A pancreatic neuroendocrine tumor diagnosed during the management of acute appendicitis

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Pancreatic neuroendocrine tumors (PNET) are increasingly being discovered. A case of PNET diagnosed and treated during the management of acute appendicitis is presented and discussed. The importance of imaging modalities in patients with acute abdominal pain is emphasized. To the best our knowledge, this is the first pediatric report of PNET and acute appendicitis combination. ***Ann Pediatr Surg*** **9:150–151 © 2013 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2013, **9**:150–151

Keywords: acute appendicitis, children, pancreatic neuroendocrine tumor

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Received 3 December 2012 **accepted** 27 August 2013

Introduction

In recent years, pancreatic neuroendocrine tumors (PNETs) have increasingly been discovered by chance on imaging studies performed for various nonspecific abdominal symptoms. A case of PNET diagnosed and treated during the management of acute appendicitis is presented and discussed.

Case report

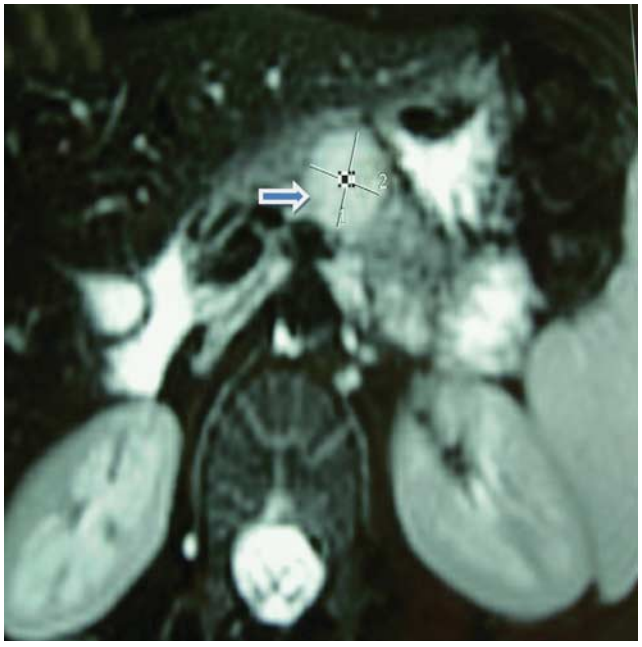
An 11-year-old boy underwent an appendectomy because of acute gangrenous appendicitis and localized peritonitis. Consent letter has been obtained from the patient's family. Before the appendectomy, all laboratory parameters were normal, except an increase in the white blood cell count. A doubtful solid pancreatic mass was reported during preoperative ultrasound examination. Histopathological examination showed only a gangrenous appendix without any neuroendocrine tissue. After the appendectomy, repeat ultrasound and MRI confirmed the presence of a solid pancreatic mass (Fig. 1). Biochemical parameters including tumor markers (24-h urine sample of 5-hydroxyindoleacetic acid, plasma levels of 5-OH-tryptamine, neuron-specific enolase, insulin, C-peptide, and gastrin) were found to be normal. Three weeks after the first operation, laparotomy was performed for the diagnosis and treatment of pancreatic mass. An encapsulated tumor with a diameter of 20 mm located between the head and the body of the pancreas was enucleated totally (Fig. 2). Frozen section of the tumor was determined to be a well-differentiated PNET with no capsular or vascular invasion and no necrosis. Immunohistochemically, the tumor showed positivity for synaptophysin and chromogranin A, and the expression of Ki-67 was found to be lower than 1%. The patient had an uneventful postoperative course. At present, he is being followed up by both us and the pediatric oncology department, and has no evidence of disease for 2 years.

Presented at the 29th Annual Congress of Turkish Association of Pediatric Surgeons, 16–19 May 2011, Istanbul Congress Center, Harbiye, Turkey.

