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Frantz-Gruber Tumour

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ABSTRACT

Cystic tumours of pancreas often present a pre-operative diagnostic dilemma. Frantz-Gruber is one such tumour which carries an excellent long term prognosis if adequate surgery is done. However, a pre-operative diagnosis is rarely made and often, patients are labelled and treated as pancreatic pseudocyst, which delays the surgical intervention or results in inappropriate surgery. We present a case of young female with cystic tumour of body of pancreas being treated as pancreatic pseudocyst elsewhere.

KEYWORDS : Cystic tumour, pancreas, Frantz-Gruber tumour, Solid pseudopapillary tumour

Introduction:

The pancreas is a retroperitoneal organ known to be affected by a variety of malignant as well as benign conditions. Solid pseudopapillary tumour (SPT) of the pancreas is a rare tumour, first described by Frantz in 1959 (1) and constitutes of 0.2 to 2.7% of the primary non-endocrine tumours of the pancreas (2). It is usually observed in young women. Due to its rareness and unusual behaviour, SPN of the pancreas is often associated with diagnostic and therapeutic challenges. We report the case of a 15 year-old female having a solid pseudopapillary tumour of the body of the pancreas.

Case History:

A 15 year-old female patient presented in the Surgical Outpatient Department with history of abdominal pain of 8 months duration. The pain was intermittent in nature and not associated with anorexia, nausea, vomiting, weight loss, or any other systemic complaints. The CT scan done elsewhere, 6 months ago had revealed a predominantly cystic mass in the body of pancreas.

The patient was told to have a pseudocyst pancreas and was accordingly put on conservative treatment and close follow-up. Patient continued to be symptomatic and noticed progressive increase in the size of tumour, which was corraborated on repeat ultrasonography of abdomen. She presented to our institute at this point. On examination, she was a well preserved and abdominal examination revealed a mass in epigastric region and left hypochondrium which was immobile and measured about 6 x 5 cms in size. There was no left supraclavicular lymphadenopathy. Although a repeat CT imaging was planned for her but due to financial reasons, the same could not be obtained. Blood investigations were within normal limits. Ultrasound of abdomen did not reveal any liver metastasis or intra-abdominal lymphadenopathy. She was planned for distal pancreatectomy.

Intra-operative findings : there was 8 x 6 cms cystic lesion in body of pancreas. (Fig 1). There was no evidence of liver or peritoneal metastasis. There were no enlarged regional lymphnodes. Distal pancreatectomy with splenectomy was done Post-operative period was uneventful.

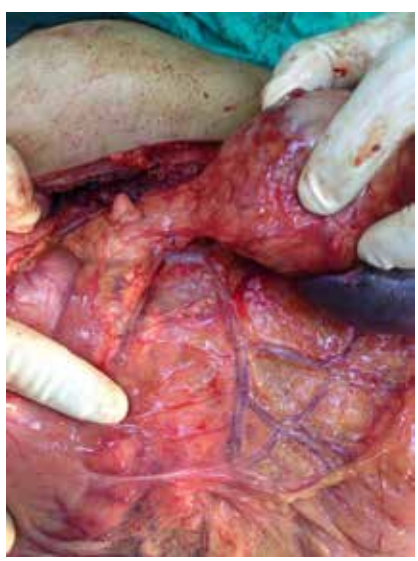

Fig 1. Operative Photograph showing tumour arising from body of pancreas.

Histopathology (Fig. 2) of the specimen was Solid pseudopapillary tumour of the pancreas.

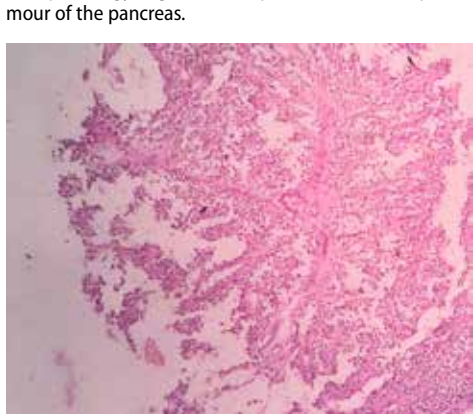

Fig 2. Section shows uniform round cells arranged in pseudo-papillary pattern.(H&E X200)

Discussion:

In 1959 Frantz described an unusual tumour arising from the pancreas termed papillary tumour of the pancreas [1]. It is known by many names solid and pseudopapillary neoplasm, solid and cystic tumour, papillary cystic tumour, solid papillary epithelial neoplasms, Frantz tumor, Frantz-Gruber tumour. It was named by the World Health Organization (WHO) as solid pseudopapillary tumors of the pancreas in 1996 [2].

SPT is tumour of low grade malignant potential with invasion of capsule and adjacent structures. SPTs are observed in young women (female: male ratio, 20:1, mean age is 28 years; with over 90% of patients reported in women under the age of 35 [3]. Hormonal influences appear to play a role in the development and the evolution of SPNs [4], but these neoplasms can arise in men, although men comprise <10% of all patients.

The most frequent symptom of these tumours is upper abdominal pain seen in nearly half the patients; this was the only presenting symptom in our case. Symptoms may be present for years before correct diagnosis is made. The most important distinction to be made is the differentiation of cystic neoplasms from other cystic lesions of the pancreas, particularly pancreatic pseudocyst. The patient in our case was initially misdiagnosed as pancreatic pseudocyst and was kept under observation. It was only after 6 months of follow-up that when the patient could feel the tumour increasing in size she was referred to our centre and offered surgery. Less than one third of cystic neoplasms are correctly diagnosed preoperatively and some patients even undergo inappropriate surgery after being wrongly labelled as pancreatic pseudocyst [5].

Difficulty in preoperative diagnosis is also because of lack of specific tumour markers known.

These tumours can be located anywhere in the pancreas with 30-40% tumours located in the head and 40-50% in the tail region [6]. Metastases are found in about 15% of the cases and can involve lymph nodes, liver, spleen, colon and mesocolon [7].

Differential diagnosis includes pseudocyst, mucinous cystic tumors, mucus secreting tumours, microcystic adenoma, islet cell tumor, acinar cell carcinoma, cystadenocarcinoma and pancreaticoblastoma [7].

These tumours carry a good prognosis with 5 year survival rate reaching upto 97% with adequate surgery. Patients presenting with atypical histology, elderly patients, men and patients with incomplete resection are at higher risk of recurrence and death. [8]

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