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Research Paper

Solid Pseudo Papillary Neoplasm of Pancreas – A Rare Variant

Mathematics

KEYWORDS : Abdominal Tumour, Immunohistochemistry, Pancreas, Pseudo papillary tumor

Dr. Rajani. Mutyala	Department of Pathology, Guntur Medical College, Guntur, Andhra Pradesh, India
G. Sailabala	Department of Pathology, Guntur Medical College, Guntur, Andhra Pradesh, India
Subbarao P	Consultant Pediatric Surgeon, Guntur
Nageswararao K	Department of Pathology, Guntur Medical College, Guntur, Andhra Pradesh, India

ABSTRACT

Solid Pseudopapillary neoplasm (SPPN) of Pancreas is a rare tumor usually benign described mostly in young women. We here report a case of SPPN in a 12 yr old child. It is important to differentiate this tumor from other pancreatic neoplasmsas it is a low malignant potential tumor and its complete surgical resection is curative.

INTRODUCTION:

Solid pseudopapillary neoplasm (SPPN) is a rare tumor, mostly encountered in young women, first described by Frantz way back in 1959 (1,2). Its various synonyms include papillary cystic neoplasm, papillary epithelial neoplasm, papillary and cystic tumor, papillary and cystic epithelial carcinoma, papillary and solid neoplasm, solid and cystic acinar cell tumor and Gruber – Frantz`s tumor (3). This tumor is low malignant potential (2,4). It offers diagnostic and therapeutic challenges because of its rarity (4). It occurs predominantly in adolescent girls and in young females with mean age of 20-25yrs. It is rare in men (3,5) we here in report a case of SPPN in a 12 yr old girl.

CASE REPORT:

A 12 yrs old girl presented with abdominal pain, nausea, obstructive jaundice. Ultrasound and MRI images showed 4 cms lesion in the head and uncinate process of pancreas with pressure effect onduodenum, CBD and pancreatic duct.

MACROSCOPIC APPEARANCE -

Received Whipples resection specimen (Pancreatico – duodenectomy) of size 14x8x5 cm cut section of pancreatic head shows large well circumscribed grey white mass with extensive areas of necrosis and hemorrhages.[Fig1&,2]

Figure1

Figure 2

MICROSCOPY :

Sections studied from tumor shows round to oval cells arranged in papillary pattern, peri vascular arrangement, gland pattern and occasional rosettes. Extensive areas of necrosis is seen. No evidence of any mitotic activity or vascular invasion[Fig 3 & 4]. Sections from duodenum show mild chronic non specific inflammation. No evidence of any tumor invasion. Possibilities considered are Solid pseudo papillary neoplasmand Acinar cell carcinoma

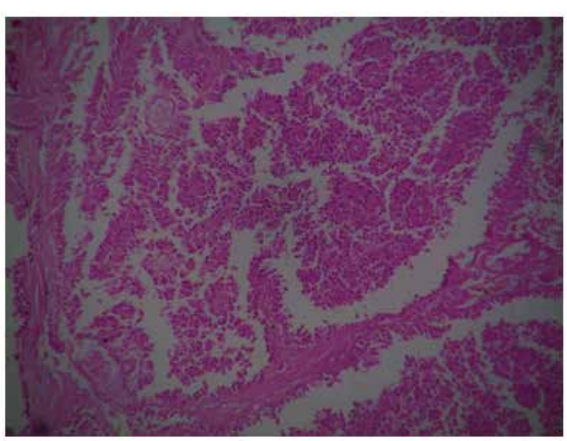

Figure 3

H&E Papillary areas

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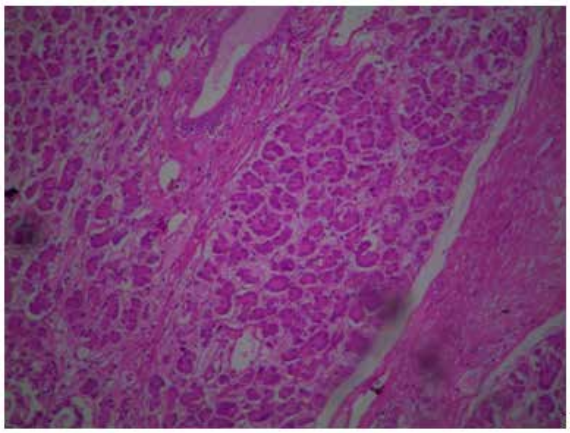

Figure 4
H&E Normal pancreatic Acini

Immunohistochemistry markers study was done with CDIO and Vimentin which were positive and confirmed the diagnosis of SPPN (fig5,6)IHCI

