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Median pancreatectomy for Solid Pseudo-papillary tumor of Pancreas - A Rare Case Scenario.

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Abstract: Solid pseudo papillary tumor of pancreas is a rare benign tumor of pancreas especially found among young females, most commonly seen in Asian countries, among the 2nd and 3rd decade of life. It has low malignant potential and excellent chance of long term survival. Here we are presenting a case of Pseudo papillary tumor over the Body of pancreas which warranted median pancreatectomy, a rare case scenario.

Keyword: Solid pseudopapillary tumor of pancreas, Median pancreatectomy, benign tumors of pancreas

Introduction

Solid Psuedopapillary tumor of pancreas has been reported first by Lichtenstein in 1933^{2,4,5}, It has been well documented in literature that solid pseudo papillary tumor of pancreas is more prevalent among young females in their second and third decade of life, based on the review of Theodossios and colleagues over 718 patients , its documented that the mean age was 21.97 ^{2,4}. Most common site of tumor localization was found more over the head and tail of pancreas accounting about (69.9%) and body for about (14.8%) ^{3,4}. Also solid psuedopapillary tumor is a potentially low grade malignancy hence has metastasis to liver, portal vein and spleen of about 19% ^{4,6}. According to literature the tumor involving the body of pancreas is relatively rare and need for median pancreatectomy alone was reported to be only 1.27% ^{4,6}.

Case summary.

A 22 yrs old antenatal female who underwent a routine antenatal scan on the 3rd trimester was found to have a mass in the body of the pancreas and was referred to our hospitals. She was evaluated. She denied any symptoms of abdomen pain, vomiting, haemetemisis, malena, steatorrhoea and weight loss. Her routine investigations, Liver function test, serum amylase and lipase were all normal. Tumor markers CEA and Ca19-9 were within normal limits. MRI of abdomen was done which showed mass lesion of size 10x8 cms involving the body of pancreas. It showed multiple solid and cystic components.Her Endo-sonography showed extrinsic compression of lesion probably arising from the body, FNAC was inconclusive.



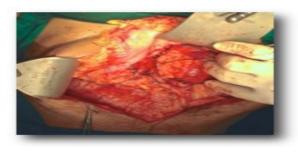
Endo-Sono Picture of tumor

Hence she was allowed to complete her pregnancy and planned for surgery later. She gave birth to a single live male baby without any complications who is healthy now. After 6 months post natal period she was referred to surgery and she was planned for distal pancreatectomy with preserving the spleen.



Pic 1- Tumor seen after opening the lesser sac

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Pic 2 - Tumor on the body of pancreas after resecting the neck Pancreatico jejunostomy on distal end with head primary closure



Intra-operatively a large 12x8 cms solid Tumor was noted found occupying the body of pancreas just from the head with lateral 25% of body and tail was relatively free of Tumor. Hence it was decided for median (central) pancreatectomy with primary closure of head and Distal end - pancreatico jejunostomy by Duncan's Technique. Two abdomen drains were left.Immediate post operative period was uneventful. On the fifth post operative period patient developed pleural effusion on tapping evidenced to have Pancreatic-Pleural Fistulae, was treated with ultrasound guided aspiration amproved well. Hence she was discharged on her 10th post operative day after removal of abdomen drains. Her biopsy revealed Solid — Pseudo Papillary Neoplasm of Pancreas with resection margins free of tumor.

Discussion:

Solid Pseudo papillary tumor of pancreas is a rare entity which has low metastatic potential as cited by Theodossios ^{4,5,6}, In 1981 Klippel and colleagues explained about it and WHO has classified it as exocrine pancreatic tumors and appropriate terminology goes by as Solid Pseudo papillary tumor 4,7,8.

They account for about 6% of all exocrine pancreatic neoplasm 2,3,4 . In children there has been a reported 8% of solid pseudo papillary tumor among pancreatic neoplasm 8,9.

The commonest age is between the 2nd and 3rd decade of life as in our case $^{3.4.7}$

Clinical presentation is usually varied, commonly presenting as abdominal mass and pain over the upper abdomen with radiation to the back yet it is usually picked up incidental as in our case ^{4,5}.

