Pancreatic peripheral primitive neuroectodermal tumor diagnosed by endoscopic ultrasound

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.

Competing interests: None

Flávio Amaro, Rogério Colaiácovo, Augusto Carbonari, Mauro Saieg, Ana Claudia Baraldi, Lúcio Rossini
Centro Franco Brasileiro de Ecoendoscopia (CFBEUS), Santa Casa de São Paulo, São Paulo, Brazil

References
2. Llombart-Bosch A, Lacombe MJ, Contesso G et al. Small round blue cell sarcoma of bone mimicking atypical Ewing’s sarcoma with neuroectodermal features. An analysis of
five cases with immunohistochemical and electron microscopic support. Cancer 1987; 60: 1570–1582
13 Ambros IM, Ambros PF, Strehl S et al. MIC2 is a specific marker for Ewing’s sarcoma and peripheral primitive neuroectodermal tumors. Evidence for a common histogenesis of Ewing’s sarcoma and peripheral primitive

CEA  Negative
D1 CYCLIN  Focal positive
SYNAPTOPHYSIN  Focal positive
CHROMOGRAIN  Negative
Alpha-fetoprotein  Negative
Beta-catenin  Negative
CK 7  Negative
Ki-67  Positive in 30% of neoplastic cells
Tdt  Negative
Alpha-antitrypsin  Negative
VIMENTIN  Negative
CD99  Positive
FLY-1  Focal positive

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.

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