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Case Study

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A CASE REPORT OF SOLID PSEUDOPAPILLARY CARCINOMA OF PANCREAS

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ABSTRACT

Background: Solid pseudopapillary neoplasm (SPN) of the pancreas is generally regarded as a low-grade malignant tumour that predominantly develops in young women in the second and third decades of their lives.It is a rare type of tumour accounting for 0.9%-2.7% of all pancreatic tumours.[1] SPN is a pancreatic cancer in which there is no differentiation into a specific cell line, and it is considered an undefined tumour histo- genesis.[2] It has a low-grade malignant potential with excellent post-surgical curative rates and rare metastasis. Despite advances in imaging, pseudocysts and other cystic neoplasms feature in the differential diagnosis. On morphology alone, other primary pancreatic tumours and metastatic tumours pose a diagnostic challenge. Recent advances in immunohistochemical characterization have made the histopathologic diagnosis more specific and, in turn, shed light on the likely histogenesis of this rare tumour. Among the few patients who have died from metastatic SPN are mostly those whose tumours harbour an undifferentiated component characterized by diffuse sheets of cells with increased nuclear atypia and proliferative index. Such high-grade tumours have been subclassified as SPN with foci of high-grade malignant transformation. **Case summary:** A 54 year old lady presented with epigastric pain. She also complained of mild weakness and frequent episodes of indigestion and bloating. On physical examination, no significant abnormality was detected. Initial laboratory tests revealed a raised white blood cell count, 16 × 109/L, and a high C-reactive protein level of 3 mg/dL, consistent with an inflammatory reaction. Other biochemical values were normal.

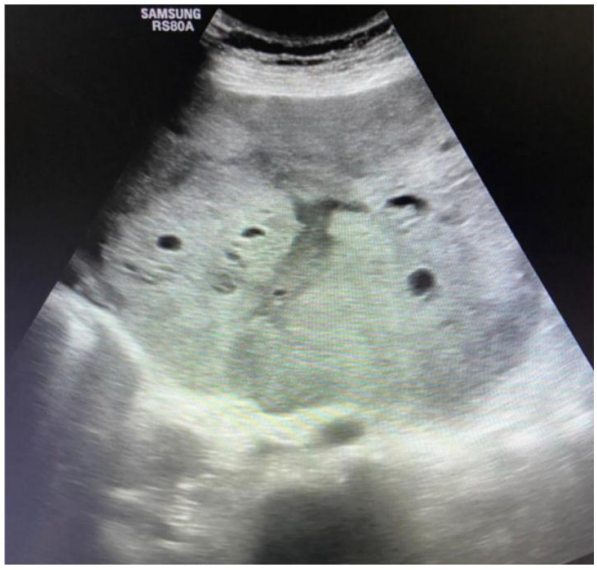


Figure 1: On USG, there was a large heterogeneously hyperechoic lesion in the epigastric region in closer relation to the pancreatic tail. There was another well defined heterogeneously hyperechoic round to oval lesion in the right lobe of liver. Both lesions exhibited significant internal vascularity on colour doppler study.

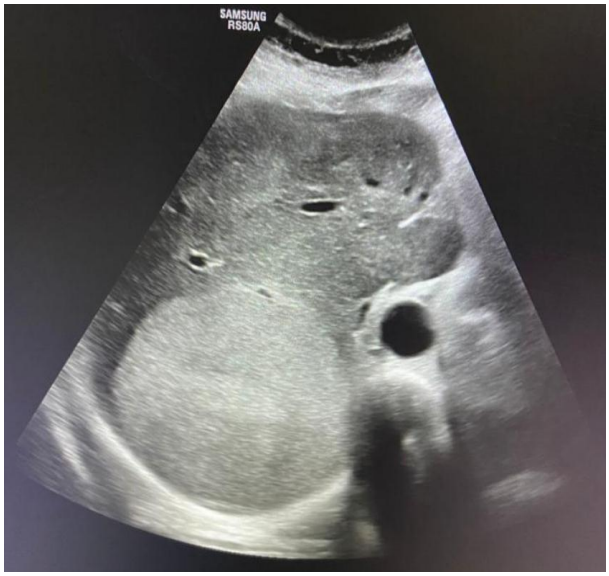


Figure 2: On contrast enhanced CT, there was a hypodense heterogeneously enhancing lesion in the abdominal cavity in close relation to the tail of pancreas. It was noted to have an exophytic component which is infiltrating into the left lobe of the liver. The lesion was also encasing and infiltrating into the left branch of the portal vein.

