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A CASE REPORT ON SOLID PSEUDOPAPILLARY TUMOR

General Surgery

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KEYWORDS

INTRODUCTION :

Solid pseudopapillary tumours (FRANTZ TUMOUR or HAMOUDI TUMOUR) constitute 3% of all pancreatic tumours and 6-10% of pancreatic cystic neoplasms. It is more prevalent in females in third decade, in the tail of pancreas . These patients have good outcomes with appropriate treatment .we present a case of SPN in young female undergone laparoscopic distal pancreatectomy.

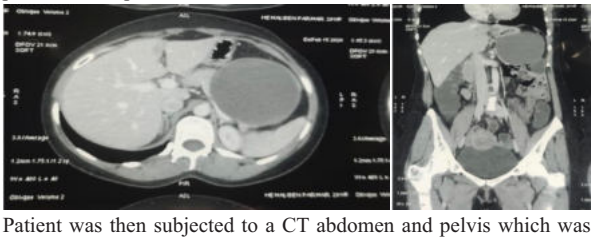
CASE REPORT:

A 29 year old female patient, a housewife, residing in morbi , gujarat came to civil hospital rajkot with the chief complaints complaints of Upper abdominal pain since last 1month .

No history of fever / vomiting / constipation / bleeding per rectum / burning micturition / history of anorexia / weight loss / yellowish discoloration of eyes / Clay colored stools.

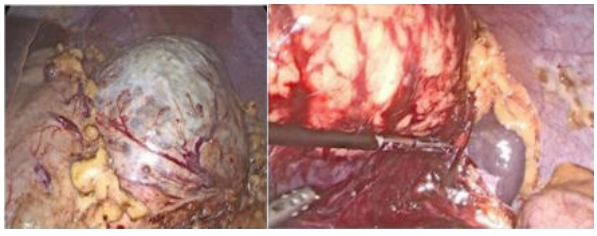
There was nothing significant in the past history, family history or the personal history of the patient .No previous operative intervention. Regular menstrual cycles and flow. Patient was vitally stable with no systemic abnormality.

Per abdomen examination was suggestive of a soft, non tender abdomen with no gaurding or rigidity. Chest and abdominal x - ray of the patient were normal. The ultrasonography was suggestive of a approximately 87x56 mm size of cystic lesion arising from the tail of the pancreas with evidence of mural thickening and thickened wall with few septas and multiple echoes within. Possibility of cystic pancreatic neoplasm.



Patient was then subjected to a CT abdomen and pelvis which was suggestive of a well defined thick walled cystic lesion of size 86 x 79 x 73 mm in the tail of pancreas with wall thickness of the lesion approximately 3.5 mm with enhancing septas with eccenteric enhancing soft tissue component within. Small foci of calcification is noted in the wall of the lesion. The lesion is displacing splenic artery and the veins posteriorly which otherwise appears normal, Possibility of cystic pancreatic neoplasm appears more likely than a pancreatic pseudocyst. A tiny calculus of size 3mm is noted in middle calyx of right kidney. All the other routine laboratory investigations were within the normal limits. Patient was undertaken a laparoscopic distal pancreatectomy.

Patient was kept in reverse trendelenburg position and the tranverse colon dissected off by dividing the lienorenal and the gastrocolic ligaments. Entering into the sac, a large cystic lesion and overlying vascularity visible along the tail of pancreas. Plane created under the inferior bundle of pancreas using harmonic scalpel while the pancreas kept in traction.



Pancreas everted and a plane created along the superior border of pancreas and splenic vein found adherent to pancreas. With meticulous dissection splenic vein dissected off the pancreatic tissue plane created posteriorly along the neck of pancreas and a umbilical tape passed behind it. After confirming the absence of any major vessels along the pancreatic tissue pancreas divided at neck using endo GI stapler. Cyst deflated and pancreatic tissue dissected off splenic hilum. Spleen examined for any devascularisation, found to be normal. Thorough wash given and a 24 Fr drain placed along the stapled line.

