Solid Pseudopapillary Neoplasm of the Pancreas: Two Cases with Review of Literature

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Solid pseudopapillary neoplasm of pancreas (SPNP) is considered a rare tumour of pancreas[1]. The incidence is increasing, possibly due to an increase in the use of cross-sectional imaging and immunohistochemistry. It is most frequent in young females[2], usually occurs in the body-tail region of pancreas, compared to the head[3,4] and carries an excellent prognosis with surgical resection. We present two cases of SPNP.

Case 1

A 22 year-old-female patient presented with dull right upper abdomen pain for 1 year. She did not have jaundice, anorexia or weight loss. There was a palpable lump in the right hypochondrium. USG abdomen was suggestive of a large mass in the head of pancreas. Blood investigations, including liver function tests and pancreatic tumour markers, were within normal limits. CT scan abdomen with pancreatic protocol showed 11x10 cm size large hypodense mass in the pancreatic head, with peripheral enhancement and central necrosis (**Figure 1a**). The mass displaced the duodenum, bile duct and portal vein; and was touching the liver, right kidney and inferior vena cava. Based on clinical and imaging features, SPNP was suspected. As the patient was symptomatic, and the mass was resectable, the patient was offered pancreatoduodenectomy instead of endoscopic ultrasound (EUS) guided fine needle aspiration cytology (FNAC). She underwent classical Whipple's procedure (**Figure 1b**). Postoperatively she had chyle in the subhepatic drain, which was then left in situ for two weeks. She recovered well. Histopathological examination revealed features of SPNP (**Figure 1c**). IHC examination confirmed the diagnosis with tumour cells expression for β-catenin,

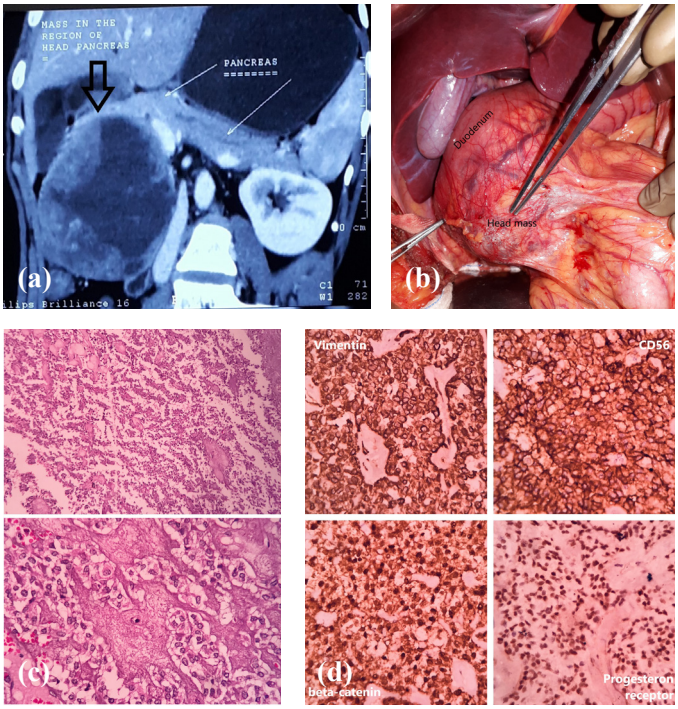

Figure 1 (a): CT scan of the abdomen, axial section, showing 11x10 cm size large hypodense mass in HOP, with peripheral enhancement and central necrosis; (b): Operative picture showing a large mass in the head of pancreas; (c): Microphotograph (upper one, 10x, HE staining) showing the papillary architecture of the tumour cells. Note the thick fibrovascular cores and medium-sized tumour cells lining the papillae. Some of the papillae have fallen away and seen as micropapillary groupings. Microphotograph (lower panel, 40x, HE staining) showing medium-sized tumour cells arranged around fibrovascular cores. These cells show round nuclear contour with vague convolutions. There is moderately abundant pale cytoplasm; (d): Cytoplasmic and membrane staining of Vimentin and CD56. Nuclear staining of β catenin and progesterone receptor (PR).

CD-56, vimentin and progesterone receptor (PR) (**Figure 2d**). We followed her up with cross-sectional imaging every 6 months. At 21 months of follow up, she did well without any recurrence.