There was another similar lesion in the segments VI and VII of the liver.

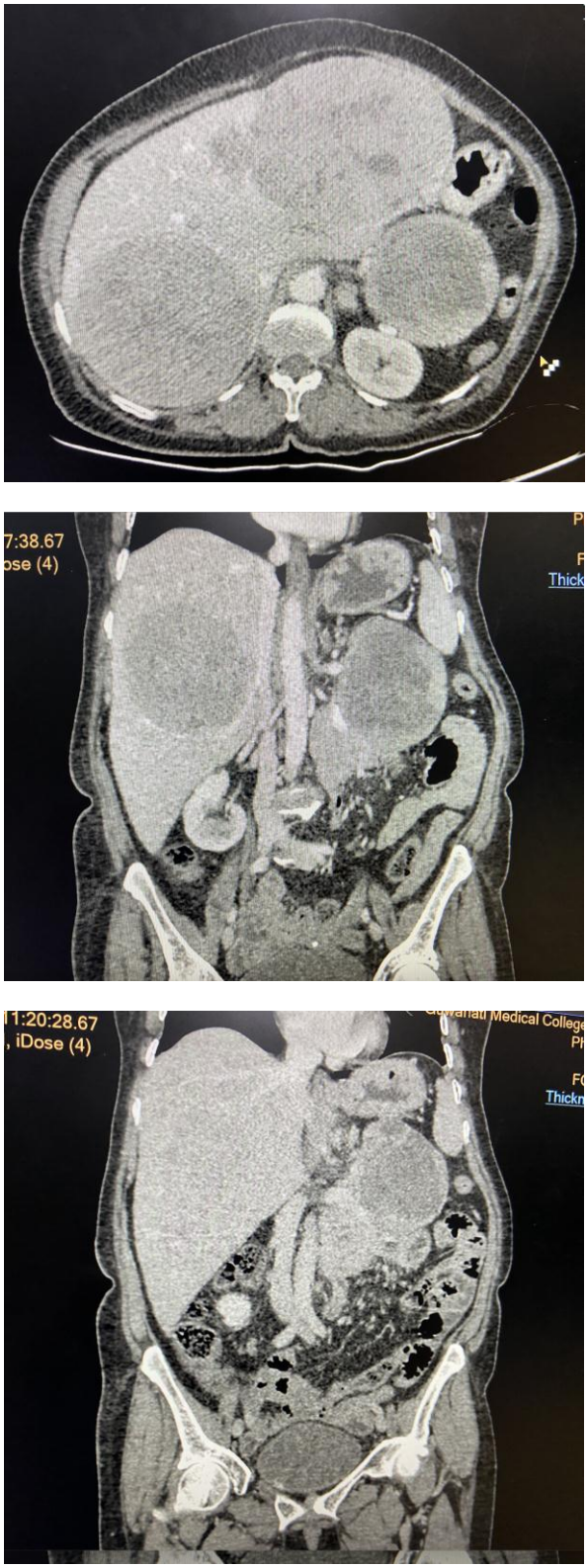
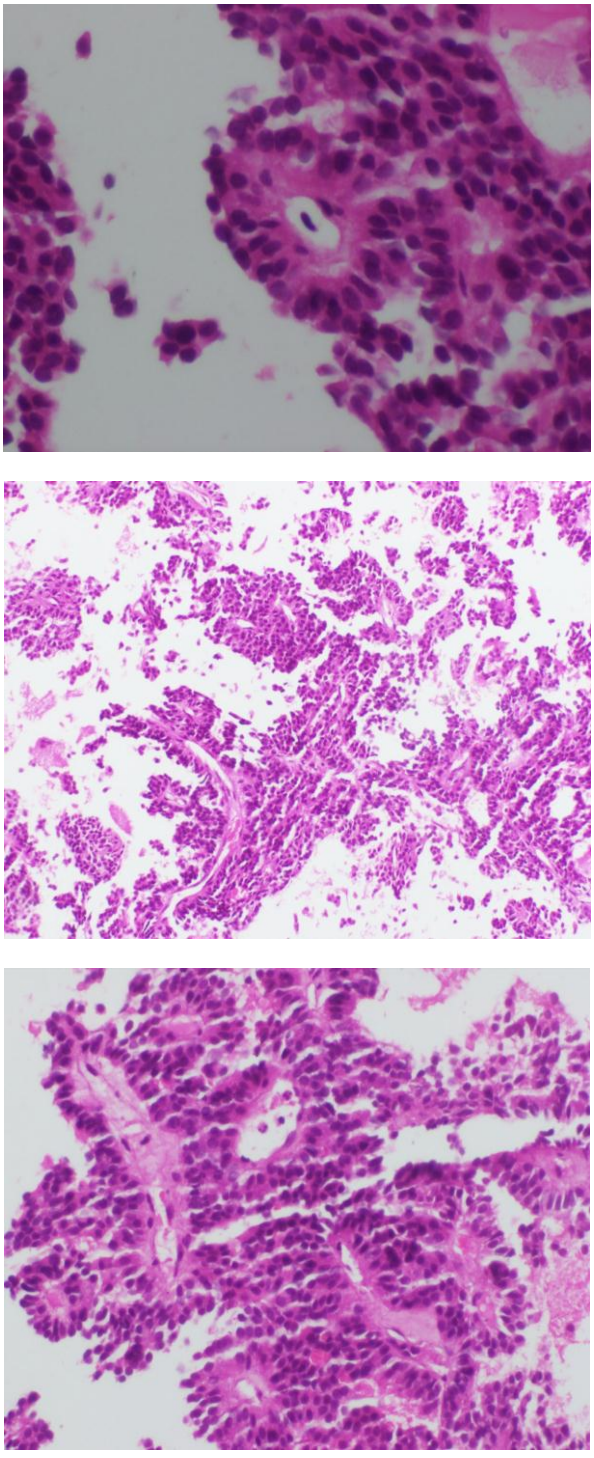


