Solid Pseudopapillary Tumor of Pancreas a Case Report

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Abstract
Solid pseudopapillary tumor of pancreas is rare neoplasm which occurs commonly in young females. It grows with indolent manner and complete surgical resection is standard of care. We report a case of 22 year old female presented to us with symptoms of dyspepsia, on examination she had lump in right upper quadrant, USG revealed hypoechogenic mass between liver and right kidney. CECT scan showed hypodense and heterogeneous mass arising from head of pancreas. Patient underwent pancreaticoduodenectomy with portal vein reconstruction. HPE report mentioned solid pseudopapillary tumor (SPN) of pancreas. After two years of follow up patients maintains good general condition with no signs of recurrence.

Keywords - pancreatic neoplasm, women, complete excision, good prognosis

Introduction
Solid pseudopapillary tumor of pancreas is uncommon tumor but with availability of modern imaging studies it’s incidence is apparently increasing. It is commonly seen in young females and with adequate resection it’s outcome is very good.

Case report
A 22 year old female presented in our outpatient department with symptoms of dyspepsia and vague discomfort in right upper quadrant of abdomen for several months with no history of trauma or any surgery. On examination she had palpable mass in right upper abdomen. Ultrasound examination
revealed heterogeneous hypoechogenic mass measuring 10 x 9 x 9 cm between liver and kidney, CT scan reported a rounded well circumscribed, heterogeneous mass at head of pancreas of 10 x 9 x 9 cm (Figure 1).

Figure 1: CT scan demonstrating large mass arising from head of pancreas

No liver metastasis, no enlarged lymph nodes nor invasion of surrounding structures noted by CT scan report. CEA and CA 19-9 levels were normal rest laboratory findings showed no abnormalities. So with the report of imaging studies pre operative diagnosis of solid pseudopapillary tumor of pancreas was suspected. Patient underwent elective surgery and pancreaticoduodenectomy was done. Intra-operatively surprisingly lesion was found to be infiltrating small length of portal vein (figure 2), so to achieve negative margin short segment of portal vein had to be excised with specimen and primary reconstruction of portal vein was done without tension. Rest of the reconstruction done in standard fashion. The resected specimen (Figure 3) was fixed in 10% formalin and submitted for histopathological examination.

Figure 2 Intraoperatively tumor is seen infiltrating to segment of portal vein

Figure 3 Resected specimen of pancreaticoduodenectomy showing solid cystic mass

Post operatively patient recovered well without any complication. Histopathological examination revealed solid pseudopapillary tumor of pancreas showing extensive hemorrhage and necrosis. On follow up of two years with clinical examination and combination of CT scan and ultrasonography at
6, 12 and 24 months interval there is no evidence of recurrence or metastasis.

Discussion

Cystic and solid cystic neoplasm of pancreas encompasses various different tumor types and among that Solid pseudopapillary neoplasms (SPN) of the pancreas is believed to be a uncommon tumor, which accounts for 1-2% of pancreatic neoplasms \[^1,2\]. Though Lichtenstein had first described this tumor\[^5\] but credit is largely shared by Frantz (1959) and Hamoudi et al (1970) who gave the pathologic description of this tumor, so this tumor is also referred sometimes as a Frantz tumors or Hamoudi-Frantz tumor\[^3,4\]. WHO defined and accepted this tumor as a separate entity with the name SPT( solid pseudopapillary tumor) in1996 and this nomenclature is still followed. Availability of modern imaging studies have proved that prevalence of this tumor is actually much more common than what was once believed. Among the several case reports and small serieses T. Papavramidis and S. Papavramidis have given the largest case series\[^1\]. As we experienced in this case, SPN usually affects young women with a female: male ratio of 10:1, and 25% cases are seen in pediatric population \[^1\] and it is diagnosed a decade earlier in females than their male counterparts (25 years versus 37 years)\[^7\]. As we noticed in our case, most of the patients with SPN of pancreas presents with non specific symptoms such as vague discomfort, mild abdominal pain or even palpable abdominal mass. As the growth of this tumor is very indolent, so the tumor commonly attains a large size and often the patients are detected incidentally on imaging studies may be advised for other reasons. Rarely tumor may present with poor appetite and nausea, loss of weight, vomiting, jaundice or hematemesis \[^8\]. Most commonly it is found at the tail followed by head and body of pancreas, rarely it could be multicentric and even extrapancreatic possibly because of synchronous tumor spread\[^1,9\]. Cell of origin for SPN is controversial, it is assumed to be arising either from primitive pancreatic cells or from female genital bud. It’s prominent histological feature is solid and pseudopapillary proliferation of homomorphous cells lacking increased mitoses or atypia. Beta catenin mutation, and abnormalities in the wnt pathway and E cadherin are found in growth of SPN\[^9,10\]. Imaging study with CT or MRI is usual way of diagnosis which would reveal a well circumscribed heterogeneous lesion with solid and cystic component demarcated by peripheral capsule. MRI is superior to CT in describing certain tissue characteristics, such as, cystic degeneration, haemorrhage and presence of a capsule. In high risk patients and those who would require complex resection image guided pre operative biopsy like EUS or CT guided FNA can be of great help in 60% to 70% \[^10\].

SPN usually has low malignant potential and an indolent growth pattern with excellent prognosis. So the aggressive surgical resection is always warranted unless there is medical contraindication and despite features of invasion of surrounding structures incomplete resection must be avoided to avoid recurrence. In patients undergoing complete resection overall 5 year survival rate has been reported near 97%. One should note that tumor size
is not the criteria for unresectability. The clear role of chemotherapy and radiotherapy in the management of SPN is unsubstantiated.

Conclusion
SPN is rare pancreatic tumor, because of availability of modern imaging studies more and more case are being reported. It usually affects young females, despite of large size it has got very good prognosis so aggressive surgical resection should always be attempted.

References