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Non functioning Neuroendocrine tumour of pancreas in a 14 year old child- A rare case report

Oncology

KEYWORDS:

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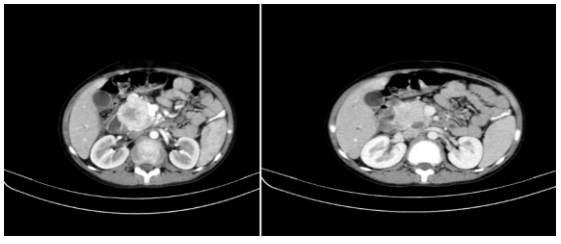
Introduction- Pancreatic NETs are rare subgroup of pancreatic tumors and represent about 1-2% of all pancreatic neoplasms. The incidence and prevalence of overall NETs has increased substantially Neuroendocrine tumors (NET) account for approximately 25% of pediatric pancreatic masses. NET arise from endocrine tissues of the pancreas and are classified as either non-functioning or functioning. Functioning NETs secrete various peptide hormones, which act at distant sites and can result in a variety of clinical syndromes. Pancreatic NETs can be benign or malignant; however the majority of non-functioning tumors are malignant. Pancreatic tumors in children rarely present with biliary obstruction, owing to the expansive rather than infiltrative growth patterns of the common pediatric pancreatic neoplasms.

Methods- A 14 year old male child presented with intermittent jaundice and epigastric pain since 6 months. The patient consulted a local doctor and was initially managed conservatively. As the symptoms persisted he was then referred to our institute. On investigating, Complete blood count was normal Total bilirubin was 5.2mg/dl with direct of 4.1mg/dl Se Alkaline phosphatase 253 IU/L,SGOT 91,SGPT 67 IU/L,Se.Albumin 2.95gm/d

The USG showed a mass in the head region of pancreas, with dilated common bile duct.

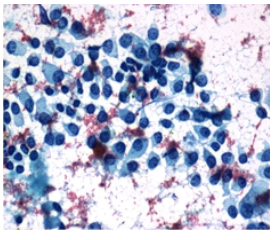
On follow up, abdominal CT scan was requested, which showed a retroperitoneal lesion in the right upper quadrant with a size of7.5 cm×6.6 cm×7 cm with intense peripheral heterogeneous enhancement on arterial phase with gradual central enhancement on delayed phase after administration of contrast media and infiltration of the duodenal wall without invasion of the large vessels, CBD measured 21mm.No evidence of metastasis.

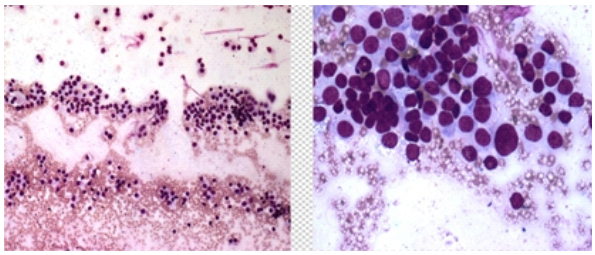


Endocrinal workup revealed non functioning tumour Se.Insulin, Se. Glucagon, Se.Lipase, Se.Amylase were negative.

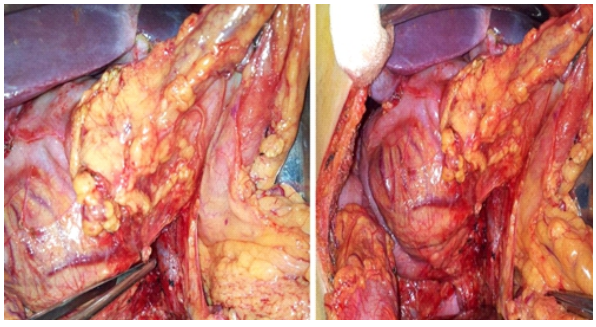
With the endoscopy result a EUS FNAC was carried out, which was reported as a neuroendocrine tumor positivity. Chromogranin and synaptophysin was positive, Ca 19-9, CK 7 and 20 were negative.

Cytologic smear showing dissociate cells with high N:C ratio and Salt-Pepper chromatin and occasional rosette like pattern.





After preoperative work up, he was subjected to elective exploratory laparotomy and pancreatoduodenectomy (Whipple procedure) was performed



with findings of a tumor of the head of the pancreas about 7 cm×4 cm×7 cm in diameter, well defined ,free in relation to large vessels, indurated, and involving the duodenum on its posterior and medial wall . The definite histopathological report was that of a neuroendocrine tumor of the head of the pancreas, with a mitotic index of 1-2 mitosis per 10HPF, and lesion-free borders . The patient had an uneventful clinical course without complications and is currently on regular follow-up without any complaints.

