Pancreatic peripheral primitive neuroectodermal tumor diagnosed by endoscopic ultrasound

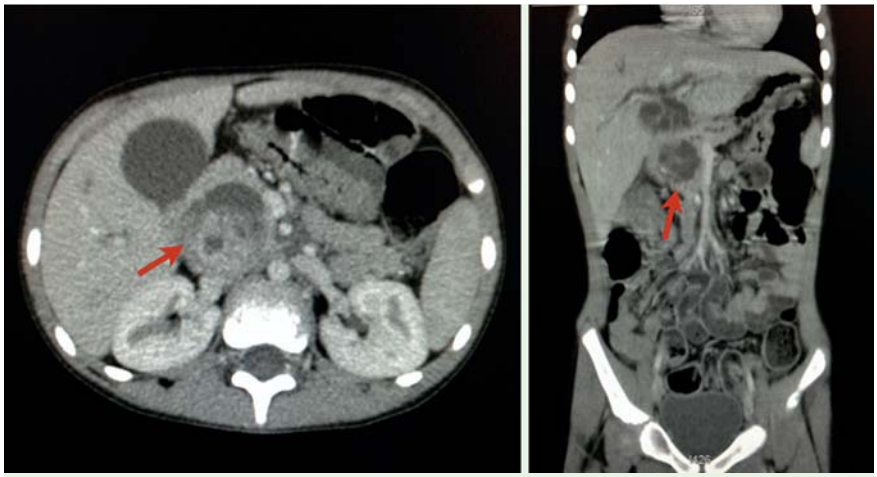

Fig. 1 Pancreatic peripheral primitive neuroectodermal tumor. Computed tomography in axial (left panel) and coronal (right panel) views showing a 4.5 × 4.0-cm well-delimited mass in the head of the pancreas (red arrows) with heterogeneous content, foci of calcification, and cystic/necrotic areas.

An 8-year-old girl presented with abdominal pain and jaundice of 1 month's duration. She had conjugated hyperbilirubinemia and negative hepatitis serology. Computed tomography showed a mass in the head of the pancreas, with foci of calcification and cystic/necrotic areas (▶ **Fig. 1**). Pancreatoblastoma and Frantz tumor were suspected. The patient underwent a cholecystojejunal anastomosis, and intraoperative biopsy of the pancreatic mass yielded inconclusive results. She was referred for endoscopic ultrasound (EUS) to re-evaluate the pancreatic mass.

EUS showed a solid–cystic lesion in the head of the pancreas without vascular involvement (▶ **Fig. 2**, ▶ **Fig. 3**). The main pancreatic duct and common bile duct were slightly dilated. EUS-guided fine-needle aspiration of the pancreatic mass was done with a 22-gauge needle (EchoTip; Cook Medical, Limerick, Ireland) (▶ **Fig. 4**). Cytopathologic evaluation of cell block material revealed a small cell neoplasm, and immunohistochemical analysis confirmed the diagnosis of peripheral primitive neuroectodermal tumor (PNET) (▶ **Fig. 5**, ▶ **Fig. 6**).

PNET belongs to a rare group of tumors called the Ewing sarcoma family of tumors [1–3]. Few PNETs arise in solid organs, and pancreatic PNETs are extremely rare [4–8]. Pancreatic PNETs are highly aggressive. Metastasis and recurrence are common, so that the prognosis is very poor. With modern multidisciplinary treatment, long-term survival can be achieved in 70% to 80% of patients with disease that has not metastasized [9].

The correlation of clinical symptoms with imaging, cytopathologic, and immunohistochemical analysis is useful to establish the diagnosis [10, 11]. An atypical rosette array of the cells, cytoplasmic neuronal secretory granules and neurofilaments, and pyknotic nuclear granules are important diagnostic criteria [4–8, 12]. Most tumors of the Ewing sarcoma family express high levels of a cell surface glycoprotein, CD99 [13, 14].

According to a 2014 review article [15], 14 cases of pancreatic PNET have been reported. This is the first case of a pancreatic PNET diagnosed by EUS.

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Competing interests: None

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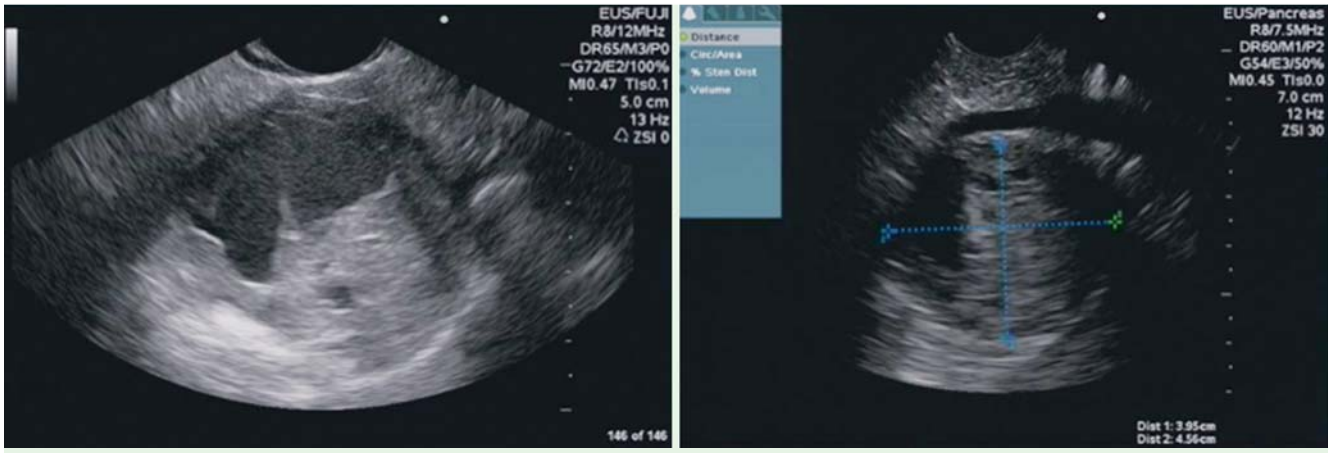

Fig. 2 Endoscopic ultrasound (stomach views) showing a solid cystic heterogeneous lesion in the pancreatic head.

