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Case Report
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Solid Pseudo-Papillary Tumor of the Pancreas in a 9 Year Old: Pylorus Preserving Pancreaticoduodenectomy for a Rare Indication

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Abstract

Solid pseudo-papillary tumor of the pancreas is rarely seen among the pediatric age group. Common presenting complaints are abdominal discomfort and abdominal pain. This is the case of a 9-year-old girl with vague abdominal pain and works up done for evaluation of pain revealed pseudo papillary tumor of the head of pancreas. Patient underwent a pylorus preserving pancreaticoduodenectomy for removal of the mass. She made unremarkable post-operative recovery and was discharged 1 week after the procedure. Pancreatic resection, depending on the location of the solid pseudo-papillary tumor, is the mainstay of management as there is a low but well defined risk of malignancy. Early diagnosis of pseudo- papillary tumor of the pancreas in pediatric age group with abdominal pain can bring significant improvement in the patient management and outcome.

Keywords: Solid pseudo-papillary tumor of the pancreas; Pylorus preserving pancreaticoduodenectomy; Pancreatic neoplasm: Pediatric pancreatic neoplasm; Papillary cystic neoplasm

Abbreviations

AACT: Alpha 1-Antichymotrypsin; AAT: Alpha 1-Antitrypsin; AFP: Alpha Fetoprotein; CT: Computed Tomography; EUS: Endoscopic Ultrasound; IVC: Inferior Vena Cava; MRI: Magnetic Resonance Imaging; NSE: Neuron-Specific Enolase; PLAP: Placental Alkaline Phosphatase; SPTP: Solid Pseudo-papillary Tumor of the Pancreas; WHO: World Health Organization

Introduction

Solid Pseudo-papillary Tumor of the Pancreas (SPTP) was first described by V.K. Frantz in 1959 as 'papillary tumors of the pancreas. These tumors are of low malignant potential and rarely seen before the second decade of life [1]. It constitutes less than 3% of all pancreatic tumors and most frequently present in the head or tail of the pancreas [2]. A malignant variant of SPTP is defined by World Health Organization (WHO) as a tumor with tissue, perineural and perivascular invasion on microscopic analysis along with metastasis [3]. Clinical manifestations include abdominal pain, discomfort, jaundice and palpable mass [4]. Diagnosis is difficult due to variability of signs and symptoms and it is usually diagnosed on imaging studies because of its characteristic morphology, moreover confirmation relies on histopathology [1]. Distinguishing radiological features noted, are enhancement patterns like heterogeneity within a well-circumscribed tumor with a fibrous pseudo capsule [5]. Surgical resection is internationally recommended as it is associated with positive long-term outcome. Metastasis is seen in 10% of cases, usually arising in liver or regional lymph nodes [2]. Five year survival is 97%, even in presence of metastasis [6].

Case Presentation

A 9 year old girl was referred to Hepatobiliary and Liver Transplant Surgery service, at Sindh Institute of Urology and Transplantation, Karachi, Pakistan, complaining of epigastric pain. She had experienced multiple episodes of vomiting, one month earlier, that resolved after a few days but was followed by intermittent dull aching upper abdominal pain of moderate intensity. There were no other associated symptoms such as nausea, vomiting or weight loss. Examination at that time was unremarkable. The patient had no significant family history including malignancies or other neoplastic lesions. Sonography, done previously, showed a well circumscribed mass at pancreatic head region measuring 4.4 cm × 3 cm. A CT scan was performed that showed a round hypodense

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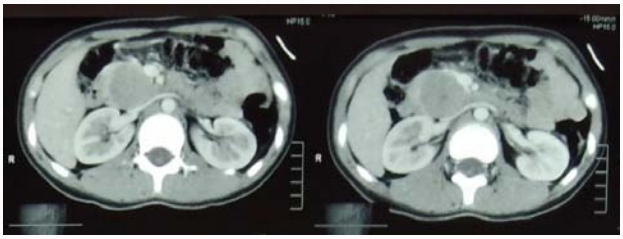

Figure 1: Axial view of abdominal CT scan (post contrast) with a mass in pancreatic head, measuring 33 mm × 31 mm × 41 mm in size compressed right kidney vein and inferior vena cava.

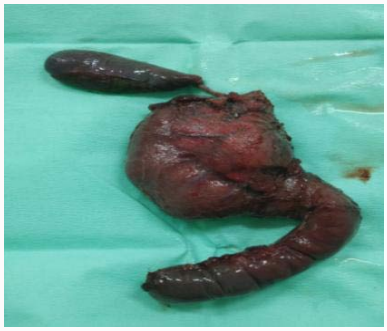

Figure 2: Specimen of resected pancreatic head with the tumor, duodenum and gallbladder.

mass in head of pancreas having an indistinct interface with the second part of duodenum, with normal body and tail of the pancreas. It was compressing Inferior Vena Cava (IVC) and right renal vein. Mass measured around 3.3 cm × 3.3 cm × 4.1 cm. significant contrast enhancement was noted with central non-enhancing scar within the lesion. Patient was admitted and underwent Endoscopic Ultrasound (EUS) guided biopsy of the mass. Histopathology revealed fragments of neoplastic lesion composed of papillary architecture of rounded to polygonal cells, exhibiting eosinophilic cytoplasm and uniform, rounded, normal chromatic nuclei in the background of fibrinous exudates and myxoid stroma suggestive of solid pseudo-papillary tumor of pancreas. All Immunohistochemistry markers were negative, including, synaptophysin, chromogranin, CD-56, Alpha Fetoprotein (AFP) and Placental Alkaline Phosphatase (PLAP) (Figure 1).

