SOLID PSEUDOPAPILLARY TUMOUR OF THE PANCREAS- A SINGLE INSTITUTION STUDY OF THREE CASES WITH A BRIEF REVIEW OF LITERATURE

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ABSTRACT

BACKGROUND

Solid Pseudopapillary Tumour (SPT) of the pancreas is a distinctive tumour of low malignant potential with a predilection for females in the second and third decades of life. We report here 3 cases because of its rarity and we attempt to briefly review the literature of this distinctive lesion.

MATERIALS AND METHODS

A total of 3 cases were diagnosed and treated in our institute over a 2-year period. A retrospective study of these cases was performed with respect to age group most frequently affected, sex ratio, common presenting symptoms and signs, investigative protocols, pathological features, treatment offered, outcome and prognosis. All the three patients were females with the age of presentation of 20 yrs., 14 yrs. and 15 yrs. respectively. The commonest presenting symptom was abdominal pain. The patients underwent distal pancreatectomy, Whipple's resection and pancreaticoduodenectomy with appendicectomy respectively. Histological diagnosis was made by performing haematoxylin-eosin staining and confirmed by immunohistochemistry.

RESULTS

All the patients were females in the age group of 13 - 25 years. Pre-operative imaging is not diagnostic. Pathological examination is the mainstay in the diagnosis. No metastasis or recurrence was detected during followup.

CONCLUSION

Solid pseudopapillary tumour of the pancreas is a rare entity, therefore proper diagnosis, evaluation and formulation of treatment protocols is difficult. A high index of clinical suspicion is necessary to suspect and diagnose it. Surgical excision offers the best chance for cure and should always be attempted irrespective of the magnitude of the resection involved. The results of immunohistochemical and ultrastructural studies indicate that it must be regarded as a tumour of uncertain histogenesis and undetermined differentiation.

KEYWORDS

Solid, Pseudopapillary, Neoplasm, Pancreas.

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BACKGROUND

Solid Pseudopapillary Tumour (SPT) is a rare primary exocrine pancreatic neoplasm occurring in about 1% of pancreatic tumours.[1] Most cases are found in young women in second to fourth decade of life[2,3] with non-specific symptoms including abdominal discomfort, subtle abdominal pain or abdominal mass. It is a well encapsulated, relatively benign tumour having a favourable prognosis. The presence of extrapancreatic invasion, distant metastasis, pancreatic parenchymal invasion, perineural or vascular invasion have been considered to be features of malignancy.

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SPT of the pancreas most commonly arise in the pancreas, while retroperitoneum is the second site of involvement.[4] We report here three cases because it is important to differentiate this tumour from other pancreatic neoplasms, as this type is amenable to cure with surgical resection, even in cases with capsular invasion, unlike malignant tumours of pancreas.

MATERIALS AND METHODS

Clinical records with follow-up information were studied for 3 cases of SPT of the pancreas, diagnosed and treated in our hospital between 2015 and 2017. The clinico-pathological, radiological, operative and survival data were obtained. A CT scan of the abdomen was performed in all the cases and the findings revealed a mass in the pancreas. Histological diagnosis was made by performing haematoxylin-eosin staining and confirmed by immunohistochemistry using a panel of markers. All the patients were followed up every 6 months. Routine blood tests, biochemical analysis, chest x-ray, ultrasound or a CT scan abdomen along with CA 19-9 were performed in the follow-up period.

RESULTS

All the patients were females in the age group of 14 to 20 years (median 15 years). Two patients presented with pain abdomen and abdominal lump gradually increasing in size (67%). One presented with a painless abdominal mass (33%) in the epigastrium. The tumour was located in the head of the pancreas in two cases (67%) and in the tail in one case (33%). The tumour size ranged from 6 to 10 cm (average 7.3 cm).

The patients underwent distal pancreatectomy, Whipple's resection and pancreaticoduodenectomy with appendicectomy respectively. The postoperative hospital stay was uneventful. All the 3 patients are disease free in the follow-up period. Details of the cases are summarised in Table 1 and Table 2.

