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Solid pseudopapillary epithelial neoplasm of pancreas in pregnancy: Case Report of a Rare Co-Occurrence

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Abstract
Background: *Solid pseudopapillary epithelial neoplasm of pancreas (SPEN) also known as papillary epithelial neoplasm and cystic-solid papillary carcinoma of the pancreas is an uncommon neoplasm of exocrine pancreas. It predominantly occurs in young women in their twenties and thirties and accounts for 1-2% of all pancreatic tumors. It is a peculiar tumor for having low grade malignant potential and an excellent prognosis. Most patients are asymptomatic at diagnosis and may present with gradually enlarging abdominal mass or vague abdominal pain. The diagnosis is usually done by imaging (CECT, MRI) and histopathology. Here we report a case of 22 years old pregnant woman who was referred to our hospital with a history of gradually increasing abdominal lump and vague abdominal pain since 1 year. She was diagnosed with SPN by imaging and histopathology.*
Case Report: *A 22 years old female with full term gestation was referred to us in view of gradually increasing abdominal mass and history of vague abdominal pain since 1 year. At 12 weeks of gestation she was investigated at her native place and was diagnosed with a mass in left renal fossa. In our hospital she delivered male child at full term by normal vaginal delivery. In postnatal period CECT was done which showed a lobulated lesion with internal calcification in left anterior para-renal space. A USG guided trucut biopsy confirmed the diagnosis of pseudopapillary epithelial neoplasm of pancreas. Distal pancreaticosplenectomy with left radical nephrectomy and left hemicolectomy was done. At the time of discharge patient was advised regular follow up. We are reporting this case because SPN are uncommon tumors and their co-occurrence with pregnancy is even rarer.*
Conclusion: *SPENs are rare but curable neoplasm of exocrine pancreas affecting young females in their 2nd or 3rd decade. The diagnosis is sometimes difficult because of a wide differential diagnosis however once diagnosed it has an excellent prognosis because of its low malignant potential. Presence of gradually increasing, well defined, cystic pancreatic mass in young females should raise the suspicion of SPN.*
Keywords: *Pseudopapillary epithelial neoplasm of pancreas, Pregnancy, Imaging, Surgical resection.*

Introduction

Solid pseudopapillary epithelial neoplasms (SPENs) of pancreas are rare neoplasms that almost exclusively occur in young women [1]. In 1959 Frantz first described the morphological features of the SPENs [2]. They are extremely rare

and thought to account for 1-2 % of exocrine pancreatic tumors, more common in Caucasian population and are usually detected in women in their twenties and thirties[3]. These tumors are characterized by their low-grade malignant potential and excellent prognosis after complete resection. These tumors are also known as Hamoudi or Franz tumor, solid and papillary epithelial neoplasm, papillary-cystic neoplasm, and cystic-solid papillary carcinoma of the pancreas. Usually these neoplasms are asymptomatic and diagnosed incidentally but they may present as gradually enlarging abdominal mass or abdominal pain. Other presenting complaints may include dyspepsia, loss of appetite, nausea and vomiting. A large tumor may compress upon stomach causing early satiety or if is compressing over bile duct it may cause obstructive jaundice. Although it can assume a very large size it has a very low malignant potential and very rarely metastasize, the prognosis therefore is excellent after complete resection [4].

The diagnosis is usually done by imaging and histopathology can be used for confirmation. On ultrasound it may appear as large well-defined mass with heterogeneous appreance due to presence of solid as well as cystic composition. The most common location is tail followed by head of pancreas. On computed tomography SPENs appear as encapsulated lesions with varying cystic and solid components due to hemorrhagic degeneration. Post contrast imaging typically shows enhancing solid areas peripherally and non enhancing cystic spaces located in central part of the lesion. MRI may show a well defined encapsulated lesion which is hypointense on T1 hyperintense on T2 and may show slowly progressive post-contrast enhancement[5]. Though in majority of the cases diagnosis is reached on the basis of imaging a histopathological confirmation is essential before operative intervention. Pathologically these tumors are characterized by presence of encapsulated lesion with cystic and solid areas[6].