Discussion

Incidentally discovered solid organ lesions with no symptoms (incidentalomas) are increasingly being detected because of the continuous improvement in imaging resolution. Although it is not the most prevalent site of such lesions, the pancreas may also be the involvement site of these tumors. The WHO classification classifies pancreatic lesions into exocrine and gastroenteropancreatic neuroendocrine tumors [1,2]. Primitive neuroendocrine tumors are rare tumors occurring at a rate of 6.5 per million in children 14 years of age or younger [3]. The most common site is the stomach (23%), followed by the pancreas, the appendix, the small intestine, and the rectum.

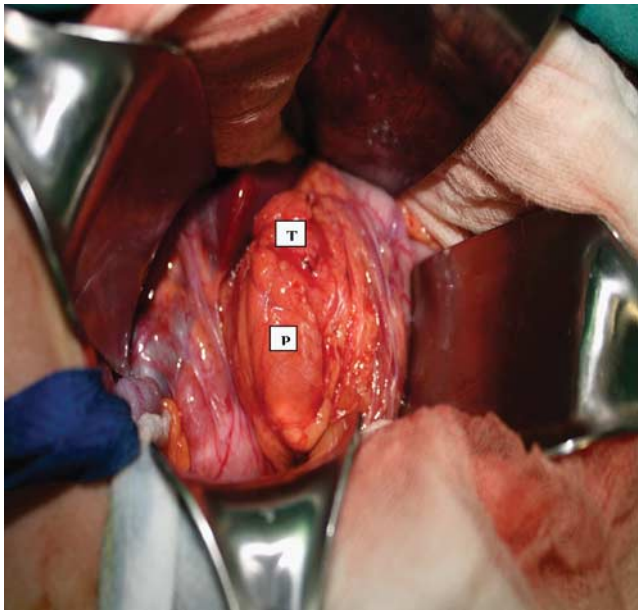
Fig. 1



MRI of the solid pancreatic mass (arrow: pancreatic mass).

1687-4137 © 2013 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000434559.51763.6c

Fig. 2



Operative view of the tumor located between the head and the body of the pancreas (P, pancreas; T, tumoral mass).

Asymptomatic lesions of the pancreas have appeared with increasing frequency in recent years. The first report of pancreatic incidentaloma in the literature was published by Kostiuk, in 2001, and several series have since been reported [4,5]. There are reports of pancreatic incidentalomas in the literature, detected mostly in adults using radiological methods [6–9]. In our patient, the tumoral mass was located between the head and the body of the pancreas, which has been reported in the literature as the second most common location for PNETs. About 46% of PNETs are classified as benign, 15.4% as uncertain, 31.9% as well-differentiated malignant, and 6.7% as poorly differentiated malignant [10]. Histopathological examination showed that PNET of our patient was a well-differentiated benign type with positivity for chromogranin A and synaptophysin, which are usual immunohistochemical markers for PNETs [11]. PNET cases usually presented with abdominal pain and jaundice [3]. Our patient was also admitted for abdominal pain; however, this was because of acute appendicitis. The pancreatic mass, a finding not consistent with acute appendicitis, was determined incidentally in radiological imaging studies. It is reported that 78–97% of appendiceal neuroendocrine tumors in children are associated with appendicitis [12–14]. However, only a gangrenous appendix without any neuroendocrine tissue was found in the histopathological examination of the appendix in our patient. Review of the literature on the basis of pancreatic incidentaloma, in the context of acute appendicitis, indicated a 21-year-old woman with findings of a solid pseudopapillary pancreatic tumor with a remarkable size at the time of diagnosis [15]. To our knowledge, our case is the first report of pediatric pancreatic incidentaloma in the context of acute appendicitis.

The importance of imaging modalities in patients with acute abdominal pain is emphasized in this report and if a pancreatic mass is found, it should be surgically excised. To the best of our knowledge, this is the first pediatric report of a combination of PNET and acute appendicitis; this information adds to the literature that PNET and appendicitis can be seen together as distinct entities in children.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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