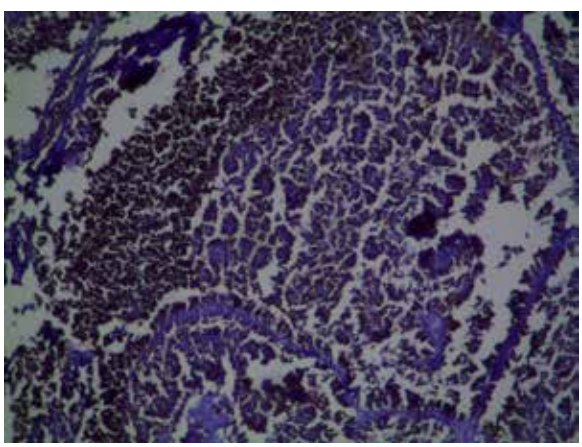

Figure 5
IHC Marker CD 10 Positive

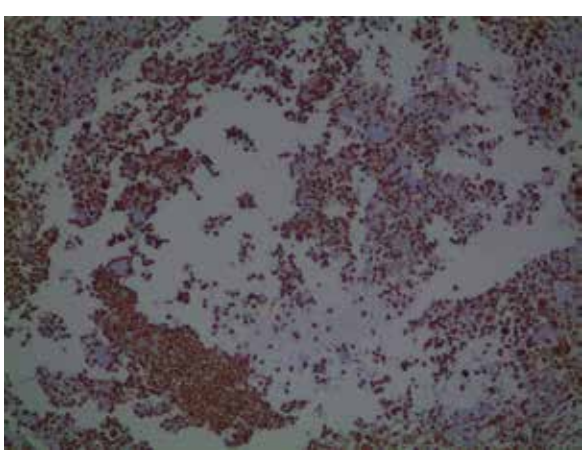

Figure 6

IHC Marker Vimentin Positive

DISCUSSION:

SPPN is rare tumor. It accounts for 1-2 % of all exocrine pancreatic tumors (3,5). Most commonly seen in adolescent girls and young women with mean age 25 yrs (3,5). In our case it is seen in a 12 yrs old girl. Large SPPN may cause symptoms like abdominal pain, nausea and jaundice. These tumor are not associated with endocrine syndrome (2,3.6-9). In our case similar complaints were observed.

Imaging studies like ultrasonography and MRI often reveal classic imaging characteristics of SPPN like large size mixed solid and cystic nature, encapsulation and hemorrhage (10-12). On imaging showed 4cms lesion in the head and uncinate process of pancreas with pressure effect on duodenum, CBD and pancreatic duct.

Zeytunlu et.al in their study of four cases of SPPN have described them in young women presenting with nonspecific symptoms located in the body and tail of pancreas as large tumors (13).

The pre-operative diagnosis of SPPN is possible by means of fine needle aspiration cytology, which reveals loose aggregates of small monotonous cells with scant cytoplasm surrounding thin walled capillaries. There is also characteristic presence of pseudo papillae intra cytoplasmic globules, stripped nuclei and non necrotic back ground which helps in cytological diagnosis of SPPN (3,14). However in our case FNAC was not done.

Grossly it is usually a well circumscribed often encapsulated tumor ranging from 3-18 cm in diameter. Its cut section has a variegated appearance with solid, cystic and papillary areas with necrosis and hemorrhages. (3,15)The tumor in our case is 4cms size grey white with areas of hemorrhages and necrosis on cut surface.

Microscopically these tumors are well circumscribed and usually encapsulated. Extensive necrosis and degenerative changes are common. The tumor cells are arranged mostly in pseudo papillary and with occasional monomorphic pattern. The nuclei are uniform and round with even chromatin pattern and small nucleoli. Often nuclear grooves are seen. They have low mitotic activity and usually do not have perineural and vascular invasion (3,15). In our case extensive areas of necrosis were observed in addition to the characterstic microscopic features like presence of Pseudo-papillae. The individual cells were round to oval with bland chromatin. It has insignificant mitotic activity with no vascular invasion or perineural invasion. Differential diagnosis includes Acinic cell carcinoma. Acinic cell carcinoma occurs in older age group and tumor lack pseudo papillary areas. Immunohistochemically Acinic cell carcinoma typically stains for trypsin which distinguishes from SPPN (14)

The histogenesis of SPPN remains uncertain without any evidence for ductal, acinar or frank endocrine differentiation (15) SPPN shows variable IHC expression for various epithelial, mesenchymal and endocrine markers (3,15).

We noted in our case of SPPN CD 10 and vimentin markers positivity.

CONCLUSION :

SPPN are rare tumors and should be considered in the differential diagnosis of Acinic cell carcinoma and other cystic pancreatic tumors. Its characteristic age manifestations, gross microscopic findings and immunohisto chemistry help in diagnosis. Further study with better follow –up is desired to characterize these common tumors.

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