Case Report of an 18 Year Old Girl with Solid Pseudo-papillary Tumor of Pancreas

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ABSTRACT

Solid Pseudo-papillary Tumor (SPT) of the pancreas is a rare tumor which typically affects young women without any significant clinical symptoms. Solid Pseudo-papillary Tumor usually shows an indolent behavior and only rare cases recur and/or metastasize after complete resection. Here is a case report of 18 years old girl who presented to our centre with complaints of severe epigastric pain and underwent pancreatic parenchyma saving surgery for a large pancreatic head mass. In conclusion, Solid Pseudo-papillary Tumor being a large tumor possess a low malignant potential in which R0 resection has excellent prognosis.

KEY WORDS

Enucleation, Frantz tumor, Solid pseudo-papillary tumor (SPT), Surgical resection

INTRODUCTION

Solid Pseudo-papillary Tumor (SPT) of pancreas is a rare form of pancreatic tumor with incidence of 1-2% of all exocrine pancreatic tumors with very low malignant potential.[1-6] The tumor was first described by Frantz in 1959 so called as Frantz tumor which later in 1996 was described as "Solid Pseudo-papillary Tumor" by WHO.[2,4,7] The name itself is suggestive that the tumor is neither truly solid nor truly cystic. It occurs predominantly in young females.[1,3] Curative surgical resection is the treatment of choice where prognosis is very good as 5 year survival of patients undergoing complete resection of tumor is more than 97%.[2,3,7,8] Advanced cases may require chemotherapy (5-FU, Cisplatin, Gemcitabine) but the use of chemotherapy has not been proved to be beneficial.[6] The mainstay of treatment remains the surgical intervention in almost all cases, including advanced cases as the prognosis is very good after extensive surgical resection.[1]

CASE REPORT

An 18 years old female married for two years presented to primary care physician with complaints of severe epigastric pain for two days which later resolved on taking analgesics. She also complained of occasional upper abdominal pain for last one year for which she did not seek any medical advice and was relieved on taking medications from local medicine shop. She has no other significant medical and surgical history in the past. Family history reveals that her grandfather's sister died of some kind of intestinal malignancy (exact diagnosis not known). General examination was unremarkable. In per abdominal examination there was a palpable non tender intraperitoneal mass on right hypochondrium around 5x5 cm which was mobile. No other obvious findings were noted except mild tenderness on epigastrium and bowel sound was heard. USG abdomen revealed a well encapsulated, lobulated, heteroechoic mass with internal vascularity which was involving head of pancreas measuring about 10.3x6.8 cm along with left ovarian simple cyst

measuring 2.8x2.5 cm. Computed tomography revealed well defined mixed density soft tissue mass lesion with both cystic and solid components measuring approximately 10.88x7.02x7.67 cm involving the head of pancreas without any calcification within the lesion. On contrast administration there was heterogeneous enhancement and appeared hypo-dense compared to the normally enhancing parenchyma of pancreatic body and tail. Anteriorly the mass was abutting the anterior abdominal wall; posteriorly it was abutting the right kidney with mild perirenal and perilesional (posterior aspect) fat smudging. The distal common bile duct was prominent in the region of pancreatic head but the main pancreatic duct and proximal common bile duct was not dilated.

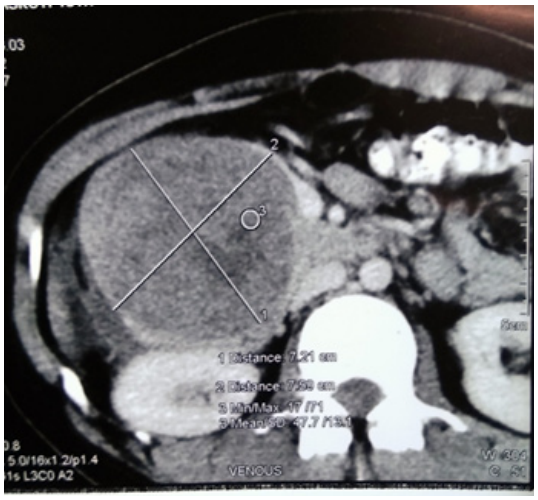

Figure 1. **CT scan showing large mass per abdomen**

Preoperative diagnosis was made with USG guided transcutaneous FNAC which showed tumor cells arranged in sheets and singly scattered with moderate amount of cytoplasm with enlarged nuclei and granular chromatin. Mild pleomorphism was noted which was suggestive of SPT of pancreas. UGI endoscopy and colonoscopy were performed to rule out the extension of tumor to the GI tract which were found to be normal and to our surprise no external compression was observed. All her hematological and biochemical tests were sent which were found to be normal except leucopenia (TLC: 2900, N: 47, L: 48, E: 01, M: 04) which resolved after a course of antibiotics (TLC: 4100). Patient was planned for surgery and prepared for it. Antibiotics and vitamin K was started along with adequate hydration three days prior surgery.