Case	Age (yrs.)	Sex	Location	Size (cm)	Treatment	Follow-up
1	20	Female	Tail	6	Distal pancreatectomy	24 months - no evidence of disease
2	14	Female	Head	10	Whipple's procedure	10 months - no evidence of disease
3	15	Female	Head	6	Pancreatectomy with appendicectomy	16 months - no evidence of disease

Table 1. Summary of Clinical Findings

Marker	Case 1	Case 2	Case3
Alpha-1 antitrypsin	+ve	+ve	Not included
Beta catenin	Not included	+ve	+ve
CD 56	Strongly +ve	Strongly +ve	Strongly +ve
CD 10	Weakly +ve	Weakly +ve	Strongly +ve
Vimentin	+ve	+ve	Not included
Synaptophysin	-ve	-ve	-ve
Chromogranin	-ve	-ve	-ve
Progesterone Receptor	Not included	Not included	Strongly +ve
Cyclin D1	Not included	Not included	Strongly +ve
CK 8/ CK 18	-ve	-ve	-ve

Table 2. Immunohistochemical Study

DISCUSSION

SPT of the pancreas was first described by Frantz in 1959.[5] It predominantly occurs in young females, but has been reported in males and children. Zhou et al reported that SPT was probably the most common pancreatic tumour in the Asian paediatric population.[6] In our series, all patients were females with a median age of 15 years [Table 1]. The most common clinical presentation is a palpable abdominal mass and abdominal pain.[7] Depending on the location of the tumour in the pancreas (head, body or tail), the differential diagnosis includes pancreatic endocrine tumours, mucinous cystadenoma, acinar cell carcinoma and pseudocyst.

Preoperative imaging is not diagnostic. However, ultrasonography, CT scan, MRI and endosonography are the commonly used radiological modalities. CT scan is more sensitive and specific and has shown greater accuracy in diagnosing SPT. In our series, CT scan was performed in all the 3 cases.

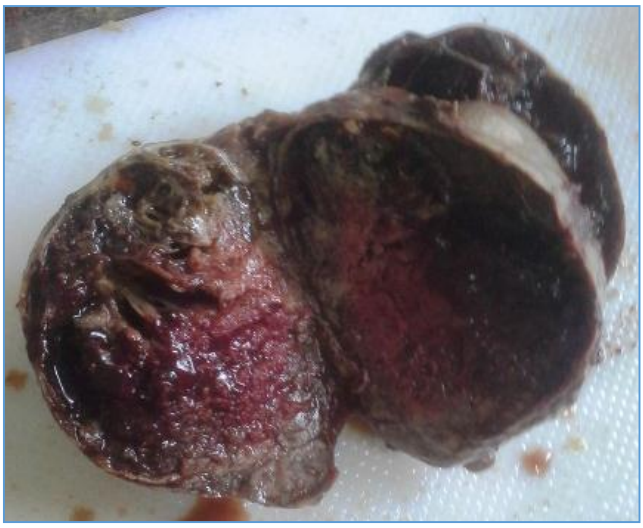

Figure 1. Encapsulated Solid/ Cystic Mass showing Haemorrhagic and Necrotic Areas

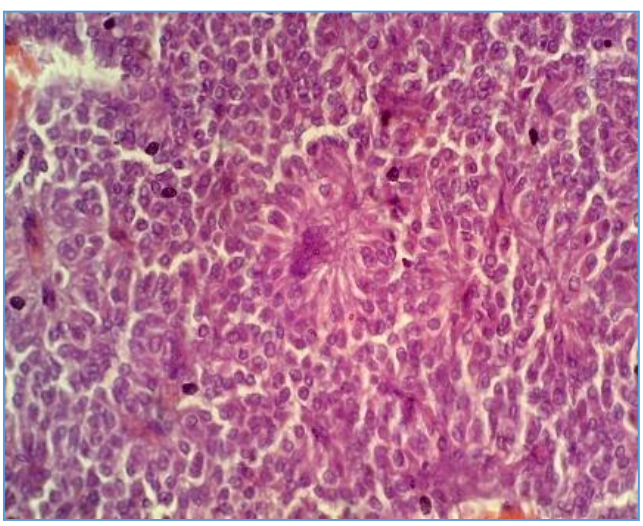

Figure 2. H and E (40X) Cells with Granular, Eosinophilic Cytoplasm and Round to Oval, Mildly Pleomorphic Nuclei. Mitotic Figures and Pseudorosettes Seen

Pathological examination is the mainstay in the diagnosis. Grossly, SPT is a well-encapsulated tumour. Cut section shows a variegated tumour with haemorrhagic and necrotic areas admixed with solid areas [Fig. 1]. Microscopically, the solid areas are composed of small and medium sized tumour cells. Tumour is very cellular and shows the presence of pseudopapilla covered by layers of epithelial cells. Nuclei is ovoid and folded with indistinct nucleoli and few mitosis [Fig.2]. It is worth noting that cystic changes are not a ubiquitous feature and instead usually occur in larger lesions secondary to long-standing tumour necrosis. The papillary appearance of the tumour is due to the cellular clustering around the microvasculature with more dyscohesive cells in the periphery and is not due to the presence of true papillary stalks. Immunohistochemical studies have shown that SPT is reactive for alpha-1 antitrypsin, beta catenin, CD 56 [Fig. 3], CD 10, vimentin [Fig. 4], progesterone receptor, S100 protein,

cytokeratin, desmoplakin, trypsin, chymotrypsin and amylase [Table 2].

