International Journal of Surgery Case Reports 86 (2021) 106327

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

# Invasive giant pancreatic desmoid-type fibromatosis with curative resection: A case report

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ARTICLE INFO

*Keywords:*
Desmoid type fibromatosis
Desmoid tumour
Fibromatosis
Pancreatic desmoid tumour
Radical pancreas resection
S100 protein
β-Catenin

ABSTRACT

*Introduction:* Desmoid-type Fibromatoses (DTF) tumours are rare, benign fibrous tumours with aggressive invasive behaviour that account for approximately 0.03% of all neoplasms. We report the success in curing a rare, invasive, and huge pancreatic intraabdominal DTF.

*Presentation of case:* A 42 years old male was medically free apart from recurrent left upper abdominal pain, anorexia, and nausea for more than ten years and no significant past surgeries, trauma, or family history of cancer. The patient has a non-tender large abdominal mass at the left hypochondria area extending down to the pelvis below the umbilicus with a rigid and smooth surface. The computed tomography scan showed a huge heterogeneous mass appears to be of pancreatic origin, measuring about 23 cm by 15 cm by 11 cm. The patient underwent radical antegrade modular pancreato-splenectomy, segmental transverse colectomy, adrenalectomy, and subsequent colo-colic anastomosis. The accurate gross size of the tumour specimen was 26 × 17 × 9 cm, and the weight was found to be 3.6 kg. Immunohistochemistry confirmed the diagnosis of pancreas DTF. The follow up to 5 years confirmed no recurrence reported clinically or by imaging.

*Discussion:* The Pancreas origin of DTF is a rarely reported subset with an incidence of around 5% of all DTF. Establishing the diagnosis is fundamentally based on the characteristic pathological and immunohistochemical studies, for the only available cure modality by complete radical resection to be promptly offered.

*Conclusion:* Our case is rare and uniquely the largest pancreatic DTF reported in the literature with curative resection despite being locally invasive.

## 1. Introduction

A desmoid tumour (DT), also known as desmoid-type fibromatosis (DTF) or aggressive fibromatosis, is considered a benign neoplasm in nature but locally invasive, giving it the aggressive behaviour, fatal extent, and an advanced stage status but without the potential propensity for distant metastasis. The origin of DT is through proliferation from well-differentiated mesenchymal fibroblasts.

Types of DTF either an abdominal wall or Extra-abdominal fibromatosis: mainly in the chest wall muscle, shoulder, thigh and back or Intra-abdominal fibromatosis from the mesentery or locating in the retroperitoneal region and pelvic region or explicitly associated with Gardner's syndrome. The consistency of DT mass varies between cystic, solid, or mixed solid and cystic forms. The majority of DT present as extra-abdominal mass (in the abdominal wall muscle) and, less commonly as intra-abdominally, occasionally can raise from any fibrous connective tissues in any location [1,2].

Several possible risk factors or associated conditions reported in the literature may attribute to the development of DT, including the history of DT in the family, exposure to irradiation, trauma, oral contraceptives bills use or pregnancy, some genetic mutation, and familial adenomatosis polyposis (FAP) in 10–20% of cases or Gardner syndrome. Nevertheless, there is no definitive aetiology of DT [1–7]. DTF tumours diagnosis is primarily based on pathological and immunohistochemical studies features. Surgical resection of the tumour is the primary curative option.

The presented case is a rare DT occupying the whole distal part of the pancreas close to the neck of the pancreas. Furthermore, it is considered the largest pancreatic mass to be ever reported in the literature and the first case of a pancreatic DT in Saudi Arabia, diagnosed in October 2016.

The work has been reported in line with the SCARE 2020 criteria: Agha RA, Franchi T, Sohrabi C, Mathew G, for the SCARE Group. The

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https://doi.org/10.1016/j.ijscr.2021.106327
Received 2 July 2021; Received in revised form 22 July 2021; Accepted 15 August 2021
Available online 19 August 2021

SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines, International Journal of Surgery 2020;84:226–230 [8].