Immunohistochemistry came poitive for

- CD 10 Ò CD 56
- Beta – catenin
- Chromogranin Ò Vimentin.
- PR

Patient was discharged successfully on POD - 10. Post operative Histopathology was s/o LOW GRADE PSEUDO PAPILLARY TUMOUR with marked cystic changes .Immunohistochemistry was positive for CD -10 ,CD -56, Beta – catenin ,chromogranin and vimentin.

DISCUSSION :

Solid Pseudopapillary Tumors:

Solid pseudopapillary tumors (SPTs) of the pancreas are rare neoplasms with low malignant potential. Other names – Frantz tumors, Hamoudi tumors, and papillary cystic neoplasm. Represent up to 3% of all pancreatic tumors and 6% to 12% of pancreatic cystic neoplasms. High prevalence among women, most commonly - third decade of life and earlier can occur throughout the pancreas, slightly more common in the pancreatic tail and, when discovered, are generally large in size(mean diameter, 5.4 cm).Almost all SPTs harbor alterations in the APC/β-catenin pathway due to a mutation involving *CTNNB1 (exon 3).Nuclear accumulation of* β-catenin has been described in 95% of SPTs.74% of tumors overexpress cyclin D1, a downstream effector of β-catenin.

BCL9L, a β-catenin stabilizing gene, is significantly decreased in SPTs, which may help to attenuate the protumorigenic effects of overactivation of the Wnt/β-catenin pathway . Downregulation of miR-194 and SOX 9 has been reported in SPTs. In addition, genes involved in the hedgehog and androgen receptor signaling pathways, as well as genes involved in epithelial mesenchymal transition have been shown to be activated in SPTs.

Clinical Features:

The most common presenting symptoms are

1. abdominal pain (45%)
2. abdominal mass (34%).

3. asymptomatic patient, tumors may be discovered as a palpable mass on routine physical examination, or
4. as an incidental finding on imaging for an unrelated complaint.

Investigations

Ultrasound shows a hypoechoic or isoechoic lesion. These lesions can range from being completely cystic to completely solid.On CT imaging, SPTs are characteristically large, heterogeneously enhancing lesions with solid and cystic components, and they frequently demonstrate peripheral enhancement and central calcification.On magnetic resonance imaging (MRI), SPTs have a low signal / intensity on T1-weighted images and a high signal intensity on T2weighted images.FNA biopsy may be useful when routine imaging is inconclusive but because of the tumor's largely necrotic composition, FNA biopsy can frequently be nondiagnostic.The radiographic differential diagnosis of an SPT should include –

- mucinous neoplasms
- serous cystadenomas,
- intraductal papillary mucinous neoplasms,
- cystic degeneration of a typically solid neoplasm, such as a pancreatic neuroendocrine tumor.

However, age is important; in a young woman under the age of 30, SPT and pancreatic neuroendocrine tumor would be most likely. In a young woman under the age of 20, SPT would clearly be the most likely diagnosis.

PATHOLOGY

Presence of solid cellular hypervascular regions without gland formation, presence of branching papillary fronds with sheets and degenerative pseudopapillae. Cells have eosinophilic granules within the nuclei, which are typically grooved.

Immunophenotype stains ;

neuron-specific enolase,

- CD10,
- atypical nuclear staining for β-catenin, which is generally a cytoplasmic protein.
- reported to positively stain for α-methylacyl coenzyme A racemase (AMACR).
- Progesterone receptors, whereas estrogen receptor positivity is more variable.

Treatment:

Surgical resection is recommended for all patients with localized SPT. Although these tumors may be extremely large and can invade critical vasculature, most lesions are usually amenable to complete resection.

Pancreaticoduodenectomy or distal pancreatectomy can be performed with en bloc resection of involved adjacent organs when indicated. Although recurrence rates are low, long-term surveillance is important. If metastatic disease occurs, the most common sites include liver, mesentery, and peritoneum. Several series have reported long-term survival after metastasectomy. For unresectable metastatic disease, anecdotal case reports have suggested that gemcitabine based chemotherapy may be successful in some patients.

CONCLUSION :

Pancreatic SPNs are rare neoplasm with malignant potential found primarily in young women .formal surgical resection may be performed safely and is associated with long term survival .Because long term survival can be achieved ,patients with SPN should be treated aggressively with complete resection ,even if this requires metastasectomy.

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