Though SPENs are slowly growing tumours malignant potential of these tumors is reported to be between 10%-15%. If metastasis occurs then liver, regional lymph nodes, omentum, and peritoneum are most commonly involved[7]. Local invasion may involve adjacent organs, including the duodenum, spleen, portal vein, superior mesenteric artery and bile duct. The treatment usually consists of complete surgical resection of tumor. Incomplete resection is fraught with the danger of recurrence and eventual dissemination. Open and laparoscopic surgeries are the available option. The choice of any one of these surgical methods depends upon hemodynamic stability of the patient, location of the lesion, tumor size and experience of the operating surgeons[8].

Co-Occurrence of these tumors in pregnancy is very rare and may present a diagnostic dilemma. It moreover may change the treatment protocol and timing of surgical management[9]. The ultimate goal of management of these tumors diagnosed during pregnancy is to minimize maternal as well as perinatal mortality. Any intervention must take into consideration the risks involved to mother as well as the developing fetus. There are no treatment guidelines for pregnant women with SPENs and decision about surgical interventions should take into consideration the risk and benefit to mother as well as fetus[10].

Case Report

A 22 years old G1P1 female with full term gestation was referred to our hospital in view of an antenatal scan showing retroperitoneal mass in left hypochondrium and lumbar region most probably arising from pancreas. She was married since 2 years and received treatment for infertility and was eventually diagnosed with this mass at 12 weeks of gestation and the treating obstetrician advised her MTP but she refused it since she had conceived after undergoing treatment for primary infertility and was desirous of continuing the pregnancy. In view of her desire to continue pregnancy the concerned obstetrician kept her

under close follow up and later referred her to us at full term gestation.

At the time of admission the patient was afebrile with haemodynamically stable condition. Blood pressure, Pulse and respiratory rate were normal. There was no pallor, icterus or edema. Respiratory, cardiovascular and CNS examination was normal. On per abdominal examination the height of uterus was corresponding to 36 weeks of gestation. Fetal heart rate was 138/minute. On palpation there was a palpable mass of approximately 10 X 10 cms. It was semisolid with restricted mobility. On per vaginal examination vertex was at -1 station and membranes were intact. Soon after admission the patient went into labor and delivered a male child weighing 2 kg. Baby cried immediately after birth and was shifted to mother. Breast feeding was started within 30 minutes of birth and baby started breast feeding well after 2 -3 assisted breast feeds.

After Delivery a computed tomography scan was done which showed presence of a large lobulated heterogeneous mass in left pararenal space with areas of necrosis and hemorrhage. A provisional diagnosis of a primary retroperitoneal sarcoma or pancreatic neoplasm was made on computed tomography and biopsy with histopathology was advised. Later ultrasound guided biopsy was done which showed tumor consisting of sheets of cells with pseudopapillary structures, hemorrhages with focal necrosis and occasional mitotic cells confirming the diagnosis of SPEN of pancreas.

In view of Imaging and histopathological diagnosis of SPEN surgery was advised. Patient refused surgery and took discharge against medical advice. She eventually turned up for surgery after 6 months. Pre-operative CT abdomen was done which showed 20 x 16 x 9 cm sized (Larger than in previous scan) lobulated lesion with internal calcification in anterior para-renal space on left side. The lesion appeared to be arising from the posterior aspect of the body and tail of the pancreas displacing it anteriorly and the adjacent structures away from it. Uterus and right ovary appeared normal.

During surgery gross examination showed a large (25 x 18 cm) solid cystic mass with multiple lobulations arising from body and tail of pancreas. It was seen involving the splenic artery and vein distal to the confluence and was found to be invading left kidney anteriorly and the left mesocolon with the left transverse and proximal descending colon were found to be draped over the tumor. Spleen was found to be draining via a tortuous and dilated gastroepiploic arcade. There was no nodal involvement or metastasis (Tumor Stage T4N0M0). In view of extension of lesion distal pancreaticosplenectomy with left radical nephrectomy and left hemicolectomy was done.Post-operative period was uneventful and the patient was discharged 1 week later. Follow up examination after 6 months revealed no abnormality and follow up ultrasound was found to be normal.

