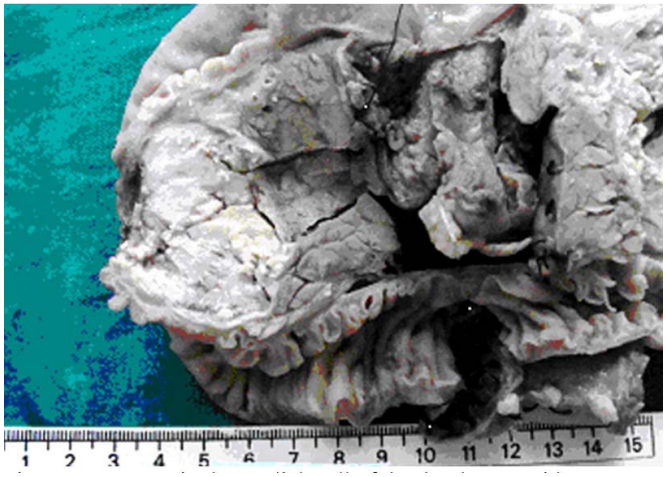

Figure 2: Tumour in the medial wall of the duodenum with creamy white, partly necrotic cut surface.

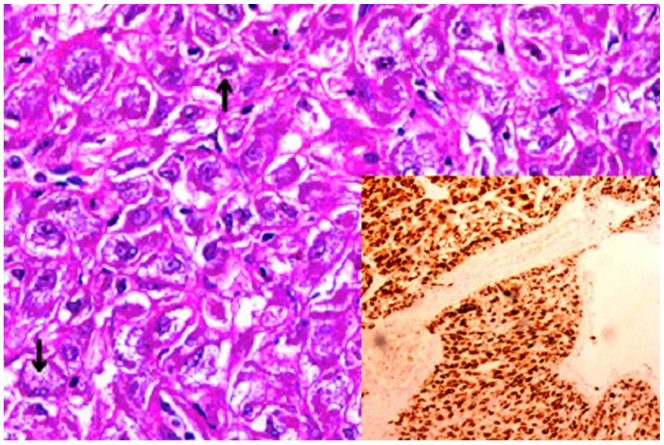

Figure 3: Tumour cells with abundant granular eosinophilic (black arrow) to focally clear cytoplasm and large centrally placed vesicular nuclei (hematoxylin and eosin, 400X magnification). Diffuse positive staining with HMB-45 antibody (inset).

tumour cells were positive for desmin and SMA. Immunostaining for CD34, S100, synaptophysin, CD117, cytokeratin, alpha-foetoprotein (AFP) and placental alkaline phosphatase (PLAP) were negative. The pathological features were consistent with PEComa. The patient was asymptomatic with normal CT scan and LFT at 6 months of follow-up.

Discussion

PEComa is composed of perivascular epithelioid cells (PECs) with myomelanocytic differentiation and strong immunoreactivity for HMB-45 antibody. It is a rare but discretely-defined neoplasm.[1] These tumours may arise in any anatomical location as PECs do not have a normal anatomic homologue. PEComa and related tumours of the duodenum are exceptionally rare, with previously reported literature limited to three case reports—one paediatric patient with PEComa and

two adult patients with angiomyolipoma.[2–4] Gastrointestinal PEComa shows a marked preponderance in women and one-third of the cases occur in the paediatric age group.[1] These tumours may be associated with neuroblastoma and tuberous sclerosis.[2,5]

PEComa seems to have a similar biological behaviour to GIST.[6] The negative reactivity for CD117 and CD34 went against the diagnosis of epithelioid GIST and KIT-negative GIST. Endocrine tumours are usually positive for NSE and synaptophysin. Cytokeratin was negative, excluding the diagnosis of carcinoma. Germ cell tumour was ruled out by the negative reaction for AFP and PLAP. Malignant melanoma and granular cell tumour could not be considered because S100 was negative. Smooth muscle tumour with epithelioid morphology was considered as a differential diagnosis due to the positivity of few cells for desmin and SMA. However, there were no spindled foci with perinuclear vacuoles or cigar-shaped nuclei. In our patient, the morphology of the tumour showed abundant granular eosinophilic to focally clear cytoplasm, delicate sinusoidal network and presence of cytoplasmic glycogen; these were in favour of PEComa.[7] The immunohistochemical reaction with diffuse strong positivity for HMB-45 antibody, and focal positive staining for SMA, highlighted the myomelanocytic nature of this tumour.[5,7]

SUDEEP BANERJEE[1]
JENNIFER PREMKUMAR[2]
MARIE THERESE MANIPADAM[2]
ANU EAPEN[3]
PHILIP JOSEPH[1]
FREDERICK LORENCE VYAS[1]
RAVISH SANGHI RAJU[1]
VENKATRAMANI SITARAM[1]

Correspondence: Dr. Venkatramani Sitaram
Division of Gastrointestinal Surgery[1] (HPB Unit),
Departments of Pathology[2] and Radiodiagnosis[3]
Christian Medical College, Vellore-632004, Tamil Nadu, India.
Email: sitaram@cmcvellore.ac.in; sur4@cmcvellore.ac.in

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Should gut malrotation be suspected in adolescents and young adults presenting with failure to thrive?

Introduction

Failure to thrive in young patients is usually attributed to chronic infections such as tuberculosis, human immunodeficiency virus (HIV) or to malabsorption syndromes. Gut malrotation (GM) as a cause of failure to thrive is usually a far-fetched suspicion. It is a congenital anomaly referring to either lack of or incomplete rotation of the foetal intestines around the axis of the superior mesenteric artery (SMA) during foetal development. Approximately 90% of patients with GM are diagnosed within the first year of life; of which 80% are diagnosed within the first month of life.[1]

Case report

Two young patients presented with failure to thrive and non-specific abdominal symptoms. Subsequently, they were diagnosed as cases of GM, which was responsible for their clinical condition.

The first case was a 13-year-old boy who presented with a history of poor appetite and failure to thrive since he was 5 years old. He was considerably malnourished and his weight was only 12 kg. Abdomen showed visible left to right peristalsis