Gruber-Frantz Tumour

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Introduction

Gruber-Frantz tumour is a rare cystic neoplasm of the pancreas that is most frequently seen in young females. Cystic pancreatic neoplasms are commonly misdiagnosed as pancreatic pseudocysts that are, by far, the most common cystic lesions of the pancreas. The presence of cystic pancreatic mass in the absence of a history of pancreatitis must be regarded with great suspicion and investigated thoroughly. These tumours are important because even benign tumours have a malignant potential and correct treatment by total surgical resection is curative. Prognosis is very good unlike in ductal adenocarcinomas of the pancreas.

Case Report

A 16 year old girl presented with a history of slowly growing swelling in the upper abdomen of one year's duration. It was accompanied by occasional, post-prandial upper abdominal pain. There was an episode of acute pain lasting for 4 days, 8 months earlier. There was no history of vomiting or weight loss and appetite was good. There was a 10 cm x 12 cm, globular lump in the epigastrium. It was firm, nontender and immobile. Ultra sound revealed cystic mass with internal septa with solid material within -? Pancreatic pseudocyst.

CT scan revealed a well defined oval, cystic lesion with irregularly enhancing wall antero-lateral to the body and tail of pancreas -? Pancreatic pseudocyst. Laparotomy revealed a lesion suggestive of Pancreatic pseudocyst and a Roux-en-Y cysto-jejunostomy was done after biopsy of the cyst wall. She had an uneventful post-operative recovery. Histopathology of the cyst wall revealed solid cystic papillary tumour of the pancreas.

Patient on follow-up 2 months after the operation was found to have a recurrent lump in the epigastrium accompanied by moderately severe abdominal pain, anorexia and loss of weight. She had occasional episodes of vomiting. The lump in the epigastrium was 12 cm x 12 cm in size, firm, nontender and immobile. CT scan (Fig-1) showed a thick walled lesion, 8.8 x 8.2 x 5.0 cm, in the body and tail of the pancreas. Air and a small fluid collection were noted (previous cysto-jejunostomy). It was reported as a possible neoplastic lesion of pancreas. Laparotomy revealed a mass lesion in the body

and tail of the pancreas about 14 cm x 12 cm x 10 cm encroaching on to splenic hilum. Liver and peritoneum were free. The lesser sac was accessed through the gastro-colic omentum. The body and tail of the pancreas along with spleen were resected and the tumour could be removed with satisfactory surgical margins. The Roux-en-Y cysto-jejunostomy was undone and enteral continuity restored. Patient had an uneventful post-operative recovery except for a short-lived pancreatic fistula. Three months post-operatively patient is asymptomatic, has gained 3 kg weight and there is no recurrence of the tumour clinically and on ultra sound. Histopathology of the resected specimen showed a tumour arising from pancreatic cells comprising variegated areas of solid pattern, pseudopapillary pattern, clefted slits and pseudomicrocysts having myxoid areas. Cells were polygonal with ovoid nuclei. No mitosis was seen. It was reported as a solid cystic pancreatic neoplasm (Gruber-Frantz tumour) (Fig 2 and Fig 3).

Discussion

Cystic pancreatic neoplasms are very uncommon entities. Robson and Moynihan classified them separately in a textbook in 1903 and Beust in 1915 [1]. Friedenwald and Cullen reported malignant progression

Fig. 1 : CT scan of the abdomen showing solid cystic tumour of the body of pancreas

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Fig. 2 : Scanner view of the tumour showing pseudopapillae in cystic space

of three cystic neoplasms. In 1978 Compagno and Oertel found two clinically and histologically distinct types in the files of Armed Forces Institute of Pathology; microcystic adenoma which is uniformly benign and mucinous cystadenoma-cystadenocarcinoma which has considerable malignant potential [2]. This classification has enabled more accurate characterization of the disease. In 1959 Frantz described a third type termed papillary tumour of the pancreas [3]. Our case belongs to the category of cystic papillary tumour of the pancreas or Gruber-Frantz tumour of which only 150 cases have been reported in the literature. It has a low grade malignant potential manifested by invasion of the capsule and contiguous structures. The cell of origin of these rare tumours is unknown; investigators have suggested an origin from primordial pancreatic cells and it has been classified under neoplasms of indeterminate type [4]. It begins as a solid mass in which there are many poorly supported tiny vessels. Cells farthest from the vessels degenerate whereas the cells near the vessels remain intact resulting in a pseudopapillary pattern and cystic spaces. Groups of foamy macrophages accumulate along with giant cells [4,5].

Gruber-Frantz tumours characteristically occur in young females as it was in our case [6]. 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. This distinction is often very difficult without an operative biopsy. In one study, less than one third of cystic neoplasms were correctly diagnosed pre-operatively; some patients underwent inappropriate operation to treat what was believed to be a simple

Fig. 3 : Micrograph of pseudopapillae comprising cells with grooved/indent nuclei

pancreatic pseudocyst [7]. In our case also a correct diagnosis could be made only after operative biopsy. Often the correct diagnosis is made only at laparotomy with incisional biopsy. Without biopsy as many as 80% of cystic neoplasms may be misdiagnosed at laparotomy [8]. This strongly supports the use of routine incisional biopsy of every pancreatic pseudocyst.