Patient underwent elective operative exploration and pylorus preserving pancreaticoduodenectomy was performed (Figure 2). Intraoperative findings included grossly enlarged, encapsulated, well circumscribed mass of the head of pancreas measuring 4 cm × 4 cm approximately. There were no obvious metastasis or ascites noted. Post-resection histopathology reported encapsulated, well circumscribed, gray white solid homogenous tumor, size of 3.5 cm × 3.5 cm in the head of pancreas with no invasion in to ampulla and 2 cm away from surgical resection margin. The histopathological feature confirmed solid pseudo papillary tumor of the pancreas.

Discussion

Solid Pseudo-papillary Tumor of the Pancreas (SPTP) is also known as solid tumor of the pancreas, Frantz tumor, papillary-cystic tumor and papillary epithelial neoplasm [1]. Three common types of pancreatic tumors are mesangial, endocrine and exocrine among which papillary tumors constitute about 1% to 2% of exocrine papillary pancreatic tumors [6]. It has unknown pathogenesis with a strong predilection for females (10:1) [1,4] and most patients are younger than 30 years of age [4]. According to a theory, progesterone receptors expression in some cases is linked with tumorigenesis in association with female sex hormones, however definitive relationship is still controversial [4]. WHO classified SPTP as potentially malignant tumors in a report in 2010 but the majority of these tumors are benign and malignancy was identified in 9% to 15% of the patients [5]. Sevenfold increase incidence has been noted in number of SPTPs since 2000 among which 90% cases were detected incidentally on imaging studies [5]. Solid pseudo-papillary tumors of the pancreas can be asymptomatic or symptomatic, depending upon the size. Symptoms frequently include abdominal pain, nausea, vomiting, jaundice among others. Abdominal examination may reveal upper abdominal mass. Among pediatric patients, based on earlier reports, the distinctions of SPTP include the ratio of female to male of 2.2:1 and a median age of 9.5 years. Presenting complains of these children were abdominal pain (50%) and abdominal mass (18.75%), whereas rest were asymptomatic and were diagnosed incidentally (31.25%). The median size of tumor noted was 7.9 cm. Pediatric patients tend to have SPTP in the head of pancreas, whereas it tends to be in the body and tail of pancreas in adults [7].

Imaging studies are of crucial importance in the diagnostic evaluation of pancreatic tumors. Ultrasound may show fluid-debris level within the mass seen in epigastric region [2]. EUS provides an opportunity of preoperative histopathological diagnosis of tumor by biopsy [1]. Contrast enhanced CT demonstrates well-circumscribed lesion with heterogenous components, however, MRI is considered superior to CT due to its better contrast resolution, which helps with visualization of cystic degeneration, hemorrhage or the capsule [1]. Contrast administration prior to CT causes enhancement of solid component at the periphery of tumor and cystic area is seen at center [2].

Confirmatory diagnosis of the tumor is based on histopathology and immunohistochemistry. On microscopy, these tumors show pseudopapillary appearance with fibrous septations and degenerative appearance with a fibrous pseudocapsule [1].

These tumors may show positive expression rates of Vimentin, Alpha 1-Antichymotrypsin (AACT), Neuron-Specific Enolase (NSE), Alpha 1-Antitrypsin (AAT), Synaptophysin and progesterone receptor. Ki-67 index is an indicator of the malignant potential of the tumor and the Ki-67 index of ≤ 5% is associated with better survival as it indicates slow growth of neoplasia [8].

Surgery is curative in more than 95% of cases. Type of surgery depends upon size and site of the tumor. Enucleation is enough for very small tumors. Lesions located in the head of pancreas require pancreaticoduodenectomy (Whipple's procedure) whereas tumors in the neck of pancreas can be treated by techniques like central pancreatectomy. Moreover, lesions in the body and tail require distal pancreatectomy (with or without splenectomy) [1,4]. Lymph node dissection is indicated in SPTPs of more than 5 cm due to chances of malignancy [1]. In this case, pylorus preserving pancreaticoduodenectomy was done for pseudopapillary tumor of the head of pancreas.

Poor prognostic features are male patients, Ki-67 expression, large tumor (more than 5 cm) [5]. Metastasis is noted in 10% to 15% of patients with advanced disease [6]. Sites of metastasis included liver, peritoneum, omentum and regional lymph nodes [1]. Five year survival is 97%, even in presence of metastasis [6].

Conclusion

Solid pseudopapillary tumors of Pancreas are well circumscribed neoplasia of low malignant potential, however metastasis should always be ruled out.

These tumors are resectable with good prognosis and early recovery.

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