Solid pseudopapillary neoplasms of the pancreas clinical analysis of 8 cases from North India and review literature

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DOI: https://doi.org/10.33545/surgery.2019.v3.i1b.17

Abstract
Solid pseudopapillary neoplasm (SPN) is rare pancreatic neoplasm. Predominantly in young female, present with abdominal lump and pain. These tumors are indolent in nature. In our case series, 8 cases in 6 years period (2012 to 2018), with one of them is unusual site in retroperitoneum. Mean age of presentation was 25.6 years. Presenting symptom was dull aching pain (80%), lump in upper abdomen (65%). Mean size was 11.6 cm on imaging abdomen. All patients underwent surgery, one patient underwent preoperative FNAC due to unusual location. Mean postoperative hospital stay was 13.9 days with no mortality. By immunohistochemistry, all cases stained positive for vimentin, CD 10 and all were negative for cytokeratin and Ki67 <2 %. Mean follow up period of 57 months, with no recurrence and metastasis. Rarely these can also present at unusual site like retroperitoneum. After surgery patients have good long term survival.

Keywords: Solid pseudopapillary neoplasm (SPN), pancreas, clinical analysis, surgery, follow up, retroperitoneal SPN

Introduction
Solid pseudo papillary neoplasm (SPN) of the pancreas is a rare neoplasm with low malignant potential with unknown etiology, which represents 1-2% of pancreatic exocrine cancers and 5% of cystic pancreatic tumours [1-3]. Papillary cystic neoplasm was the first name given to this tumor by Virginia Frantz in 1959 [4]. Later on in 1970 Hamoudi separated it different entity on finding of the electron microscopic features of tumor [5]. SPN is classified as low malignant potential tumor of exocrine pancreas according to WHO classification given in1996 [6]. At present, term SPN is most widely used for this tumor [7].
In recent few years the detection of incidental small lesion of pancreas has been increased with the advent of high resolution non invasive imaging technique. So the incidental pancreatic lesion is around 0.2-0.7% in recent publication [8]. But still there is debate on etiology [7]. In our case series we are presenting clinical and histopathological features, immunohistochemistry and treatment, followed by discussion with review of literature.

Materials and Methods
Total 8 patient records were obtained by retrospective analysis from year 2012 to 2018 (6 years). All patients were females. As a imaging, triphasic CECT (pancreatic protocol) was done in all cases. CT guided FNAC was done in 1 patient with a unusual location, that was in retroperitoneum. The clinico-pathological, radiological, operative and survival data were obtained and analyzed. Follow up was done in all patients, 6 monthly for 2 years and then annually thereafter. Clinical evaluations, routine blood investigations, serum CEA, CA19-9, CECT abdomen and chest were done in follow up.

Results
Total patients were 8 and all were females with age group from 20 to 35 years (mean 25.6 years). 80% patient had dull aching pain in upper abdomen, 65% had lump in upper abdomen and 16% presented with early satiety. As location wise most common site was tail 37.5%, head and body with 25% each and one (12.5%) in retroperitoneum. On imaging mean size of tumor was 11.6 cm. All patients underwent surgery. Patients with head mass underwent classical pancreaticoduodenectomy. Patient with tail and body mass underwent distal pancreatectomy with splenectomy, one patient underwent enucleation of mass from retroperitoneum.
Mean hospital stay were 13.9 days. 2 patients (25%) developed grade A POPF (postoperative pancreatic fistula) , 2 patients (25%) developed grade A DGE (delayed gastric emptying). 1 patient (12.5%) had grade B POPF which was managed conservatively .There were no postoperative mortality .Patients were follow up with mean of 57 months with no recurrence or metastasis.

**Fig 1.1:** CECT scan of the abdomen showing the solid cystic lesion in the pancreatic tail and an area of cystic degeneration is noted centrally.

**Fig 1.2:** The specimen has been resected en bloc with the spleen through a open approach.

**Fig 2.1:** USG abdomen showing well encapsulated solid cystic lesion in head pancreas.

**Fig 2.2:** CECT scan of the abdomen showing the solid cystic lesion in the pancreatic head, abutting the portal vein and central cystic degenerated area seen.