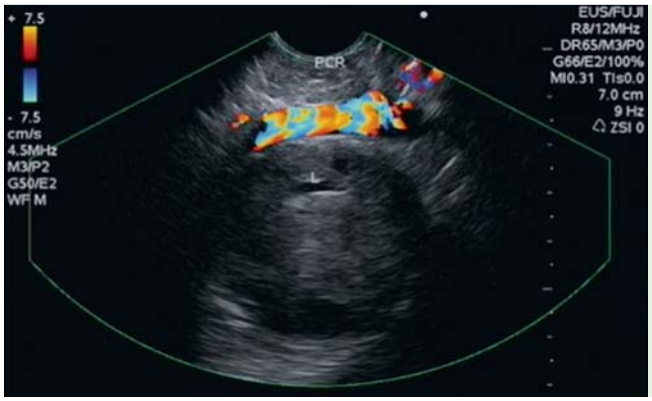

Fig. 3 Endoscopic ultrasound (stomach view) showing no echographic signs of portal vein impairment.

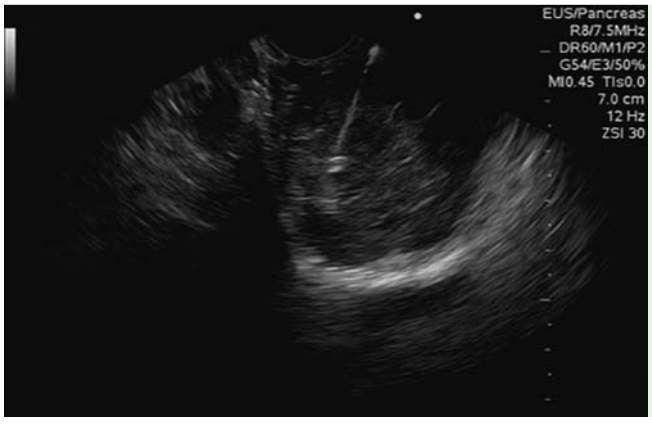

Fig. 4 Endoscopic ultrasound (stomach view) showing endoscopic ultrasound-guided fine-needle aspiration (22-gauge needle) of the solid cystic mass.

CEA	Negative
D1 CYCLIN	Focal positive
SYNAPTOPHYSIN	Focal positive
CHROMOGRANIN	Negative
Alpha-fetoprotein	Negative
Beta-catenin	Negative
CK7	Negative
Ki-67	Positive in 30 % of neoplastic cells
Tdt	Negative
Alpha$_1$-antitrypsin	Negative
VIMENTIN	Negative
CD99	Positive
FLY-1	Focal positive

Fig. 5 Immunohistochemical profile suggestive of primitive neuroectodermal tumor. CEA, carcinoembryonic antigen; CK, cytokeratin; Tdt, terminal deoxynucleotidyl transferase; CD, cluster of differentiation.

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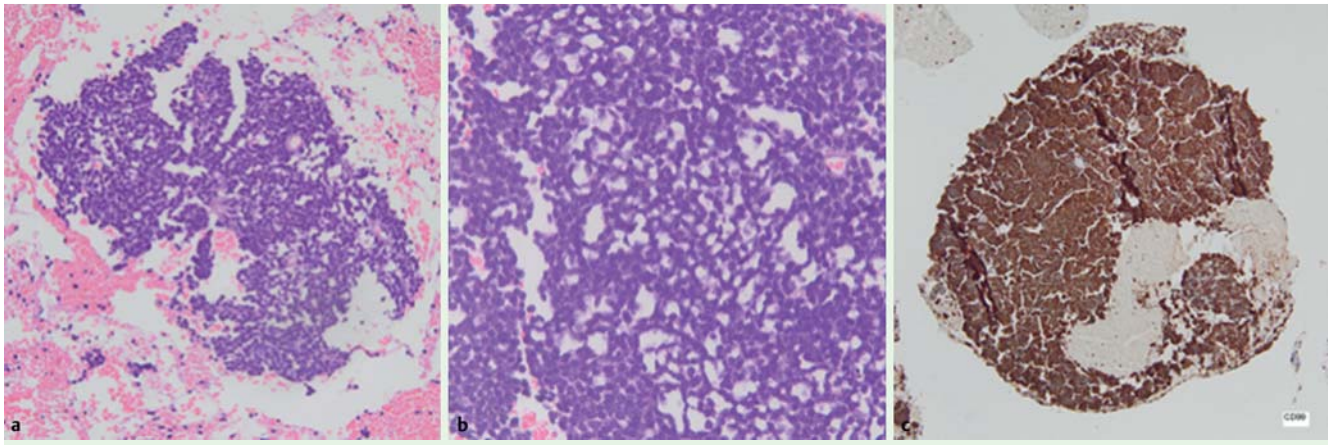

Fig. 6 Pancreatic peripheral primitive neuroectodermal tumor. **a** Cell block section showing clusters of rather uniform neoplastic cells arranged in a lobular pattern (hematoxylin and eosin, original magnification × 10). **b** Details of the neoplastic cells, showing scant cytoplasm, mild atypia, and a trabecular architecture. **c** Immunohistochemical reaction showing strong diffuse positivity for CD99.

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