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SOLID - PSEUDOPAPILLARY NEOPLASM OF PANCREAS- A RARECASE REPORT

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Abstract :
Solid-pseudopapillary neoplasm of pancreas is a very rare entity of unknown pathogenesis . It is reported at an incidence of less than 2 percent of all pancreatic tumors , almost exclusively seen in adolescent girls and young women in the second or third decades. These tumors with low grade malignant potential carries excellent prognosis. Herein, we report a case of solid-pseudopapillary neoplasm of pancreas in 22 years old female admitted with vague abdominal pain of 4 months duration. CT abdomen revealed a well defined heterogenous mass arising from the tail of pancreas. Distal pancreatectomy with splenectomy was done. Histopathological examination and Immunohistochemistry confirmed the diagnosis.

Keyword :
Solid - pseudopapillary neoplasm, pancreas, Distal pancreatectomy, splenectomy.

INTRODUCTION:

Solid- pseudopapillary neoplasm of pancreas is a rare tumor of uncertain cellular differentiation that accounts for <2% of all exocrine pancreatic tumors[1,6]. Also known as solid and papillary epithelial neoplasm, solid-cystic papillary tumor, papillary cystic tumor or Gruber-Frantz tumor[2]. It is almost exclusively seen in adolescent girls and young women in the second or third decades and rare in children,older women,and men.They are detected incidentally or present with abdominal pain and discomfort .These tumors have a low grade malignant potential and carries excellent prognosis.We reported a case of Solid pseudopapillary neoplasm of pancreas by histopathological examination and immunohistochemical analysis in a 22 year female who presented with vague abdominal pain of 4 months duration.

CASE HISTORY:

A 22 year old female presented to the surgery department with vague abdominal pain of four months duration. On

examination,the patient was afebrile ,had normal vital signs and epigastric tenderness. Serum amylase level was normal (65U/L). CT abdomen revealed a well defined heterogenous mass with necrosis arising from the tail of pancreas.Distal pancreatectomy with splenectomy was performed and the specimen was sent for histopathological analysisin a 22 year female who presented with vague abdominal pain of 4 months duration.

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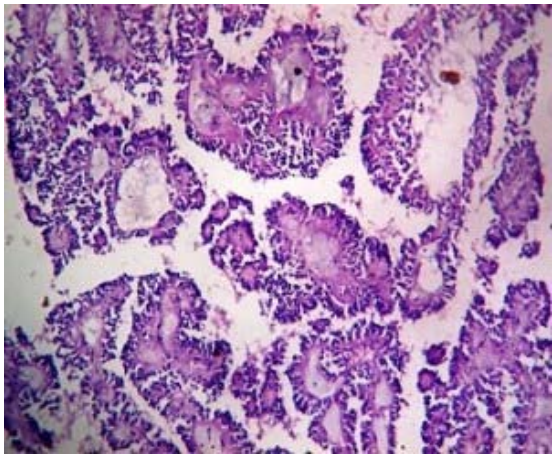
GROSS EXAMINATION:

Figure 1-Gross appearance- grey brown, grey yellow ,bosselated soft tissue mass with intact capsule and attached spleen. Cut surface shows a well circumscribed, solid yellow-brown mass with few mucin filled cystic spaces admixed with hemorrhage, necrosis and xanthomatous areas.

Recieved a grey brown, grey yellow ,bosselated soft tissue mass measuring 15x8x5cm with intact capsule and attached spleen measuring 14x9x4cm. Cut surface of the soft tissue mass revealed encapsulated, well circumscribed, solid yellow-brown mass with few mucin filled cystic spaces admixed with hemorrhage, necrosis and xanthomatous areas. Grossly there was no invasion of splenic parenchyma. (Figure1)



MICROSCOPIC FEATURES:

Figure 2- Tumor cells arranged in a pseudopapillary pattern-10 X.

