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Pathology

SOLID CYSTIC PSEUDOPAPILLARY NEOPLASM (SPN) OF THE PANCREAS IN A PAEDIATRIC PATIENT : A RARE CASE REPORT.

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ABSTRACT Solid cystic pseudopapillary neoplasm of the pancreas is an extremely rare tumor of the exocrine pancreas, occurring predominantly in young women. SPN has a low malignant potential and patient can expect an excellent prognosis after adequate surgical excision. Awareness of this tumor entity with good prognosis is crucial to make proper diagnosis to offer better treatment and reduce morbidity. We report here on a previously healthy 12 year old female presented to our institution with lump in the left side of upper abdomen and abdominal pain for 8 months. Imaging revealed a solid & cystic mass arising from the distal body and head of pancreas. Immunohistochemical staining (IHC) shows positive for CD10 and PR. Final histopathologic evaluation following resection confirmed this lesion to be a Solid Cystic Pseudopapillary Neoplasm(SPN) of pancreas.

KEYWORDS : Child, Frantz's tumor , IHC, Solid Cystic Pseudopapillary Neoplasm

INTRODUCTION

Solid cystic pseudopapillary neoplasm(SPN) of the pancreas is a very rarely diagnosed tumor, representing less than 2% of all exocrine pancreatic neoplasms.(1). It almost exclusively seen in young women of all races and has been mistaken for an endocrine neoplasm or a cystic tumor. Rarely it is seen in paediatric patient . This tumor was first described in 1959 by Virginia Frantz as "papillary cystic tumor of the pancreas". The patient was a 2 years old boy undergone to pancreatic-duodenectomy [2] . It was finally defined by the World Health Organization (WHO) in 2000 as a solid pseudopapillary neoplasm of the pancreas (3).

This tumor are characterized by solid and pseudopapillary growth pattern covered by poorly cohesive uniform cells. The origin of this neoplasm is not well known but it has been postulates that this tumor may arise from pluripotent embryonic cells of the pancreas or from the ridge ovarian analog-related cells, which were attached to the pancreatic tissue during early embryo-genesis [4].

Case report

A previously healthy 12 -year- old female child presented to our institution with lump in the left side of upper abdomen and on & off abdominal pain for 8 months. **CT scan** revealed a pancreatic solid & cystic mass arising from the distal body and tail of the pancreas. **MRI** further characterised the lesion as enhancing round lesion in relation to the tail of the pancreas extending upto the lesser sac, suggestive of either solid pseudopapillary tumor , pancreatoblastoma or pseudocyst of pancreas. **Fine needle aspiration cytology** revealed degenerated, hyperplastic cellular infiltrate. A distal pancreatectomy was done and sent for **histopathological examination**. We received a globular, encapsulated specimen measuring (10x9x5.5)cm^3. The cut surface are soft, composed predominantly of solid areas with few cystic areas, frequently with degenerative cystic foci, hemorrhage, and necrosis.

FIG: Gross appearance- the tumor is encapsulated as well as show predominantly solid and cystic cut surfaces.

Microscopic examination:

Sections studied show a solid , trabecular and pseudopapillary pattern formed by poorly cohesive round to polygonal cells with small uniform nuclei , inconspicious nucleoli having nuclear grooves at places. The cytoplasm is vacuolated to eosinophilic with extracellular eosinophilic hyaline globules.

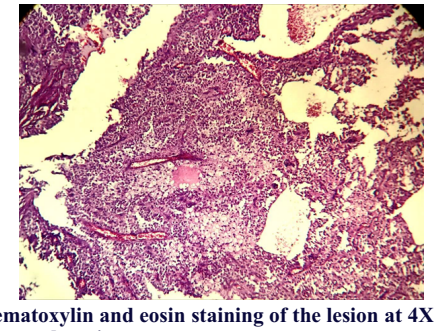

FIG: Hematoxylin and eosin staining of the lesion at 4X showing solid areas and cystic spaces.