**Fig 2.3:** The specimen of classical whipples procedure having head mass.

**Fig 3.1:** CECT abdomen revealing mass in retroperitoneum close to pancreatic tail and left kidney with cystic degeneration and multiple septa with peripheral enhancement.
Discussion
Solid pseudopapillary tumor are rare neoplasm of pancreas but recent advancement in imaging had lead to increase the prevalence of the disease [8]. It generally has a good prognosis after complete surgical resection [9]. More common in young females. A study of 718 patients having SPN showed that the head and tail disease is predominant site (more than 90%), with average age of 22 years [10]. In our study all patients were females and most common site was body and tail (fig 1.1, 1.2) followed by head (fig 2.1, 2.2, 2.3) and one at unusual location retroperitoneum (fig 3.1, 3.2, 3.3). There is dilemma about the origin of these tumor. Due to it female predominant nature, its origin is attributed to primordial pancreatic cells to the ovarian ridge during development [11]. Radiologically CECT and MRI both are almost equally diagnostic, MRI is slightly better than CT in identifying capsule, hemorrhage and cystic degeneration [12]. On CT scan these tumor are well encapsulated, hypodense with various solid cystic components [12]. On MRI feature are well defined lesions with high and low signal intensity on T1 and high signal intensity on T2 [12]. In our study triphasic CECT was done in all cases.

As the site is concern, this tumor involve any part of pancreas in which head and tail are more common locations [10, 12]. A study of 718 patients with SPN showed that most common site is tail 35.9% followed by head 34% and 10.3% in body and tail and least common and rare extra pancreatic site 1% [10]. In our study pancreatic tail was most common site and 1 case of retroperitoneal SPN was also present. Symptom with which patients present are of vague type which include increased abdominal girth, abdominal discomfort, abdominal pain, poor appetite, and nausea. These symptoms are due to involvement of adjacent organ stomach. Dull pain was most common symptoms in around 80% then followed by lump in upper abdomen in 65% in our study. On gross these tumor are well encapsulated (fig 1.2, 2.3, 3.3). On microscopic examination there is solid cystic mass with center of hemorrhagic and necrotic material and peripherally by solid tissue [14]. Pseudopapillary formation, foamy histiocytes, nuclear groove are characteristic finding of SPN [7]. On IHC they are characteristically positive for alfa 1-antitrypsin, CD56, CD10, and vimentin are present in 90% of patients [11]. In our study all the patients were stained strongly for vimentin, CD 10 and all were negative for cytokeratin and Ki 67 <2%. Malignant transformation is more common in elderly males.15% of adults and 13% of children are at risk of malignant transformation [12, 15]. On histology feature of malignant tumor are angioinvasion, perineural invasion, and deep pancreatic tissue invasion [12]. 10 to 15 % patient have metastasis to liver, lymph nodes and peritoneum [7, 12, 16, 17]. Tumor size more than 5 cm, angioinvasion are poor prognostic factors [7, 17, 18]. 5 year survival rate after complete resection of tumor is more than 95%, so even in the presence of local invasion, limited metastasis and recurrence complete resection is done [1, 17, 16]. In our study mean follow up was 57 months with no recurrence or distant metastasis. Surgical debulking is also done and in contrast to other pancreatic neoplasm this tumor is resected even after portal vein and superior mesenteric artery involvement [19, 20, 21]. Role of adjuvant therapy is unclear in SPN. Some study had shown role of gemcitabine and radiotherapy to downstage the tumor [22, 23].

Conclusion
Pancreatic SPNs are rare neoplasms with malignant potential. Primarily it is a young women disease. Differential diagnosis should be kept in mind when a young women present with pancreatic mass. FNAC has a role if there is dilemma in diagnosis. Surgical resection provide curative treatment with excellent long term prognosis. So patients with SPN should be treated aggressively with complete resection to achieve long term survival.

References

Fig 3.2: Open midline incision given through mass has been enucleated and removed.

Fig 3.3: Excised specimen of mass, well encapsulated.


