VOL. 21 | NO. 3 | ISSUE 83 | JULY-SEPT. 2023

Central Pancreatectomy for Bilobed Solid Pseudopapillary Tumor of Pancreas

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Citation

Jha MK, Ramtel R, Adhikari B, Laudari U, Thapa A, Rawal SB. Central Pancreatectomy for Bilobed Solid Pseudopapillary Tumor of Pancreas. *Kathmandu Univ Med J.* 2023;83(3):348-51.

ABSTRACT

Solid-pseudopapillary tumor of pancreas is a rare pancreatic tumor that occurs predominantly in a young female with only a small minority concerning children. These tumors have low malignant potential, found incidentally which are usually single lobed. Surgical resection remains the mainstay of treatment. Our case is 26 years female with vomiting and abdominal pain localized to the right upper quadrant. Computerized tomography scans showed two well-defined hypodense lesions at neck and body of the pancreas with greatest diameter of six centimeter. The patient underwent central pancreatectomy and histopathology confirmed solid-pseudopapillary tumor with complete resection with all resected margin free of tumor. Symptoms may be present such as abdominal pain or vomiting due to compression, mainly in large tumors. In benign cases parenchyma sparing surgery such as central pancreatectomy can preserve the exocrine and endocrine function of pancreas which helps patient to have better postoperative quality of life.

KEY WORDS

Bilobed mass, Central Pancreatectomy, Solid-pseudopapillary tumor

INTRODUCTION

Solid pseudopapillary tumors (SPT) were first described by Frantz in 1959. Solid pseudopapillary tumor is a rare tumor of the pancreas, accounting for 0.2 to 2.7% of all pancreatic tumors. Most patients are female usually within second or third decade of life with only a small minority being children.[1-3] Frequently they are found incidentally, presenting as a slowly growing abdominal mass with nonspecific symptoms, such as abdominal pain and vomiting. These are considered to be a low-grade malignant tumor, with an incidence of malignant transformation of around 15% and surgical resection is the treatment of choice with an excellent long-term prognosis.[4-7]

CASE REPORT

A 26-year-old female, presented to our surgical outpatient department of Nepalese Army Institute of Health sciences on September 28, 2017 with abdominal pain and vomiting of 1-month duration which was localized to the right upper quadrant. She had no history of weight loss or diarrhea and had no jaundice with no relevant past medical or surgical history. On physical examination, a mass was palpable on the right hypochondriac region. Computed tomography (CT) scan done showed the presence of two well defined heterogeneous, iso-echoic lesion composed of solid and cystic components at neck and body of the pancreas of size 4.1 x 3.9 cm & 5.5 x 6.0 cm approximately.

However, no encasement of adjacent vascular structures was seen in CT (Fig. 1).

In an endeavor to confirm the diagnosis, the patient underwent fine needle aspiration cytology (FNAC) of a mass which confirmed the diagnosis of a solid pseudopapillary tumor (SPT). There was no evidence of pancreatic insufficiency, abnormal liver function, cholestasis or elevated pancreatic enzymes. Her laboratory values including tumor markers: alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), CA-125, CA19-9 was found to be within normal limits. So, patient underwent an elective surgery which revealed a bilobed mass at body and tail of the pancreas, without evidence of

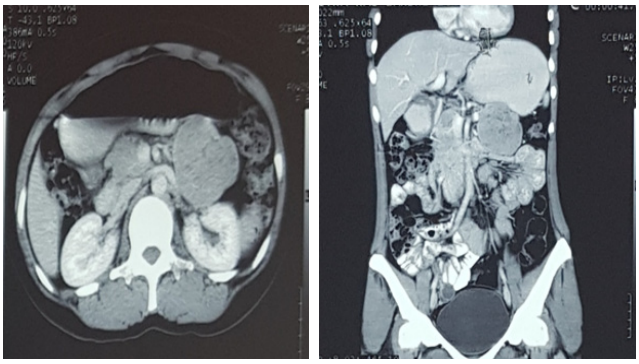

Figure 1. CECT pancreatic protocol showing pancreatic mass

metastatic disease, and a central pancreatectomy with pancreaticogastrostomy was performed (Fig. 2 and 3). The mass was completely resected without any complications. The postoperative course was uneventful and no adjuvant therapy was given. The patient was discharged from the hospital on the 15th postoperative day. Histopathology showed a well-circumscribed tumor with a fibrous pseudo capsule, composed of sheets of polygonal cells with pseudopapillary formations, confirming an SPT with complete resection with negative margin and no evidence of malignancy.