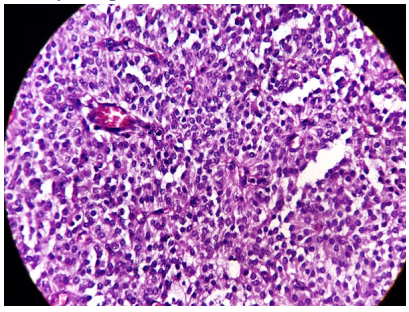

FIG: haematoxylin and eosin stain, 40X ; Uniform poorly cohesive cells with formation of pseudopapillae

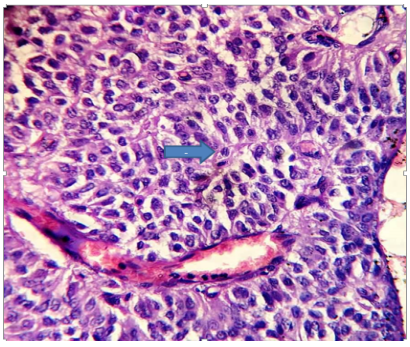

FIG: haematoxylin and eosin , 40X showing nuclear groove.

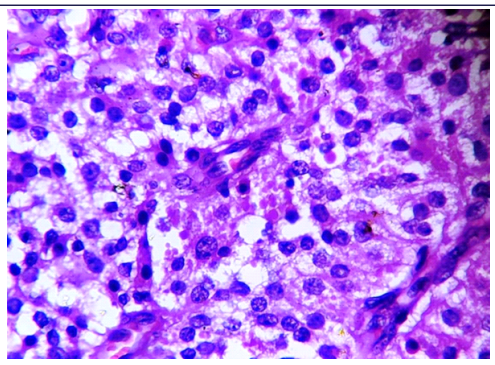

FIG: haematoxylin and eosin stain, 40X showing extracytoplasmic hyaline globules.

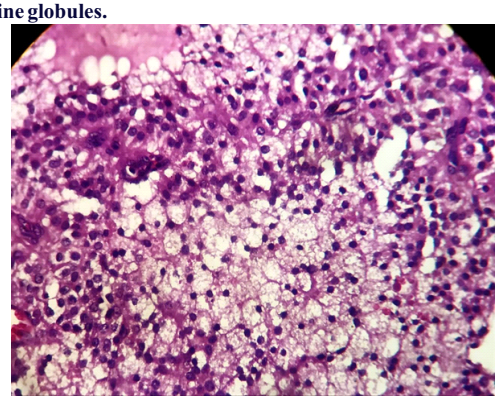

FIG: haematoxylin and eosin stain, 40X showing foamy macrophages.

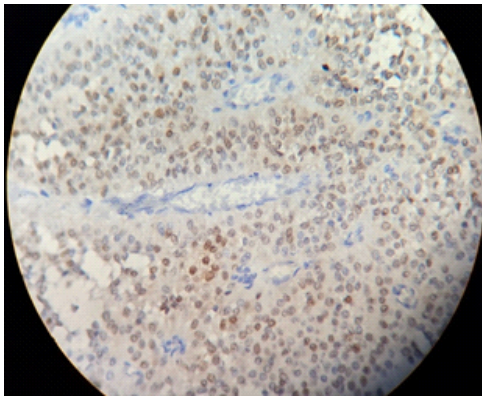

Fig: Immunoreactive for PR

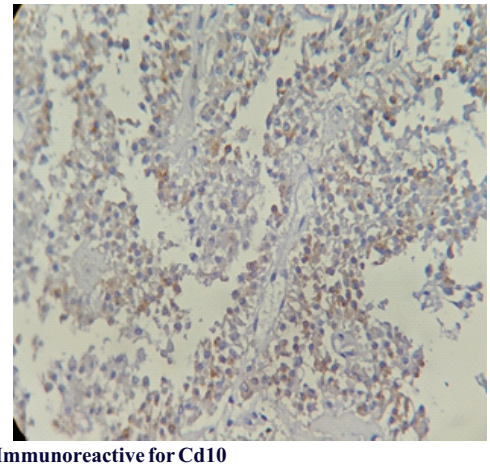

Fig: Immunoreactive for Cd10

DISCUSSION

SPN is defined by WHO as a usually benign tumour . It affect 80% women and 85% the patients under 30 years old. Rarely it occurs in males or in pediatric people [5]. Patients present with few nonspecific digestive-type symptoms, including abdominal discomfort or pain, dyspepsia, and bloating accompanied by an enlarging mass (6). The common localization of solid cystic pseudopapillary neoplasm is the tail of the pancreas , followed by head and body.