Case 2

A 39 year-old-female patient developed fever and abdominal discomfort two weeks prior to presentation. USG abdomen revealed a large cystic mass in distal body-tail of the pancreas. Further evaluation with contrast-enhanced CT scan showed a large 10x8 cm size cystic lesion with enhancing wall and internal septation in the distal body of pancreas. To further clear the diagnostic dilemma, she underwent MRI abdomen (**Figure 2a** and **b**), which showed, 10x8 cm size complex cystic lesion with a peripheral hypointense rim on T2W images. The lesion revealed hypointense signals on T1W images and hyperintense signals on T2W images. Given the possibility of complex cystic neoplasm of pancreas, she underwent laparoscopic distal pancreatectomy (**Figure 2c**) and discharged on postoperative day 4. Histopathology examination revealedfeatures of SPNP, confirmed on IHC with tumour cells expressed β-catenin, Cyclin D1 and PR (**Figure 2d**). At seven months of follow up, she was doing well without recurrence.

Discussion

SPNP is considered a rare neoplasm of the pancreas; it consists of 2-3% of pancreatic neoplasms[1]. Although a recent increase in the incidence of SPNP is apparent, it may be due to the rise in the detection rate because of the increased use of cross-sectional imaging and IHC.

It is more common in young females, as were both of our patients; only 10% of SPNP occurs in males[2]. The origin of the tumour is debatable; and is often classified as an indolent tumour but potentially malignant. SPNP can occur anywhere in the pancreas, but it is more common in body and tail of pancreas compared to the head[3,4] one of our cases had a tumour in the head while other had in the body-tail region of the pancreas.

SPNP are either detected incidentally or present with a variety of non-specific symptoms like abdominal

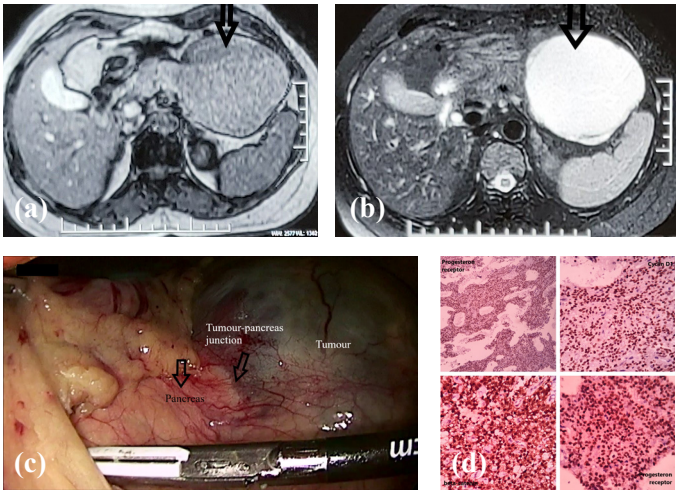

Figure 2 (a): MRI picture showing 10x8 cm size complex cystic lesion with hypointense signals on T1W; (b): MRI picture showing the lesion with hyperintense signals on T2W image with peripheral hypointense rim; (c): Intra-operative picture of laparoscopic distal pancreatectomy, note pancreas and the lesion; (d): Nuclear staining of progesterone receptor, cyclin D1 and β catenin.

pain, nausea, vomiting and rarely jaundice. Both of our caseswere symptomatic with abdominal pain, without jaundice despite the involvement of head of pancreas by a large tumour in case 1.

Among cross-sectional imaging, MRI is the preferred choice over a CT scan[5]. Characteristic findings on imaging include an encapsulated mass with solid and cystic components, there may be calcification or haemorrhage within the tumour. The solid areas enhance on pancreatic phase; slightly lesser than usual pancreatic parenchyma, the enhancement increases in delayed venous phase[6]. SPNP is usually oval in shape in females while lobulated in males[7]. EUS guided FNAC along with IHC helps in making preoperative diagnosis[8] where there is a dilemma, although in many cases; clinical and imaging information provides enough evidence to go ahead for surgery.

The overall prognosis of SPNP is excellent[9]. These tumours are indolent, with low malignant potential; up to 15% develop metastasis[4]. Young age, male sex, size more than 5 cm size, capsular invasion and high mitotic rate suggest possible malignant behaviour[10,11].

Surgical resection is the treatment of choice, parenchyma preservation with either enucleation or central pancreatectomy can be attemptedwhen feasible[4]. Spleen is preserved if there is no splenic vascular or splenic hilar involvement. Routine lymphadenectomy is not required as lymph node metastasis is rare[12]. Kim MJ *et al*. found a low recurrence rate with only 2 out of 106 patients having recurrence after a median follow up of 56.9 months[13]. Tang LH *et al*. reported a 97% five-year survival despite the presence of metastasis[9]. Role of adjuvant therapy in SPNP is unclear, while neoadjuvant treatment for down-staging of unresectable tumours using combination of gemcitabine and radiotherapy has been attempted[14].

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