Figure 3: Subsequently, the patient underwent USG guided biopsy in the department of Radiology, GMCH and the collected samples were sent for HPE.

On HPE, the sections showed small fragments of a tumour composed of sheets of tumour cells at places surrounding the vascular core. The cells had moderate amount of cytoplasm with ill-defined border and round to oval nuclei with finely granular chromatin. The findings were concluded to be suggestive of solid pseudopapillary neoplasm of pancreas with differential diagnosis as NET.



DISCUSSION

SPNs are usually found incidentally hence it is very important that an accurate radiologic diagnosis is given for their appropriate and timely management. Symptoms may be vague such as abdominal pain or vomiting due to compression, namely in large tumors. A surgical

approach is usually indicated aiming complete resection, with tumor size not predicting resectability.

Majority of SPNs (conventional SPNs) are low-grade malignant tumours that show an excellent long-term prognosis for localized or even metastatic or recurrent disease after complete surgical resection.[3,4]

However, as in the present case, a few patients have died from metastatic SPN, mostly those whose tumours harbour an amorphous, undifferentiated component lacking typical pseudopapillary structures.[5,6,7] Such fatal tumours have been subclassified as SPN with foci of high-grade malignant transformation. The tumour identified in the present case seems to be consistent with this high grade type. Such tumours are found in the elderly age group as opposed to the more common low grade tumours which predominate among the younger age groups.

The differentials for these tumours include solid pancreatic tumours with cystic degeneration which are pancreatic adenocarcinoma, cystic islet cell tumour (insulinoma, glucagonoma, gastrinoma), metastasis, cystic teratoma and sarcoma.

CONCLUSION

This case report on pseudopapillary neoplasm of the pancreas highlights the enigmatic histogenesis of this indolent neoplasm and the need to distinguish it from more aggressive pancreatic tumors that may be considered in the morphologic differential diagnosis. There is a need for better understanding of these tumours and their inclusion in the common differentials for solid cystic tumours of this region as their timely diagnosis and management can go a long way in reducing the morbidity and mortality associated with the disease.

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