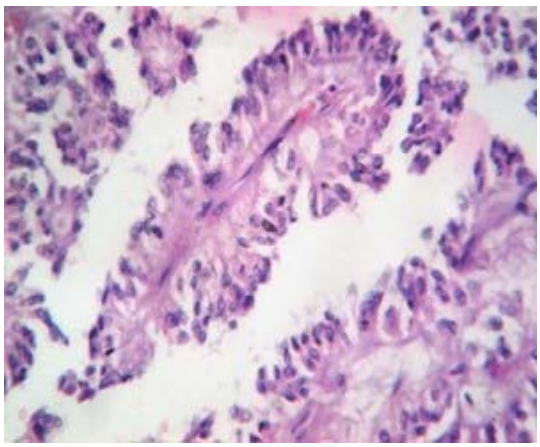
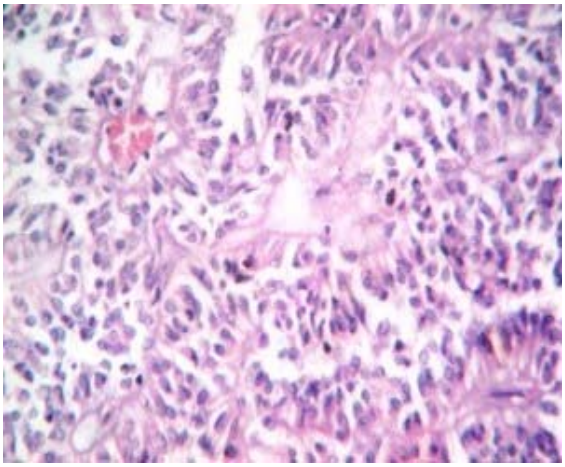


Figure 3-Pseudopapillae -40 X.
Sections from the soft tissue mass rvealed tumor cells arranged in solid nests and pseudopapillary pattern(Figure 2&3)
Tumor cells around small blood vessels (Figure 4) .



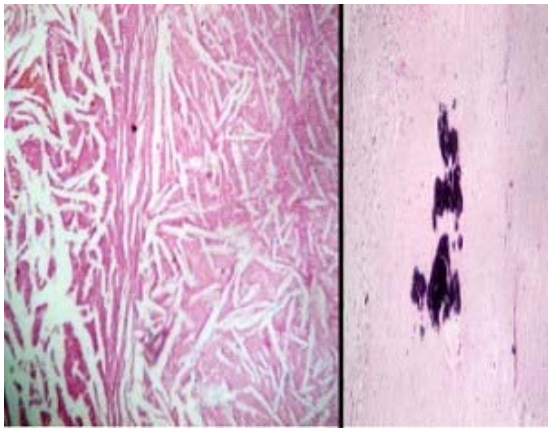
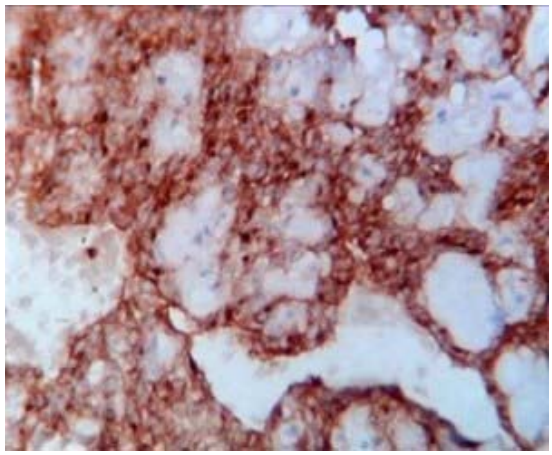


Figure 4- Tumor cells around fibrovascular core-40 X.
Figure 5- Cholestrol clefts & Calcification -10 X.
The cells are uniform, small, polygonal with clear to eosinophilic cytoplasm, ovoid nuclei, inconspicuous nucleoli and occasional nuclear grooves. Cholesterol clefts and psammoma bodies seen(Figure 5)
Final diagnosis of Solid-pseudopapillary neoplasm of pancreas was made..

IMMUNOPROFILE:
The tumour cells exhibited diffuse cytoplasmic positivity for CD10 (Figure 6) and Vimentin(Figure 7) .



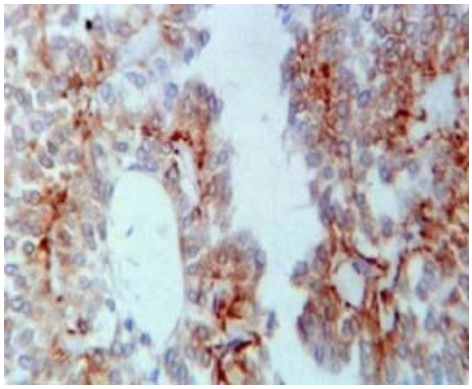


Figure 6-IHC- CD 10 positive-40 X.
Figure 7- IHC -Vimentin positive-40 X.