Pathogenesis is said to be probably due to chromosome abnormality in form of a karyotype unbalance translocation ¹⁰.

Imaging – the tumor is well demonstrated in ultrasound, but CT scores better in visualizing the tumor, respectability and surrounding vascular and structural invasions. Also it has better reliability on picking up the metastasis, yet since our patient was a antenatal mother MRI was done for her. By reference MRI also has same reliability for picking up the tumor ¹¹. Endo-Sonography guided FNAC by literature has better diagnostic value when it is feasible, here the FNAC findings were inconclusive and since the need of tissue diagnosis was not necessary when there is clear cut finding in imaging, CT guided FNAC was avoided ^{4,11}.

Surgical resection with free margins were the Treatment of choice in the literature 12,13,14 .

Yet the location of tumor over the Body for which median pancreatectomy was conducted was reported very rarely as in our patient, a total of 14.8% has been reported to have tumor in the body of pancreas and Median Pancreatectomy was conducted in only 1.27% of cases ⁴. Follow up with tumor markers has been reported useful especially in the follow up for the patients such as CA 19-9 are reported to be useful but was found to be normal in our patient ^{4,13}.

The mainstay of treatment according to literature still is with surgical resection, however certain reports have stated that for metastatic tumor of the same neoadjuvantchemotherapy has improved the resectability. Yet it still remains a debate. The Sloan-Kettering caner institute, USA has published that long term survival relies on the Complete surgical excision with removal of metastasis if any present 17,18.

Post Median Pancreatectomy complications:

The major complication after median pancreatectomy is possibility of leak from both ends - head and pancreatic jejunostomy ^{19,20}, Also the rate of Pancreatic fistula is also higher as in Review of Literature by Bassi et al the incidence is 10 - 20% respectively for head and distal resection where as 56% for median pancreatectomy ²⁰,

Conservative management such as retaining abdominal drains and if not present percutaneous drainage procedure has been reported as the mainstay of treatment and patients usually respond well as seen in our patient. As well as ICD insertion and pleural tapping is indicated if Pancreatic-Pleural fistulae develops ²¹.

As shown by Claudio Bassi and colleges 21 - there has been no reported mortality.

Advantages of this resection are good endocrine and exocrine long term function $^{19,20}. \\$

Sperti et al. showed no case of impaired endocrine function in 59 evaluated post operative patients.

Also possibility of spleen preservation also is very high thus preventing post splenectomy complications²¹.

Prognosis: In general, the prognosis is good, even in SPT with metastases or invasions. The doubling time of the tumor, as calculated by Kato and colleagues,81 is 765 days. More than 95% of patients with SPT limited to the pancreas are cured by complete surgical excision 1,4,6,7,12,17.

Local invasion, recurrence, or limited metastases are not contraindications for resection, and some patients with "unre-sectable" tumors can survive for more than 10 years after the operation 15,17,18. In patients with liver metastasis, longterm survival seems to have a better chance when excisional therapy 13,14,15,17 is used on the metastatic tumors; in the few unresectable cases in which radiotherapy 17 or chemotherapy 16,17 were used, results were encouraging. Prognosis for SPTs with treated liver metastases usually surpasses 5 years (ranging from 6 months91 to more than 17 years83). International literature 1-4,8,13,15,17 supports the concept that, despite the large size of these tumors and their ability to extend locally, complete surgical excision offers benefits in almost all patients. SPTs are of low-grade malignancy and are potentially curable by extended resections of the primary tumor mass. Operations must be with particular respect to radical oncology as conservative as possible because of the relatively low malignancy and the encapsulated form of the neoplasm.

Conclusion

As per the literature review, Pseudo papillary tumor of pancreas is a rare tumor of pancreas with a very rare presentation over the body and the need of median pancreatectomy is rarely reported. Hence the benefits of preserving the maximum amount of pancreas is emphasized especially in young individuals 1,4,7,15,16.

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