## 2. Case presentation

A 42-year-old African origin male patient with no comorbidities, previous surgeries, drug history or history of trauma and no family history of cancer, Familial adenomatous polyposis syndrome or Gardner syndrome. His childhood was unremarkable, fully vaccinated, and never been hospitalised. Presented with complaints of recurrent left upper abdominal pain for more than ten years and transferred to our hospital for suspicious large splenic mass for investigation. The pain was disturbing his daily activities, sleeping, and associated with nausea and vomiting and constipation as well as loss of appetite. However, there was no history of fever, and the rest of the systemic review is unremarkable.

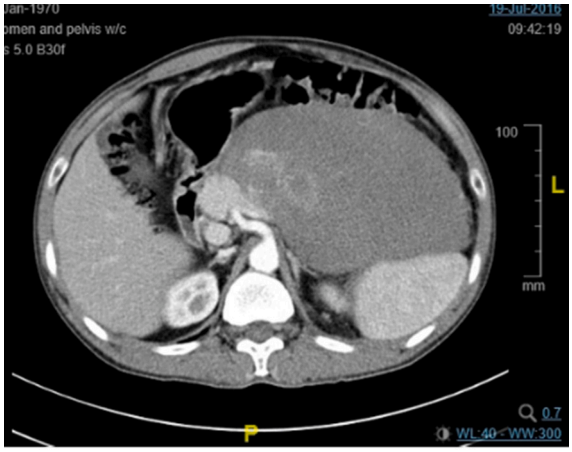
The patient looks underweights, conscious, oriented with stable vital signs, not pale or jaundiced by physical examination, and his cardio-respiratory system was unremarkable. There was no lymphadenopathy or scares. A large abdomen mass mildly tender, palpable at the left hypochondria area extends to the left hypochondrium, the umbilicus with a hard and smooth surface extending from the left upper quadrant of the abdomen till the hypochondriac region. By palpation, the mass was not mobile, stiff consistency, round borders, and smooth surface. No lower limbs findings, including varicose vein, oedema, or ischemic changes. All laboratory blood investigations revealed normal range values for the blood count, liver, and renal function tests.

Furthermore, all tumour markers were within normal ranges, e.g., CA19-9 equal to 7.10 U/ml. Contrast pancreatic protocol CT scan (Fig. 1) showed normal liver and spleen size and density. It revealed a large heterogeneous mass of pancreatic origin from the body and tail measuring about 23 cm by 15 cm by 11 cm (the pancreatic duct is compressed distally and not dilated proximally), displacing and compressing the stomach medially, spleen posteriorly and left kidney and invading the left adrenal as well. The small bowel loops were also displaced to the right side. No enlarged lymph nodes were seen. Biopsy was taken by trans-gastric approach under endoscopic ultrasound (EUS) guidance. The histopathology showed epithelial cells clusters and sheets of cells characterised by oval to round to spindle-shaped nuclei in Hg background; however, no malignant cells were demonstrated. The preliminary working diagnosis was most likely Interaabdominal fibromatosis of pancreatic origin for confirmation by detailed immunohistochemistry studies in the postsurgical resected specimen.

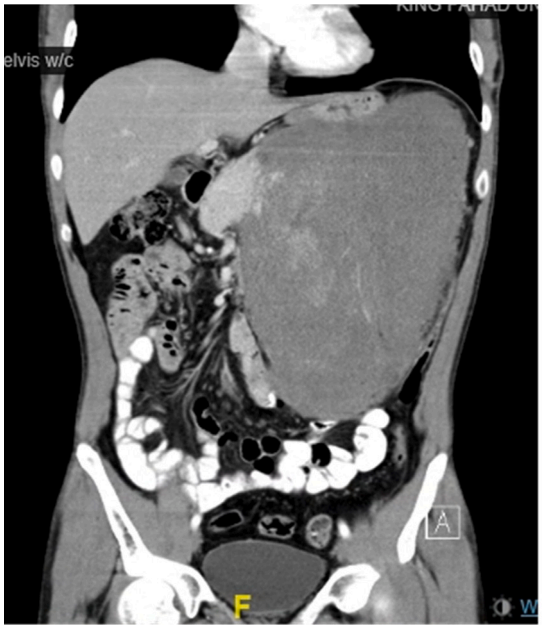
Preoperatively the patient was prepared for surgery and encouraged to take a high protein diet, and his electrolytes imbalance was corrected. The patient had radical antegrade modular pancreato-splenectomy (RAMPS) of the tumour through a midline laparotomy. The entire surgical procedure was performed by the author (hepatobiliary and multiorgan transplant consultant surgeon with ten years' experience post fellowship at that time, the training fellowship was three years clinical fellowship in Hepatobiliary and liver transplant in Royal Prince Alfred Hospital, Sydney, Australia and the University of British Columbia, Vancouver General Hospital, Vancouver, Canada).