Immunohistochemically, the tumour is intensely and diffusely positive for vimentin, CD10 and CD56 (but chromogranin is characteristically negative), oestrogen and progesterone receptor, and focally positive for neuroendocrine markers. Chymotrypsin and trypsin also stain positive. Others positive stains include: nuclear and cytoplasmic beta-catenin, cyclin D1, nuclear E-cadherin, and paranuclear dot-like CD99. Positivity for progesterone receptor results in predilection of SPN for females.

The prognosis of patient with SPN is good, with overall 5-year survival rate of more than 95% (7,8). Surgical resection remains the main treatment for SPN most commonly local resection, distal pancreatectomy or pancreatoduodenectomy. Paediatric patients reported in the current literature have typically presented with resectable disease, with an overall five-year survival. Long-term clinical follow-up is recommended(9)

CONCLUSION

Although Solid Cystic Pseudopapillary Neoplasm of the pancreas is a rare form of pancreatic neoplasm, but it is important to remember that paediatric patients with pancreatic neoplasms are more likely to have SPN than other diagnosis. If completely resected, SPNs behave in a benign fashion in the vast majority of patients. However, approximately 15% of the reported cases have resulted in local recurrence and/or liver metastases, but even patients with metastatic tumors often do well. Therefore, SPN should be regarded as a carcinoma of low malignant potential. Histological and immunohistochemical features are well-defined and sufficient for a definitive diagnosis.

REFERENCES:

1. Andren-Sandberg A, Dervenis C. Pancreatic pseudocysts in the 21st century. Part I: classification, pathophysiology, anatomic considerations and treatment. JOP 2004;5(1):8–24.
2. V.K. Frantz, Tumors of the pancreas, in: C.W. Bumberg (Ed.), Atlas of TumorPathology, VII. Fascicles 27 and 28, Armed Forced Institute of Pathology,Washington, DC, 1959, pp. 32–33.
3. Klöppel G, Solcia E, Longnecker DS, et al. Histological typing of tumours of the exocrine pancreas: World Health Organization international histological classification of tumours [Internet]. New York: Springer-Verlag; 1998 [cited 2015 Feb 7]. Available from: http:// www.joplink.net/prev/201007/ref/08-004. html.
4. Kosmahl M, Seada LS, Janig U, Harms D, Klöppel G. Solid–pseudopapillary tumor of the pancreas: its origin revisited. Virchows Arch 2000, 436: 473–480.
5. Nikolaos Vassos, Abbas Agaimy, Peter Klein, Werner Hohenberger, Roland S.Croner, Solid-pseudopapillary neoplasm (SPN) of the pancreas: case seriesand literature review on an enigmatic entity, Int. J. Clin. Exp. Pathol. 6 (6)(2013) 1051–1059
6. Yu PF, Hu ZH, Wang XB, et al. Solid pseudopapillary tumor of the pancreas: a review of 553 cases in Chinese literature. World J Gastroenterol 2010;16(10):1209–1214.
7. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: Review of 718 patients reported in English literature. J Am Coll Surg. 2005;200(6):965–72.
8. Nakagohri T, Kinoshita T, Konishi M, et al. Surgical outcome of solid pseudopapillary tumor of the pancreas. J Hepatobiliary Pancreat Surg. 2008;15(3):318–21.
9. Butte JM, Brennan MF, Gönen M, et al. Solid pseudopapillary tumors of the pancreas. Clinical features, surgical outcomes, and long-term survival in 45 consecutive patients from a single center. J Gastrointest Surg 2011;15(2):350–357.