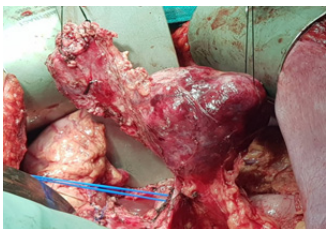

Figure 2. Intraopertive picture showing pancreatic mass

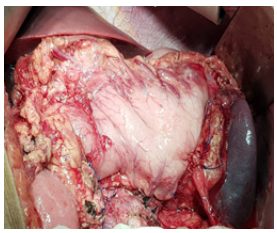

Figure 3. Pancreticgastrostomy after resection of pancreatic tumor

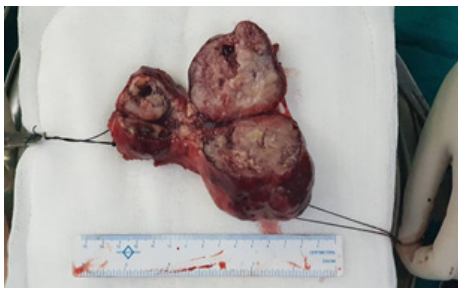

Figure 4. Resected bilobed mass

DISCUSSION

This is a case of 26 years old female initially presented with complaints of abdominal pain in the epigastric region for 2 months. On evaluation with ultrasound of abdomen, there was an incidental finding of a mass in the head of the pancreas whereas CECT abdomen showed a mass in neck and body of the pancreas. USG guided FNAC was done to rule out malignancy. The patient underwent exploratory laparotomy with Central Pancreatectomy with pancreaticgastrostomy. The resected tumor consisted of bilobed mass involving the body and part of the neck of the pancreas with mass abutting the splenic artery and splenic vein. Currently, the patient is asymptomatic on her regular follow up after two years of surgery.

SPT of the pancreas is a rare exocrine pancreatic neoplasm, constituting of 5 percent of all cystic pancreatic neoplasm and about 1 to 2 percent of exocrine pancreatic neoplasm. Predominantly encountered in young females on second to the fourth decade of life, and has also been seen in males and children.[8-10] The tumor is usually distributed in the pancreatic head (39.8%), tail (24.1%), body and tail (19.5%).[9] Our patient was also a young female of 26 years old which is a common age in presentation in most of the literature.

The predominance of SPT of the pancreas in female suggests that some gender-specific factor should play a role in its pathogenesis. A study done by Machado et al. concludes that the neoplasm in males is more aggressive when compared to females.[11] However, a study done by Cai et al. states males with SPT of the pancreas are older in age than female patients with no difference in prognosis following surgery.[12] A study was done by Klimstra et al. suggest that there is no gender-based relation for the pathology of this neoplasm.[13]

Due to a wide range of appearance of this tumor from solid to cystic, there have been various changes regarding the nomenclature of this tumor from Frantz tumor to a papillary cystic neoplasm. However, the neoplasm is known as “solid pseudopapillary tumor” of pancreas after World Health Organization defined this term.

Most patients have unclear clinical features of abdominal pain or discomfort, poor appetite, nausea, jaundice, and intestinal obstruction mostly due to tumor compression to the adjacent organ with an abdominal mass on examination. Out of these, the common presenting complaint is of abdominal pain and abdominal mass.[14] However, there are considerable patient populations who are asymptomatic and the neoplasm is incidentally detected.[9] The presentation of the case here was with the symptom of vague abdominal pain for 2 months with increasing intensity since 12 hours with no symptom of abdominal mass or jaundice and an incidental finding of SPT of the pancreas on evaluation.

For the diagnosis of SPT, various modalities included are CT scan, ultrasonography, endosonography, MRI. CT scan and Endosonography are more specific and sensitive modalities in diagnosing SPT.[9,15] In our case study, Incidental finding on CT scans along with ultrasonography and USG guided FNAC was used for the preoperative workup and conclusive diagnosis. Though Biopsy and FNAC are controversial and rarely performed, our main aim of doing FNAC was to rule out malignancy.[6] EUS was not done due to its unavailability in our setup.