During surgery, extensive adhesion and infiltration to adjacent organs were encountered. The pancreatic mass was invading nearby organs but assessed to be resectable safely; therefore, radical en mass resection with extended left pancreatectomy (1 cm Lateral to the neck of the pancreas), transverse colectomy. Splenectomy and left adrenalectomy (due to invasion by the tumour). Lymph node mapping included the gastrosplenic omentum nodes, the splenic hilum nodes, gastroduodenal ligament nodes, the infra-pancreatic nodes (behind the body of the pancreas), furthermore, dissection of the aortic lymph nodes around the celiac, superior mesenteric arteries, and above the left renal vein.

The estimated blood loss was 450 ml, and no blood was transfused intraoperatively. The patient had a smooth postoperatively recovery and




A




B

**Fig. 1.** A) a CT scan sagittal view with I/V & oral contrast showing huge heterogeneous hypodense mass originating from the pancreas, the body and the tail is replaced by the mass reported as measuring 23 cm in craniocaudal, 15 in the transverse plane and 11 cm in the anteroposterior plane. B) Coronal section demonstrates the pancreatic mass compressing the left kidney—no evidence of intraabdominal or pelvic lymph node enlargement.

resumed bowel sounds on the third postoperative day, and the diet progressed accordingly and mobilised on the second postoperative day. The hospital length of stay was ten days, and no significant events were recorded.

The accurate gross size of the tumour specimen was 26 × 17 × 9 cm,

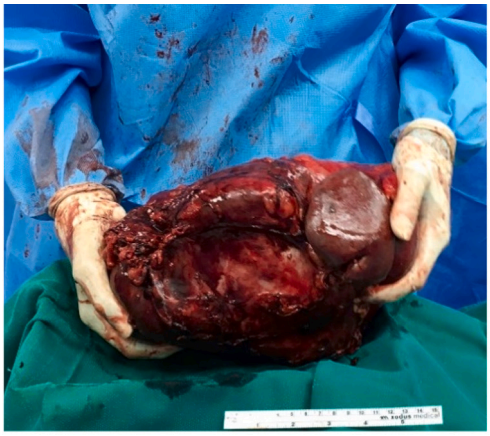
and the weight was found to be 3.6 kg (Fig. 2). The histopathology demonstrated Intera-abdominal Fibromatosis by immunohistochemistry positivity for B-catenin and C-Kit (CD117) and, as specific, negativity for S-100, Desmin, CD34, Bc12, CD99, ALK-1, Pan-Ck. All surgical resection margins were free of tumour, no metastasis to adjacent organs and eleven lymph nodes examined from the specimen showed no malignant cell (Fig. 2).

Because the resection was curative and, the histopathological nature of the tumour and the negative lymph node and no distant metastasis, no chemo- or radiotherapy recommended for the patient in the tumour-board decision, the three-month follow-up CT scan and annual ultrasound for five years did not demonstrate any recurrence; therefore, curative management is achieved. Furthermore, the patient had excellent satisfaction and improved quality of life, as he stated and confirmed by the oncologist. He did not develop diabetes and never required any hypoglycaemic agent during the entire postoperative period.