Discussion

In 1959 Franz first reported a pancreatic tumor of uncertain lineage which was predominantly seen in young Asian or Caucasian females .The definite origin of these neoplasms are not known and various hypotheses with regards to its origin have been postulated. Many authors favor the theory that SPNs originate from multipotent primordial cells. Many others believe these tumors to arise from extra-pancreatic origin from genital ridge angle-related cells[11]. Santini et al considered their origin to be from pluripotent embryonic cells of the pancreas with multipotential differentiation. However they didn't find a clear-cut evidence of terminal differentiation to either acinar or endocrine cells. Various other theories of their origin includes its origin from stem cells or from primitive ovarian cells within the pancreatic parenchyma. But the low malignant potential and slow growth goes against the theory of stem cell origin and occurrence of these tumors in males goes against the theory of its origin from primitive ovarian cells [12].

SPENs are most commonly seen in females in their twenties or thirties. In a large retrospective

review of more than 700 patients it was found that 90% of the affected patients were females with a mean age of 22 years [13]. The commonest site is head or tail of the pancreas. Very rarely it may occur in ectopic pancreas. Sudeep Khaniya et al reported an interesting case of SPEN occurring in ectopic pancreatic tissue. They reported the lesion to be arising from the root of small bowel mesentery medial to the duodenojejunal flexure. Lymph node metastasis was also present [14].

There are no specific signs and symptoms associated with these tumors and many a times they are diagnosed incidentally. Common clinical features include abdominal pain and gradually growing abdominal mass, abdominal pain and recurrent pancreatitis. If the tumor enlarge in size it may compress upon stomach causing vomiting, early satiety and abdominal pain while compression over bile duct may cause obstructive jaundice. The most common and consistent yet non-specific symptom associated with these tumors is abdominal pain which is found to be present in more than 80% of the patients [15].

The diagnosis of these tumors depends upon a classical history of gradually increasing abdominal lump which on ultrasound imaging may appear as well-defined mass with heterogeneous appearances. The heterogeneity is due to presence of solid as well as cystic areas within the tumor. On computed tomography SPENs appear as encapsulated lesions with cystic and solid components. Post contrast a characteristic peripheral enhancement is seen. High-Signal intensity T1 weighted images showing hypointense fibrous capsule is the characteristic MRI feature of these neoplasms[16]. Biopsy and immunohistochemistry may further confirm the diagnosis. On histopathology the tumor appears as encapsulated lesion with cystic and solid areas. On immunohistochemistry majority of these tumors exhibit progesterone receptors. The other markers which were commonly present in these tumors include vimentin, AAT and NSE which were each found in more than 90% of the tumors. Though it is thought to be originating from epithelial origin cytokeratin was demonstrated only in 70% of the cases [17].

Treatment usually consists of complete surgical resection of the tumor. Due to its low grade malignant potential more than 90% patients are completely cured after complete resection. In case of Pancreatic SPEN local resection with pancreatectomy or pancreaticoduodenectomy are frequently done. Surgery must ensure complete excision and the surgical procedure is usually decided depending upon the size and extent of tumor [18].

The co-occurrence of SPEN and pregnancy is quite unusual and require meticulous management planning. Progesterone positivity of these neoplasms may be the reason for such an occurrence. The SPEN occurring during pregnancy may be detected relatively late because in many cases abdominal distension caused by tumor is attributed to pregnancy by patients [19]. It may also complicate the management as the standard treatment involving complete excision of the tumor needs to be weighed against the possible harm to the fetus. Moreover there are no universally accepted treatment guidelines for pregnant women with SPENs. On one hand since it's a low malignant potential a small tumor may be left alone till the baby is delivered while on the other hand possibility of malignant transformation and tumor rupture should not be ignored in a rapidly growing tumor [20].

Conclusion

Solid pseudopapillary epithelial neoplasms (SPENs) are rare pancreatic tumors usually occurring in young females in their 2nd and 3rd decade. Since they have a very low malignant potential a complete excision cures the condition. Pregnancy complicated by SPEN is a very rare phenomenon and may require excision during pregnancy. Though there are no universally accepted guidelines for management of SPEN in pregnant women resection if planned should preferable done in 2nd trimester.

Conflict of interest: None

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