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How to cite

LITCHINKO, Alexis et al. Large Desmoid Tumor of the Pancreas: A Report of a Rare Case and Review of the Literature. In: The American journal of case reports, 2022, vol. 23, p. e937324. doi: 10.12659/AJCR.937324

This publication URL: https://archive-ouverte.unige.ch/unige:166548
Publication DOI: 10.12659/AJCR.937324

American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2022; 23: e937324
DOI: 10.12659/AJCR.937324

Received: 2022.05.23
Accepted: 2022.09.05
Available online: 2022.10.17
Published: 2022.11.15

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Financial support: None declared
Conflict of interest: None declared

Patient: Male, 41-year-old
Final Diagnosis: Desmoid tumor of the pancreas
Symptoms: Abdominal discomfort
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Desmoid tumor (DT), also known as aggressive fibromatosis or desmoid-type fibromatosis, is a rare disease. It can occur in anyone at any age, and is more likely to appear in female patients. DTs are sometimes associated with familial adenomatous polyposis and rare syndromes such as Gardner syndrome. Arising from specific cells, fibroblasts, they tend to develop in patients with a history of abdominal surgery and rarely produce metastases.

Case Report: We present a case of a 41-year-old man who was referred for abdominal discomfort with no digestive or general symptoms. An abdominal CT scan revealed a mass in the left hypochondrium, corresponding to an intraperitoneal tumor extending to left colon. Based on MRI, we suspected a gastrointestinal stromal tumor. Colonoscopy showed no intraluminal tumor in the colon. A PET-CT scan revealed tumor hypermetabolism and no metastases. The mass was diagnosed as a DT after percutaneous biopsy. Six weeks after diagnosis and as the tumor continued increasing despite pharmacological treatment, the patient underwent surgical pancreatic tail resection with splenectomy and left colonic segmentectomy. Histological examination revealed a 7.047-g DT with severe infiltration of pancreatic parenchyma and transmural colic barrier, with no high-grade differentiation and negative resection margins. The postoperative recovery was uneventful, and we proposed surveillance with MRI.

Conclusions: The DT was surgically removed and patient remains under MRI surveillance. Other reported management approaches consist of radiotherapy, tyrosine kinase inhibitors, anti-hormonal therapies/non-steroidal anti-inflammatory drugs, chemotherapy, or close surveillance only. This is, to our knowledge, the largest and heaviest DT reported in the modern literature.

Keywords: Colorectal Surgery • CTNNB1 Protein, Human • Fibromatosis, Aggressive • Gardner Syndrome • Radiotherapy

Abbreviations: **DT** – desmoid tumor; **CT scan** – computed tomography scan; **FAP** – familial adenomatous polyposis; **NSAID** – non-steroidal anti-inflammatory drug; **ECOG PS** – Eastern Cooperative Oncology Group Performance Status

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/937324

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Background

Desmoid tumors (DTs) are rare tumors which develop from the connective tissue: well-differentiated mesenchymal fibroblasts or myofibroblasts. DTs represent < 3% of all soft tissue tumors and have an estimated incidence of 2-4 new cases per million population per year [1]. Also known as aggressive fibromatosis or desmoid-type fibromatosis, DTs are benign tumors which can develop anywhere in the body, invade surrounding tissues, and can be very difficult to control because of their rapid growth. McFarlane was the first to describe a DT, which was located in the abdominal wall of a young patient in 1832 [2]. The WHO classifies DTs as intermediate (locally aggressive) fibroblastic and myofibroblastic tumors [3].

DTs can localize intra- or extra-abdominally, mostly in the abdominal wall, and there is no evidence of their metastatic potential. DTs typically affect young adults between 20 and 30 years of age [4]. The clinical presentation of DTs ranges from fully asymptomatic to swelling, left hypochondrium pain, epigastric pain, nausea, vomiting, and back pain. The etiology of these tumors is not well understood and they have 2 typical presentations: sporadic, mostly associated with post-traumatic (postoperative) etiology [1] or associated with Gardner syndrome, a subtype of familial adenomatosis polyposis (FAP). FAP develops in patients with mutations in the APC gene [5]; 20% of these APC mutations are de novo [6]. In some rare cases, DTs are associated with the peripartum period.

