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.

Endoscopy_UCTN_Code_CCL_1AF_2AZ_3AB

**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, Schrecl 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...


Bibliography
DOI http://dx.doi.org/10.1055/s-0034-1377982
Endoscopy 2015; 47: E11–E13
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

Corresponding author
Flávio Amaro, MD
Rua Haddock Lobo 807, Apt. 14
São Paulo, São Paulo 01414-001
Brazil
flavioamaro.obs@gmail.com