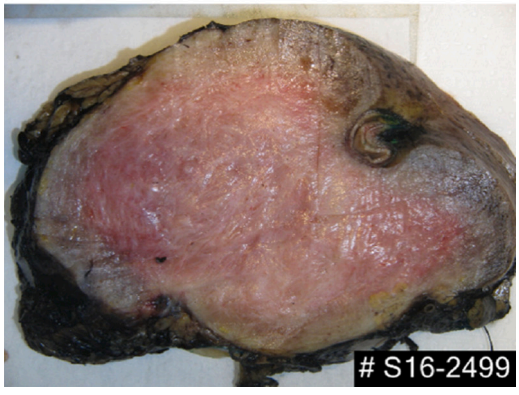
## 3. Discussion

DTF tumours are rare benign fibrous tumours that account for approximately 0.03% of all neoplasms in the USA. The pancreas is an even more rarely reported subset with around 5% of all DTF [9–11].

The unique aggressive local invasion is characteristic of DT that render the tumour unresectable or with high local recurrence, but not unknown to case distant metastases as thes is a benign tumour.



A).




B).

**Fig. 2.** Intraoperative photo demonstrates the gross specimen, including the distal pancreas, spleen, part of the transverse colon, and the adrenal gland. B). The cut surface is whirly homogenous tan to pink to white, reaching the spleen capsule but not invading it. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

These tumours are more commonly present in young adolescent patients [5,12]. Intraabdominal DT occurs in around 8% of all DT and is more likely to be in adults than in paediatric age groups, affecting any part of the gastrointestinal and genitourinary tracts [13]. The commonest origin of intraabdominal DT is from mesenteric or retroperitoneum connective tissue. The pancreatic origin DT remains an extremely rare subset of DTs. The English literature from the 1980s report 27 pancreatic DTs, and therefore our case is case number 28th [1,2,4,5,11,14–19].

Clinical presentation of intraabdominal DTs is none specific in symptoms and signs. In general, the clinical findings are related to the anatomical location of the DT mass or invasion of nearby structures, causing bowel obstruction, ischemia, fistulas, gastrointestinal bleeding or perforation, and ureteric obstruction. Pancreatic-origin DT behaves like pancreatic cancer in presentation as asymptomatic mass or nonspecific mild epigastric pain or painless jaundice if the tumour location is in the head or the uncinate process [1,5]. Our patient presented with mild to moderated abdominal pain associated with nausea, weight loss and splenic mass for investigation [20].

Imagining is valuable, especially CT scans or MRI, to determine invasiveness, aid in biopsy approach and assess resectability but not diagnostic [1,4,5].

In FNA histopathology, as in our case, the presence of spindle cells is frequent but not pathognomonic for DT. The Immunohistochemistry feature of intraabdominal DT is characterised by positive vimentin and β-catenin. It is characteristically negative for S100, CD34, CD99 and Bcl2 and Focal reaction (positive or negative) for CD117, SMA, Desmin, Caldesmon, SMA (smooth muscle myosin antibodies), estrogen receptors (ER), and PGR. DT can be further confirmed by demonstrating mutations of β-catenin gene in exon three, which is positive in 85% of the case is an essential diagnostic feature [5,12,14,21].

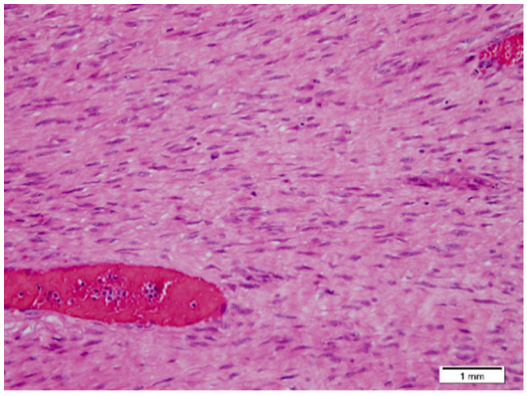
Our case was positive for CD 117; this finding was reported in earlier studies and demonstrated high positivity of 75% of CD117 (c-kit) in intraabdominal DT (Fig. 3). However, other more recent larger volume studies indicated either none or very low incidence of positive CD 117. They postulated to considered intrabdominal DT as a CD117-negative tumour; therefore, no role of a c-Kit tyrosine kinase inhibitor therapy in management, i.e., matinib mesylate Gleevec (STI571, Novartis, Basel, Switzerland) [22–25].

