

CASE REPORTS

Duodenal Fibrosarcoma Mimicking Franz Tumour Complicated by Post-resection Chylous Ascites

A Case Report

JM Plummer¹, KO Bonadie¹, N Williams², PA Leake¹, DIG Mitchell¹

ABSTRACT

This case report presents a young woman who underwent a Whipples resection for a large pan-creato-duodenal tumour. Pathology and immunohistochemical analysis of the tumour suggest duodenal fibrosarcoma. The patient's postoperative management was complicated by chylous ascites. A brief literature review is given to highlight this unusual case.

Keywords: Duodenal fibrosarcoma, chylous ascites, Whipples resection

Fibrosarcoma Duodenal con Apariencia de Tumor de Franz Complicado por Ascitis Quilosa Post-resección

Reporte de un Caso

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RESUMEN

Este reporte presenta el caso de una mujer joven a la cual se le practicó una resección de Whipples a causa de un tumor pancreato-duodenal grande. La patología y el análisis inmunohistoquímico del tumor sugieren la existencia de un fibrosarcoma duodenal. El tratamiento postoperatorio del paciente estuvo complicado por una ascitis quilosa. Se ofrece una breve revisión de la literatura para resaltar este caso inusual.

Palabras claves: Fibrosarcoma duodenal, ascitis quilosa, resección de Whipples

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INTRODUCTION

Duodenal fibrosarcoma is a rare tumour of varying malignant potential. Depending on its exact location, affected patients present with the usual features of a pancreato-duodenal tumour namely obstructive jaundice or features of gastric outlet obstruction (1). Pancreatoduodenectomy is the treatment of choice. We present a case of duodenal fibrosarcoma,

presumptively thought to be a Franz tumour, whose management was complicated by chylous ascites after a Whipple's resection. We believe the unusual combination of the patient's pathology and postoperative complication is worthy of sharing with the surgical community.

CASE REPORT

A 21-year old woman known to have the sickle-cell trait presented to the University Hospital of the West Indies (UHWI) with a one-year history of a rapidly enlarging upper abdominal mass associated with anorexia, weight loss and a history of recent onset jaundice.

Examination findings revealed an icteric woman with pale mucus membranes. Significantly, the patient had a 12 x

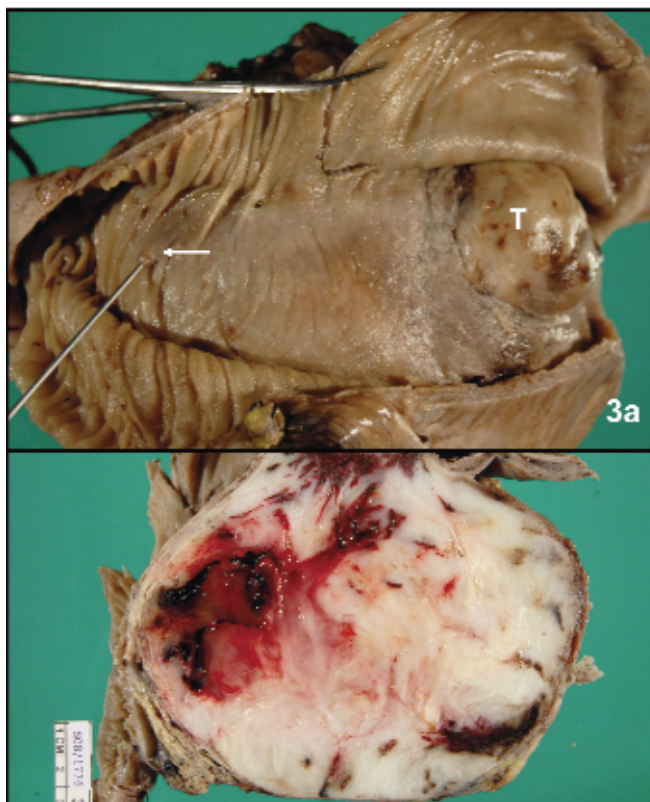
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vealed a 12 x 11 x 10 cm well-circumscribed, heterogeneous, abdominal, midline mass that was intimate to the pancreatic head, stomach and duodenum (Fig. 1). Additionally, upper gastrointestinal endoscopy revealed no mucosal abnormality in the stomach or duodenum.

Postoperatively, large volumes (between 1800 ml – 3000 ml) of fluid were noted from the lesser sac drain. Fluid analysis was not in keeping with a pancreatic fistula as the amylase content was 56 IU/L. On commencing oral diet, the fluid became milk-like in appearance and its volume increased (Fig. 2). The diagnosis of chylous ascites was confirmed by the appearance of the fluid plus chemical analysis of the fluid which revealed triglycerides 15.5 mmol/L, cholesterol 1.8 mmol/L, total protein 63 g/L with albumin of 39 g/L. Culture of the aspirate revealed no growth. The patient was started on octreotide with no significant response. Total parenteral nutrition (TPN) was commenced. Over the next four weeks, with reduction of her drain volume, TPN was weaned, lesser sac drain was gradually shortened and removed. Oral intake of low fat and short chain fatty acids was reinstituted.

spindle cells with only 1 mitosis/50 hpf. Occasional lymphocytes and plasma cells were scattered throughout the tumour and its borders were well-circumscribed (Figs. 3 and 4).