There is little controversy about the optimal treatment of neoplastic pancreatic cysts. They all possess malignant potential and thus must be treated aggressively [2,9]. Unlike pancreatic ductal adenocarcinoma, resection often results in cure. Any lesser procedure results in invasion into porta hepatis, liver, major vascular structures and viscera. All these tumours should be viewed as malignant tumours and treated with anatomic pancreatic resection [1]. This was possible in our case. The diagnosis of a "benign" cystic neoplasm can only be made confidently after total extirpation and examination of the entire lesion because a small focus of malignant epithelium may be found in an otherwise benign appearing tumour [2]. Resection of Gruber-Frantz tumours affords an excellent prognosis to the young women in whom these lesions develop, with 10 year survival approaching 100% [6,10].

References

1. Turange RH, Zweng TN, Strodel WE. Cystic Pancreatic Neoplasms. In : Zuidema GD, Turcotte JG, editors. Shackelford's Surgery of the Alimentary tract Vol III. 4th ed. Philadelphia : WB Saunders, 1996;134-46.
2. Compagno J, Oertel JE. Mucinous cystic neoplasms of the pancreas with overt and latent malignancy (Cystadenocarcinoma and cystadenoma) : A clinicopathological study of 41 cases. *Am J Clin Pathol* 1978;69:573-8.
3. Frantz VK. Tumours of the Pancreas. In: Atlas of Tumour Pathology, First Series, Fasc. 27 and 28, 1959. Washington DC, Armed Forces Institute of Pathology.
4. Oertel JE, Oertel YC, Heffes CF. Pancreas. In : Sternberg SS,

- editor. Diagnostic Surgical Pathology. 3rd ed. Philadelphia : Lippincott, Williams and Wilkins, 1999;1469-1509.
5. Kloppe IG, Maurer R, Hoffman E et al. Solid-cystic (Papillary cystic) tumours within and outside pancreas : Report of two patients. Virchow's Arch (A) 1991;418:179-83.
 6. Kaufman SI, Reddick RL, Steigel M et al. Papillary cystic neoplasms of pancreas : A curable pancreatic tumour. World J Surg 1986;10:851-6.
 7. Tuma RH, Eckhauser FE, Ninik AL et al. Diagnostic dilemmas in patients with cystic neoplasms of the pancreas. Int J Pancreatol 1988;3:477-82.
 8. Strodel WE, Eckhauser FE. Cystic neoplasms of the pancreas. In : Dent TL, editor. Pancreatic disease : Diagnosis and Therapy. 1st ed. New York : Grune and Stratton, 1981;455-67.
 9. ReMine SG, Frey D, Rossi RL et al. Cystic Neoplasms of the pancreas. Arch Surg 1987;122:443-8.
 10. Kloppel G, Morohoshi T, John HD. Solid and cystic acinar cell tumour of the pancreas : A tumour in young women with favourable prognosis. Virchow's Arch (Path Anat) 1981;392:171-82.

Events Calendar

5th July 03

Interactive Symposium On Organ Donation

Venue : INHS ASVINI

Mumbai

Contact Person : Surg Cdr MSN Murthy

INHS Asvini, Colaba

Mumbai - 400 005

Tel : 22151666/3227

6th July 03

Update on Renal Transplantation

Venue : INHS Asvini

Mumbai

Contact Person : Surg Cdr MSN Murthy

INHS Asvini, Colaba

Mumbai - 400 005

Tel : 22151666/3227

7th July 03

Work Shop On Laparoscopic Donor Nephrectomy and Live Related Renal Transplantation

Venue : INHS Asvini

Mumbai

Contact Person : Surg Cdr KSK Patrule

INHS Asvini, Colaba,

Mumbai-400 005

Tel : 22151666/3094

5th -7th September 03

MAMICON - 2003

9th annual Conference of the Maharashtra Chapter of the Indian Association of Medical Microbiologists & Preconference CME

Venue : Armed Forces Medical College,

Pune 411 040.

8th -9th September 03

MICROCON - 2003

27th Annual National Congress of Indian Association of Medical Microbiologists to be held at Mumbai.

15th,16th September 03

National Medicine Update

Venue :INHS Asvini

Mumbai

Contact Person : Surg Capt YD Singh

INHS Asvini, Colaba

Mumbai - 400 005

Tel : 22151666/3215

22nd - 23rd Nov 2003

International Conference of Paediatric Anaesthesia Forum of ISA in collaboration with Asian Society of Paediatric Anaesthesiologists. AIIMS, New Delhi

Last date of registration : 15 Aug 2003

Contact : Dr DK Pawar, Addl Professor, Dept of Anaesthesiology, AIIMS, New Delhi.

Email : dkpawar@hotmail.com

27th November - 30th November 2003

33rd Annual Conference of Indian Academy of Cytologists & Pre Conference Workshop and CME

Venue : Armed Forces Medical College

Pune - 411 040

Chairman : Col SK Nema

Associate Professor and Head

Department of Pathology

Armed Forces Medical College

Pune - 411 040

Organising Secretary : Col S Satyanarayana

Associate Professor

Department of Pathology

Armed Forces Medical College

Pune - 411 040

Workshop : Adjunct techniques in cytology

CME : Genito-urinary system

Prof Bernard Naylor, Ann Arbor - USA & Prof S Vante, R Orell, Australia are delivering guest lectures.

For details contact organising Secretary /

Email : Cytocon2003@rediffmail.com

26th December - 30th December 2003

51st Annual conference of Indian Society of Anaesthesiologists at Bhubaneswar

Last date for regular registration : 30 Sep 2003

Contact : Dr N Acharya

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