CT scan usually shows solid and cystic component of a

well-encapsulated mass with enhanced solid portions on its periphery after contrast administration. The size, depth of penetration; pancreatic anatomy and also invasion to the surrounding structures can be defined by the help of CT scan. CECT abdomen of the patient showed two heterogenic enhancing lesions in the neck (4.1 x 4.1 cm) and body (5.6 x 6.2 cm) of the pancreas. Foci of calcification in the lesion of the body with no encasement of adjacent vascular structures. Other major pancreatic cystic neoplasms should be taken into consideration, such as cystadenocarcinoma or intraductal papillary mucinous neoplasms. Endoscopic ultrasound (EUS) and endoscopic retrograde cholangiopancreatography (ERCP) can help clarify the diagnosis.[6]

Magnetic resonance imaging is extremely useful in diagnosing cystic lesions. The lesion appears large, well defined, encapsulated with heterogenous high or low signal intensity on T1 weighted, heterogeneous high signal intensity on T2 weighted, and early peripheral heterogeneous enhancement with progressive fill in on gadolinium-enhanced dynamic MR imaging. These features are useful to differentiate this rare tumor from other pancreatic neoplasms.[17] Immunohistochemical studies of the neoplastic cells of the tumor regularly express positive for b catenin, vimentin, alpha 1 antitrypsin, antichymotrypsin, neuron-specific enolase, and cyclin D1. This modality of investigation is not useful in differentiating with any normal pancreatic cell. Thus, Routine immunohistochemical staining is not useful.[13]

The SPT of the pancreas has low malignant potential and has good prognosis among female patients. Less than 15 percent of the cases with this tumor commonly metastasizes into the liver, lymph node and can also have peritoneal dissemination.[10] Patients with local recurrence, liver and peritoneal metastasis are also reported to have a good prognosis with long-term survival. The study has suggested that the older age of the patient and size of the tumor is proportional to each other in case of metastasizing SPT of the pancreas. High degree of cellular pleomorphism along with elevated mitotic rate and tumor involving blood vessels and adjacent organs are usually found to be associated with metastasis.[13] The patients presented here was a young female with no metastasis to any adjacent organs.

SPT has malignant potential, therefore surgical resection is the main mode of treatment.[18-20] Extensive procedure such as Pancreatoduodenectomy (PD) for pancreatic head tumor and Distal pancreatectomy (DP) for pancreatic body and tail tumor are sensible treatment for malignant lesion but can result in early development of postoperative exocrine and endocrine insufficiency hence it is unnecessary for benign lesions of neck and proximal body of the pancreas.[21]

Enucleation and central pancreatectomy (CP) are the operative procedure that spare the normal pancreatic parenchyma and have high morbidity; however, these procedures lower the risk of early development of exocrine and endocrine insufficiency.[22] According to study done by Yasugi et al.[23] Patient developed Hyperglycemia immediately when pancreas was resected more than 70% whereas the patient did not develop diabetes when pancreas was ressected less than 70%. Such results have highlighted the need of CP and enucleation for benign and borderline tumor of body and neck tumor of pancreas.

Indication for enucleation is that lesion has to be benign, a tumor located in head and body, neuroendocrine tumor, branch-duct intraductal papillary-mucinous neoplasm (IPMN) , cystic tumor and proximity of lesion to the pancreatic duct CP is an operative procedure for management of tumor of pancreas located in body and neck region.[24-26] In 1984 Dagradi and Serio removed a benign insulinoma by a central pancreatectomy, making it the first such resection for a pancreatic neoplasm.[27]

Indication for CP are that a tumor must be benign, where simple enucleation has risk of injury to the main pancreatic duct and in young patients with cystic lesion who are not suitable for enucleation such as symptomatic serous cystadenoma , mucinous cystadenoma, intraductal papillary mucinous neoplasm, pseudopapillary tumor, small tumor which are present in deep region which cannot be cleared by enucleation, fibrotic stenosis of wirsung duct, solitary metastases in the pancreatic neck.[27-29] In a study done by Wang et al. enucleation was done in 31 cases of SPT with the better postoperative outcome and endocrine function compared to conventional resection.[30]

In our case tumor was bilobed which was present in the neck (4.1 x 4.1 cm) and body (5.6 x 6.2 cm), therefore enucleation was not possible because of tumor location and size. CP was opted instead of Pancreaticoduodenectomy and distal pancreatectomy in order to preserve pancreatic endocrine and exocrine function which can improve postoperative quality of life of the patient and can aid in longer life expectancy. CP is a safe procedure with good long-term functional reserve but it increases the risk of postoperative pancreatic fistula.[31,27] Follow-up is essential due to the potential of local recurrence and appearance of metastasis, most commonly at the liver, regional lymph nodes, mesentery, omentum and peritoneum.[32-34]

Solid pseudopapillary tumors of the pancreas are extremely rare and are usually single lobed tumor that typically affects young females without significant symptoms. This patient had bilobed mass involving neck and body of pancreas. Pancreatic parenchyma sparing surgery like Central Pancreatectomy is useful in tumor such as solid pseudopapillary tumor of pancreas to improve the postoperative quality life of the patient where complete surgical resection of the tumor is the only effective

treatment option.

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