Immunohistochemical (IHC) stains on external consultation for vimentin and CD 10 were positive while significantly CD 117, PDGFRA, CD 34, desmin, caldesmon, SMMS, CD 30, pan-cytokeratin, CD 68, CD 163, S 100, tyrosinase, calretinin

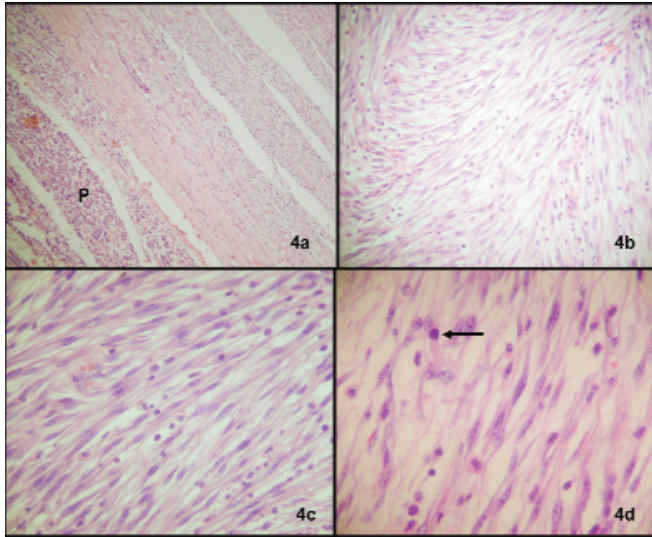


Fig. 4: Histological sections showing tumour composed of spindle-shaped cells with overlying pancreatic tissues (P) (4a); disposed as sweeping fascicles (4b). The component cells are uniformly bland (4c) with rare mitotic figures (T) (4d). H/E

and CD 99 were all negative. DNA sequencing for mutations in c-kit and PDGFRA were negative.

The lack of CD 117 and CD 34 immunoreactivity and failure to find a mutation in either the c-kit or PDGFR genes militate against a diagnosis of GIST while the lack of an inflammatory component and appropriate IHC ruled against inflammatory myofibroblastic tumour. The tumour was thus reported as a low grade fibrosarcoma. Presently, the patient is doing well 12-months after resection and has no evidence of recurrence.

DISCUSSION

The most common sarcomas affecting the small intestine are gastrointestinal stromal tumours (GIST) but various other sarcomas have been diagnosed based on electron microscopy and immunohistochemical (IHC) characterization of the differentiation of the spindle-shaped cells. It is difficult to accurately determine the number of duodenal fibrosarcoma reported in the literature but it is a rarity (2) with less than 20 cases identified in the English literature via a PubMed search for this review. The index patient was distinguished from GIST by the lack of CD117 and CD34 immunoreactivity and the failure to find a mutation in either the c-kit (CD117) or platelet derived growth factor receptor (PDGFR). The lack of an appropriate inflammatory component and appropriate IHC suggest that this was not an inflammatory myofibroblastic tumour. The presentation was similar to previously published duodenal fibrosarcomas (3) and is typical for a large mass in that location. The initial diagnosis as a Frantz tumour is acceptable as the local experience is usually in favour of this diagnosis in a young woman with a large

pancreatoduodenal mass (4). A Frantz tumour is an unusual tumour of the pancreas with varying malignant potential, which is usually well-circumscribed with regions of necrosis, haemorrhage and cystic degeneration. Histologically, solid and pseudopapillary components are evident as is a well formed fibrous capsule. Its cell of origin remains uncertain (4). Resection for cure for malignant tumours in this location is almost always with a Whipple's resection. Furthermore, given the absence of tumour necrosis and only 1 mitosis/50 hpf, we expect the index case to have a good prognosis as complete tumour resection was achieved.

The development of chylous ascites after pancreaticoduodenectomy though uncommon is well recognized (5, 6). It has an overall incidence of up to 11% post pancreaticoduodenectomy when studied prospectively, with as many as 4% requiring medical treatment (7). Because the cysterna *chili* are located at the same level as the pancreatic head, anterior to the first and second vertebrae, damage is likely to occur during a pancreaticoduodenectomy. This is more likely to occur if an extended lymphadenectomy is done (7). We believe that the index case suffered this complication because of the large size of the pancreatic head mass though a standard pancreaticoduodenectomy was done. Chylous ascites may adversely affect patient outcome by causing nutritional depletion, prolong hospital stay and increase risk of other complications. If the tumour is malignant, this may even lead to a delay in any adjuvant therapy.

This patient had a lesser sac drain producing amylase-poor serous fluid, and once oral diet was commenced the appearance of a milky white discharge from the surgical drain is typical and a well-described sign in patients with this diagnosis (8). Biochemical analysis of the fluid, with its high triglyceride content is confirmatory (9).

Several treatment options are available for the treatment of chylous ascites including dietary manipulations with short chain fatty acids, total parenteral nutrition (TPN) and surgery with the placement of a peritoneo-venous shunt or with direct ligation of the lymphatics. We choose management with TPN as it is our experience (10) and that of others (11) that administration of TPN over a variable period with the patient kept *nil per os* will allow for healing of the chylo-peritoneal fistula and clinical improvement with the majority of post-Whipples chylous ascites having successful conservative treatment. Total parenteral nutrition may be complemented with the addition of somatostatin which has been shown to be a safe and effective adjunct to non-operative treatment (12).

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