Discussion

Pancreatic tumors in children and adolescents are exceedingly rare. Investigators from Memorial Sloan-Kettering published a retrospective review of pediatric patients less than 21 years of age with malignant pancreatic tumors [1]. Patients with MEN were excluded. They identified sixteen patients with pancreatic neoplasms over a 35-year time period. Solid pseudopapillary tumors (SPT) were the most common pathological subtype, followed closely by pancreatoblastomas (PB) [1]. Only 2 patients (12.5%) had NETs: one was non-functional and the other was a malignant vasoactive intestinal peptide-secreting neuroendocrine tumor (VIPoma]. The majority of children or adolescents with pancreatic neoplasms present with abdominal pain, emesis or a palpable mass [1,5]. Biliary obstruction is rare [1,4]. Unlike ductal adenocarcinoma, the majority of pediatric pancreatic tumors exhibit an expansive growth pattern, and push nearby structures aside rather than invading them. Furthermore, they do not have a predilection for the pancreatic head, with only 38% found in this location in the series from Memorial Sloan-Kettering [3]. In particular, NET have a predilection for the pancreatic tail [5].

Complete surgical resection is the mainstay of treatment for malignant tumors of the pancreas in children. Due to the infrequent nature of these neoplasms and the histologic heterogeneity, the role of chemotherapy and radiotherapy is not proven, and their role in the adjuvant setting has yet to be clearly established [1,3]

Approximately 25% of pediatric pancreatic neoplasms are NETs [3,2,5]. The WHO grading system for NET depends entirely on histopathologic criteria, including proliferation index (Ki-67 index), mitotic count, and the presence or absence of tumor necrosis. The most common symptom at diagnosis is abdominal pain (85%) and weight loss (46%), while jaundice is rare [7]. The majority of pancreatic NETs in children and adolescents are non-functioning tumors [7]. In some case series, up to 50% of children had metastatic disease at presentation, and localized disease is less common in pancreatic NET than with other sites of extra-appendiceal NET [6,7]. These tumors may be associated with genetic syndromes such as MEN, Von Hippel-Lindau syndrome, Neurofibromatosis type 1, or tuberous sclerosis [5,6]. It is unknown whether high-grade tumors (WHO grade 2 or 3) are more common in the pediatric population.

The gold standard of treatment for extra-appendiceal NET is complete surgical resection. If complete surgical resection is not achieved, these tumors have a tendency to recur locally or metastasize

[8]. NETs respond poorly to chemotherapy. Over-expression of the somatostatin receptor is common in these tumors, so peptide receptor radionucleotide therapy is possible [6]. There is no standardized chemotherapy regimen in children, but in adults, both streptozotocin and fluorouracil are first-line treatments for stage III or greater [7]. Chemotherapy may have many adverse effects on the developing child, including failure to thrive, infertility and a significant risk of secondary malignancy.

Conclusion- Pancreatic tumors in children and adolescents are rare. Pancreatic NETs only represent 25% of these neoplasms.. The majority of children or adolescents with pancreatic neoplasms present with abdominal pain, emesis or a palpable mass. Unlike ductal adenocarcinoma, the majority of pediatric pancreatic tumors exhibits an expansive growth pattern, and push nearby structures aside rather than invading them. In particular, NET have a predilection for the pancreatic tail .

Complete surgical resection is the mainstay of treatment for malignant tumors of the pancreas in children. Due to the infrequent nature of these neoplasms and the histologic heterogeneity, the role of chemotherapy and radiotherapy is anecdotal, and their role in the adjuvant setting has yet to be clearly established. In contrast to the adult population, most children have resectable disease at the time of presentation and these tumors rarely present with biliary obstruction and have a good prognosis if complete surgical resection can be achieved, hence should be kept as strong differential .

References

[1] Shorter NA, Glick RD, Klimstra DS, Brennan MF, LaQuaglia MP. Malignant pancreatic tumours in childhood and adolescence: the Memorial Sloan-Kettering experience, 1967 to present. J Pediatr Surg 2002;37(6):887e92.
[2] Yu DC, Kozakewich HP, Perez-Atayde AR, Shamberger RC, Weldon CB. Childhood pancreatic tumors: a single institution experience. J Pediatr Surg 2009;44: 2267e72.
[3] Marchegiani G, Crippa S, Malleo G, Partelli S, Capelli P, Pederzoli P, et al. Surgical treatment of pancreatic tumors in childhood and adolescence: uncommon neoplasms with favorable outcome. Pancreatology 2011;11:383e9.
[4] Friedlander J, Quiros JA, Morgan T, Zhang Z, Tian W, Kehr E, et al. Diagnosis of autoimmune pancreatitis vs neoplasms in children with pancreatic mass and biliary obstruction. Clin Gastroenterol Hepatol 2012;10:1051e5.
[5] van den Akker M, Angelini P, Taylor G, Chami R, Gerstle JT, Gupta A. Malignant pancreatic tumors in children: a single-institution series. J Pediatr Surg 2012; 47:681e7.
[6] Rojas Y, Warneke CL, Dhamne CA, Tsao K, Nuchtern JG, Lally KP, et al. Primary malignant pancreatic neoplasms in children and adolescents: a 20 year experience. J Pediatr Surg 2012;47:2199e204.
[7] Redlich A,Wechsung K, Boxberger N, Leuschner I, Vorwerk P. Extra-appendiceal neuroendocrine neoplasms in children-data from the GPOH-MET 97 study. Klin Padiatr 2013;225(06):315e9.