Focal positivity has been observed with NSE and islet cell hormones, such as insulin and glucagon. This points that SPT arises from primitive pancreatic cells having dual differentiation, but with a predominance of exocrine features.[8,9] Presence of progesterone receptors with its predilection for females suggests that SPT is a hormone dependent tumour.[10,11] Histogenesis of SPT is unknown. Ultrastructurally evidence of acinar, ductal and endocrine cell differentiation has been found.

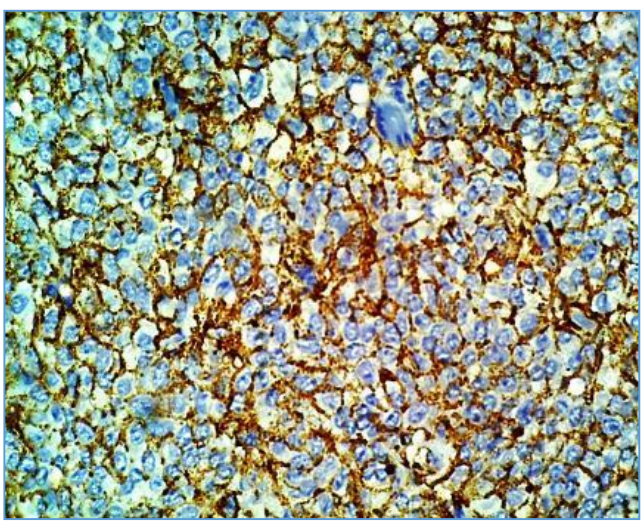

Figure 3. CD 56 Positive

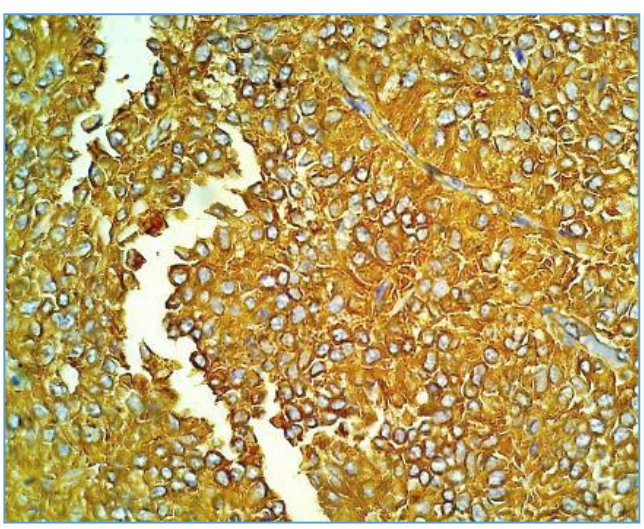

Figure 4. Vimentin Positive

Treatment of choice is complete surgical resection. A study from Memorial Sloan-Kettering Cancer Center, New York, USA, recommends complete surgical excision with even metastatectomy if required.[12] In our study, none of the cases had metastasis. A local recurrence rate of 6.2% is reported in cases treated by radical surgical excision.[13] In our series, all the patients are disease free on follow-up with no evidence of recurrence. SPT has an excellent prognosis with a low malignant potential. Aggressiveness is associated with cellular atypia, mitotic activity and lymphovascular, perineural or invasion of neighbouring organs. Metastasis is seen in regional lymph nodes, liver and omentum. On analysing our study, the results are in close proximity to the earlier studies conducted by Patil TB et al[14] and Ozguven BY et al[15] [Table 3].

Criteria	Patil et al	Ozguven et al	Present Study
Cases	14	09	03
Duration of study	10 years	15 years	02 years
Sex	All females	8 females, 1 male	All females
Age (yrs.)	13-45 (median 20)	14-73 (median 32)	14-20 (median15)
Site	Equivocal (1:1)	Tail (7:2)	Head (2:1)
Size (cm)	3-18 (average 6.8)	4-9 (average 5.8)	6-10 (average 7.3)
Post-operative morbidity	01 case	Nil	Nil
Post-operative mortality	Nil	Nil	Nil

Table 3. Comparison between the Parameters in the Present Study with those Reported in Two Previous Studies

CONCLUSION

Solid pseudopapillary tumours have a low malignant potential and their prognosis is extremely good unlike other pancreatic tumours. A high degree of clinical suspicion is necessary to suspect and diagnose SPT. The widespread use of imaging techniques together with a better knowledge of the disease has prompted a rapidly increasing number of case reports. Surgical excision offers the best chance for cure and should always be attempted irrespective of the magnitude of the resection involved. Immunohistochemical and ultrastructural studies indicate that it must be regarded as a tumour of uncertain histogenesis and undetermined differentiation.

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