Laparotomy was performed via a rooftop incision. As tumor was not invading the pancreatic head deeply; pancreatic parenchyma saving enucleation of tumor was done. Whole of the tumor was removed along with its capsule. Per operatively there was a cystic mass of size 11.5X7X7 cm arising from the head of pancreas (fig. 3) with minor adhesions to the first part of duodenum (fig. 4). Capsule of the tumor was found to be highly

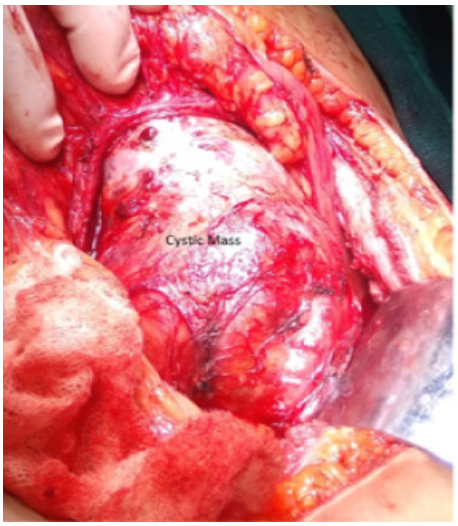

Figure 2. **Initial per-operative appearance of the tumor**

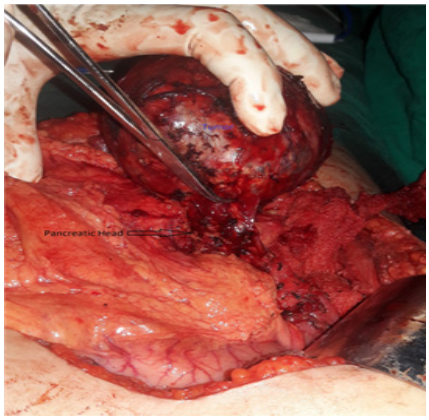

Figure 3. **Tumor arising from the pancreatic head**

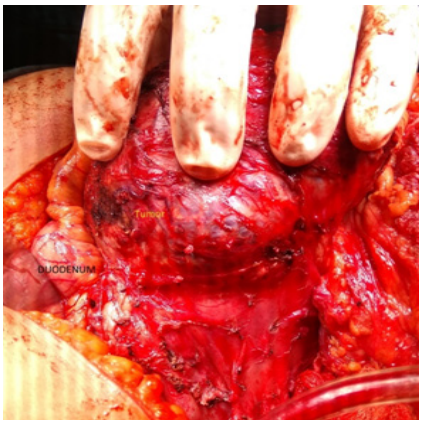

Figure 4. **Tumor adhesion with first part of the duodenum.**

vascular and friable due to which minor tears occurred leading to spillage of hemorrhagic content (around 100 ml). There was no ascites and metastases in abdomen.

Gross examination of the tumor showed a large encapsulated mass of size 11.5X7 cm (fig. 5) with hemorrhagic and necrotic areas in cut section (fig. 6).

Specimen was sent for pathological study and microscopic examination which confirmed our diagnosis. Microscopy showed tumor tissue arranged in papillary architecture with prominence of fibrovascular core and

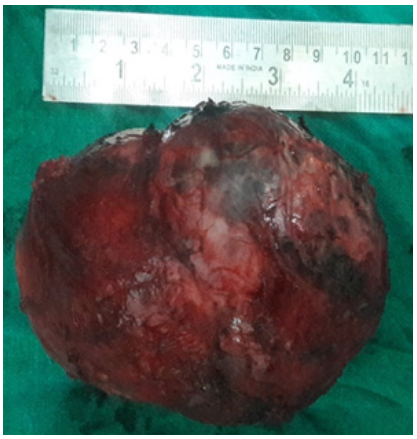

Figure 5. **Gross appearance of tumor after removal.**

Figure 6. **Cut section of the tumor.**

lined by cuboidal cells with fine nuclear chromatin and indistinct nucleoli with mild to moderate amount of pale cytoplasm and accumulation of foamy cells were seen admixed with mucinous deposition in focal area. Immunohistochemical analysis showed that cytoplasm was positive for CD-10 and alpha-1antitrypsin. Post-operative period was uneventful and patient was discharged on 8th post operative day.