FOLLOW UP:
In this case we followed the patient in three consecutive visits and the patient was found to be asymptomatic and there was no evidence of recuence.

DISCUSSION:
Solid-pseudopapillary neoplasms of pancreas are rare, constituting <2% of exocrine pancreatic tumors[1,6,7].It arises from an uncommitted cell, similar to intercalated duct cells or centroacinar cells.

Consistent -catenin gene abnormalities are detected[1]. Mostly affects adolescent girls and young women in their second and third decades[3,4,5]. The body and tail are most frequently affected[2,10]. Mostly, they are detected on routine physical examination or on imaging study; sometimes present with abdominal discomfort and pain, or occasionally following abdominal trauma. Usually no abnormalities in laboratory tests (eg, serum amylase) or in pancreatic cancer markers (eg, CA19–9, fetoprotein, carcinoembryonic Antigen) are detected.[3,8,11]

Grossly, most of the tumors present as >10cm diameter in size[(1,2,6,7,8,14)], round, well demarcated encapsulated mass. The cut surfaces are typically soft, friable , well circumscribed with variegated appearance exhibiting cystic, solid and solid-cystic areas, frequently with hemorrhage, degenerative cystic foci and necrosis.

Microscopic examination reveals a variety of patterns- solid,cystic, pseudopapillary, pseudomicrocystic and trabecular [(1,2,6,7,8,14)]. The basic architecture is solid nests of cells with small blood vessels. The solid area contains sheets, cords and trabeculae of monomorphic, uniform, tumor cells seperated by vascular hyalinized stroma. The cells adjacent to the blood vessels are attched to the stroma and are intact, while those farthest from the blood vessels are detached from each other, undergo swelling and degenrative changes, resulting in a pseudopapillary cystic pattern.Slit like spaces seperate the pseudopapillae. Foamy macrophages, cholesterol crystals surrounded by foreign body giant cells and areas of haemorrhage are seen. Myxoid, loose, hypocellular connective tissue proliferates around the tiny blood vessels, creating a pseudomicrocystic pattern. Collagen is deposited in the tumor along the blood vessels resulting in a trabecular patter. Gland formation is not seen.

The cells are small to medium-sized and polygonal to elongated, with clear to eosinophilic cytoplasm lacking glycogen and mucin. Nuclei are ovoid to round, grooved or indented with finely dispersed chromatin and inconspicuous nucleoli. Rarely, few cells have large or irregular nuclei. Mitotic figures are rare. Intracellular and extracellular eosinophilic, PAS positive , hyaline globules[12] are diagnostic .

The neoplasm can invade vessels in the surrounding capsule or extend into adjacent normal pancreas. Recurrences and metastases are uncommon limited to the liver and peritoneum (<10%).

They are usually benign, rarely malignant. Features associated with malignancy are perineural invasion,vascular invasion,deep invasion into surrounding soft tissues, nuclear atypia, elevated mitotic rate and metastasis.

IHC[6,9,10]- The neoplastic cells are positive for vimentin, NSE, CD10, progesterone receptor, nuclear catenin ,nuclear cyclinD1 and E-cadherin.

Eosinophilic cytoplasmic globules are immunoreactive with $_1$-antitrypsin and $_1$-chymotrypsin.Negative for chromogranin, epithelial membrane antigen, and cytokeratin.Due to the indolent nature of the tumor, long-term clinical follow-up is recommended. They have an excellent prognosis after surgical excision[3,4,5,13].

DIFFERENTIAL DIAGNOSIS [(6,7,8)]:

1.Acinar cell carcinoma

2.Mixed acinar-endocrine carcinoma

3.Pancreatoblastoma

4.Pancreatic Endocrine neoplasm

5.Adrenal cortical neoplasms

SUMMARY:

A clinical diagnosis of Solid-pseudopapillary neoplasmis suspected when young female presents with a pancreatic mass.Surgical excision is curative. Prognosis is good.

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