Favourable prognosis reported by Jia et al. with overall survival of 100% and disease-free survival of more than 80% [5]. However, the accurate prognosis is not well reported, and sporadic cases reported a high postoperative recurrence rate around 19–77%, mainly when associated with Gardner syndrome and FAP with recurrence rate up to 90% [5,17].

To date we lack consensus or guidelines for the management of DT. The first-line management of DT was and remained wide free margin complete resection (radical resection) given the aggressive nature and excessive local invasive behaviour of the tumour with the guidance of frozen section support considering the locally invasive behaviour of this tumour. Observation without surgical resection, also called the wait-and-see approach, mainly in the static tumour were recommended by some experts to avid morbidity and the possible high recurrence associated with surgery, especially in asymptomatic cases with mild, manageable symptoms.

As a possible second line of management and specifically in high-risk patients for surgical intervention or in advance stages, adjuvant systemic chemotherapy is based primarily on a combination of methotrexate, vinblastine doxorubicin, and dacarbazine and/or radiotherapy and molecular target therapy as Tamoxifen. The use of COX2 inhibitors, e.g., NSAID and celecoxib, demonstrated promising successes in some studies; however, the mechanism by which it achieves regression is not reported [4,5,17,20].

A).

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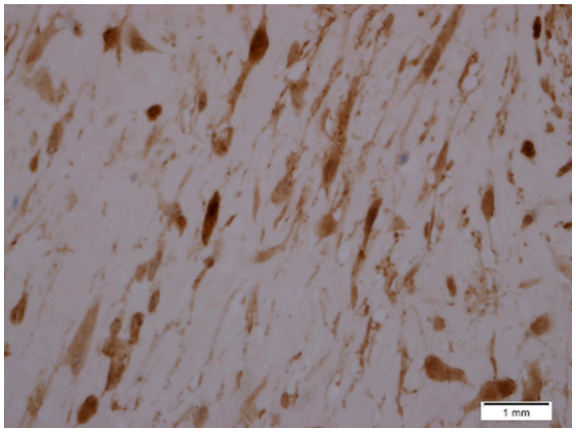


**Fig. 3.** Histopathology showing. A). Interaabdominal fibromatosis. All surgical resection margins are free of tumour size 26 × 17 × 9 cm.
B). Immunohistochemical staining of the tumour showing positive satin for anti-beta-catenin ×400.

## 4. Conclusion

DT in the pancreas is an exceedingly rare form of already rare intraabdominal form of desmoid type fibromatosis. Mainly due to nonspecific clinical presentation or asymptomatic incidentally discovered mass, secondly, the lack of specific clinical investigation, the diagnosis of DT is considered challenging. Pathological Immunohistochemical staining demonstrating positive β-catenin nuclear or negative S100 protein is currenly the principal diagnostic tool .

Radical-margin-free surgical resection remains the mainstay management approach. At the same time, chemotherapy and/or radiotherapy are not effective, and no standardised protocol of consensuses and considered the last resort for only high risk for or in advanced cases.

Long-term follow up is warranted due to the recurrence behaviour of the tumour. As demonstrated in this case, tumour size and extensive invasion are not contraindications to expert hands' curative resection.

Our case presents the largest pancreatic DTF to be published in the midline English literature.

## Abbreviations

| | |
|---|---|
| DT | desmoid tumour |
| CT scan | computer tomography scan |

## Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. Or from any other source.

## Ethics approval

It was waived because it's a case report according to the policies of the affiliated University institutional review committee.

## Consent

Written informed consent was obtained from the patient to publish this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Authors' contributions

**Hanan Alghamdi**: performed the operation, study concept, study design, data collection, writing the paper, reviewing and editing the final manuscript.

## Research registration

not appliacble

## Availability of data & material

Available.

## Guarantor

The author is the Guarantor who accepts full responsibility for the work and the conduct of the study; I had access to the data and controlled the decision to publish it.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## Declaration of competing interest

The author has no conflict of interest to declare or commercial associations.

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