DISCUSSION

SPT of pancreas is a very rare form of pancreatic tumor with incidence of 1-2% of all exocrine pancreatic malignancy with low malignant potential among which 20% can end up in malignancy.[1,2,7,9] Malignant disease is called Solid Pseudo-papillary Carcinoma of Pancreas. It predominantly affects females (F:M ratio being 10:1) of young age (mean age being early 20s).[1] It is found to be common in Asian females but the geographical distribution has not been established yet.[2] Before WHO described it as Solid Pseudo-papillary Tumor it was known with different names in different times i.e. Frantz or Hamoudi tumor, solid and papillary tumor, papillary cystic tumor, solid-cystic tumor, solid cystic and papillary tumor.[2,4,7] Differential diagnoses include pseudocyst of pancreas and other pancreatic cystic tumors.[5,10] Some typical features like low malignant potential and rare metastasis differentiates it from other pancreatic cancers.[2] Though slow growing, patients usually presents after tumor becomes significantly large.[11] Patients commonly presents with vague symptoms like upper abdominal pain, some may present with abdominal lump in right or left hypochondrium and in some cases it may be the incidental finding.[1,3,5,6,12] Jaundice and vomiting may occur due to obstruction of bile duct and duodenum respectively by the growth but is rare.[3] In majority of cases all biochemical and hematological reports are normal.[3] In contrast to other pancreatic neoplasm, serum tumor markers (CEA and CA19-9) are not usually elevated.[5] Initially it starts as a solid tumor but later undergo massive degeneration giving rise to cystic appearance in radio-imaging. It is usually a large tumor with average size of 8-10 cm which could easily be figured out in simple radiological investigations. USG abdomen shows a heterogeneous (due to both solid and cystic components) well encapsulated mass arising from the pancreas.[13,14] Extrapancreatic tumor site has also been noted i.e. mesocolon, ovary, retroperitoneum, liver, stomach and duodenum but pancreas being the commonest site and manifestation in our patient, it has been discussed here.[14] Preoperative diagnosis can be made with the CECT abdomen which shows a well-encapsulated lesion with varying solid and cystic components owing to hemorrhagic degeneration; following intravenous contrast administration, enhancing solid areas are typically noted peripherally, whereas cystic spaces are usually more centrally located. Mural nodules for solid structure, “floating cloud” signs, or solid and cystic crossing distributions may be the typical findings in case of tumors having predominant cystic component or equal proportion of solid and cystic components.[15] Pathological confirmatory diagnosis can be made by USG guided fine needle aspiration and cytological examination which shows tumor cells arranged in sheets and singly scattered with moderate amount of cytoplasm with enlarged nuclei and granular chromatin.[5] Preoperative precise pathological diagnosis is always important as surgeon will be able to decide the extent of surgery which will reduce postoperative morbidities.[4] Despite the technical advances in medical field etiology and pathogenesis of SPT has not been established yet but sex and age distribution point to genetic and hormonal factors.[2] Metastasis is rare but if it occurs it commonly metastasizes to liver, regional lymph nodes, peritoneum and greater omentum.[9,12] Complete surgical resection of the tumor is the modality of treatment followed all over the world. Radical resection is commonly performed i.e. pancreatoduodenectomy for tumor arising from head and distal pancreatectomy with/out splenectomy for tumor arising from the tail. If complete curative surgical resection is achievable some surgeons prefer local resection or enucleation referring to its benign nature as performed in this case.[1,8] First

surgical resection was performed by Grosfelad and was described by Hamoudi in 1970.[2] Overall survival of patients who undergo complete surgical resection is 95-98% and 93% respectively including advanced cases with metastasis.[2,8] Even in patients who had local spread, recurrences or metastases; long disease free periods have been reported after initial diagnosis and resection.[2] Malignant behaviour of the disease is characterized by perineural invasion, angio-invasion and deep invasion into surrounding tissue.[1,5,12] Metastatic disease and unresectable tumors are usually treated with chemotherapy but significance of adjuvant chemotherapy has not been established yet.[3,6] Some authors' also give verdict in use of radiotherapy in advanced malignant disease but no proper literature is found regarding the significance of radiotherapy.[3,6] Macroscopic examination of the cut section of surgical specimen reveals lobulated, light brown solid areas, zones of hemorrhage and necrosis and cystic space filled with necrotic debris.[1] On microscopy, SPT show cells arranged in the form of solid sheets, microcysts and pseudo-papillary areas which show characteristic pseudo-papillae with the fibrovascular stalks surrounded by several layers of polygonal epithelioid cells.[1,16] The nuclei are ovoid and folded with indistinct nucleoli and few mitoses.[16] Different sizes of cystic and hemorrhagic degenerations are also noted along with hyaline globules and collection of foamy cells.[1,16] The thick fibrovascular core often shows prominent mucinous changes.[1] It is not clear whether this tumor originated from ductal cells, acinar cells or primitive ones.[1,16] Immunohistochemistry (IHC) can be sent for the confirmation of diagnosis. Pancreatic lesions are positive for α_1-antitrypsin, α_1-antichymotrypsin, phospholipase A_2, CD10 and CD56 from which combination of β-catenin and CD10 and absence of e-cadherin is most specific for SPT.[1,5,6,7,12] These findings suggest that the tumor cells predominantly have exocrine features but has capacity for dual differentiation.[1] Some authors mention that positive immunoreactivity for Ki-67 may predict the malignant potential and poor outcome of SPT of pancreas but is not approved by other authors.[3]

SPT is a rare pancreatic exocrine tumor which has been increasingly spotted in recent years. Complete curative surgical resection is the mainstay of treatment as prognosis of the disease is very good even in metastatic disease and metastasis should not be considered the contraindication of the surgery.

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