Specific factors have been identified as abnormalities in histological analysis showing fibroblastic proliferation and then confirmed by immunohistochemical staining for β-catenin, as the β-catenin gene (CTNNB1), present in more than 75% of DTs cases, is mutated in FAP or Gardner syndrome [7].

There is no standard of care for DTs, but the first-line pharmacotherapy consists of selective estrogen receptor modulators such as Tamoxifen or Toremifene and non-steroidal anti-inflammatory drugs (NSAID) as well as cytotoxic chemotherapy and sometimes radiotherapy [1]. Treatment with oral tyrosine kinase inhibitor drugs such as Imatinib or Sorafenib has shown promising results [4,8]. An emerging technique uses high-intensity ultrasound waves to remove small DTs. This non-invasive approach can be combined with other treatments [9,10].

However, in the vast majority of symptomatic cases, surgery plays a major role in the treatment of DTs. In 25-60% of patients who undergo surgery, DTs recur locally [11], most frequently near the surgical resection. It is well accepted that the aim of surgery is to remove the entire tumor with R0 resection margins and minimize the risk of recurrence. Based on data from 495 patients with DTs, physicians at Memorial Sloan-Kettering Cancer Center have developed a nomogram that can be used to evaluate that risk [12].

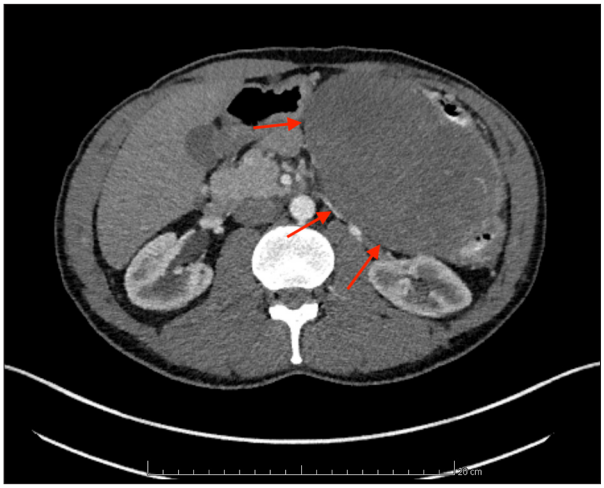

Figure 1. CT scan with contrast in arterial phase showing the DT in the left hypochondrium bulging from the pancreas; axial view. Red arrows indicates the tumor mass.

We report the largest case of pancreatic DT, diagnosed on 22 November 2021, at the Geneva University Hospitals, Switzerland. The DT was removed surgically with negative resection margins. We expand this case report with a literature review on DTs of the pancreas.

The work has been reported in line with the SCARE 2020 criteria: Updating Consensus Surgical CAse REport (SCARE) Guidelines, International Journal of Surgery 2020;84: 226-230 [13].

Case Report

A 41-year-old male patient with no significant medical or surgical history apart from an asymptomatic megaureter and a bicuspid aortic valve presented to his family physician with nausea and diarrhea persisting for 1 week, 3 months prior to our consultation. He was prescribed simple symptomatic management. Because of the persisting abdominal discomfort and postprandial nauseas, the patient was referred to the Division of Digestive Surgery of the University Hospitals of Geneva. We noted a visible bulking in the epigastric and left hypochondriac region. The mass was easily palpable and solid, without any signs of abdominal guarding. The patient was in good condition, height 178 cm, weight 77 kg, and ECOG PS 0. He reported no active smoking or alcohol consumption, no allergies, and no regular medication. The patient had no personal or family history of desmoid tumor. The first abdominal CT scan (**Figure 1**) showed a massive intra-abdominal heterogeneous tissue tumor located in the left hypochondrium and invading the left colon. We also observed a moderate amount

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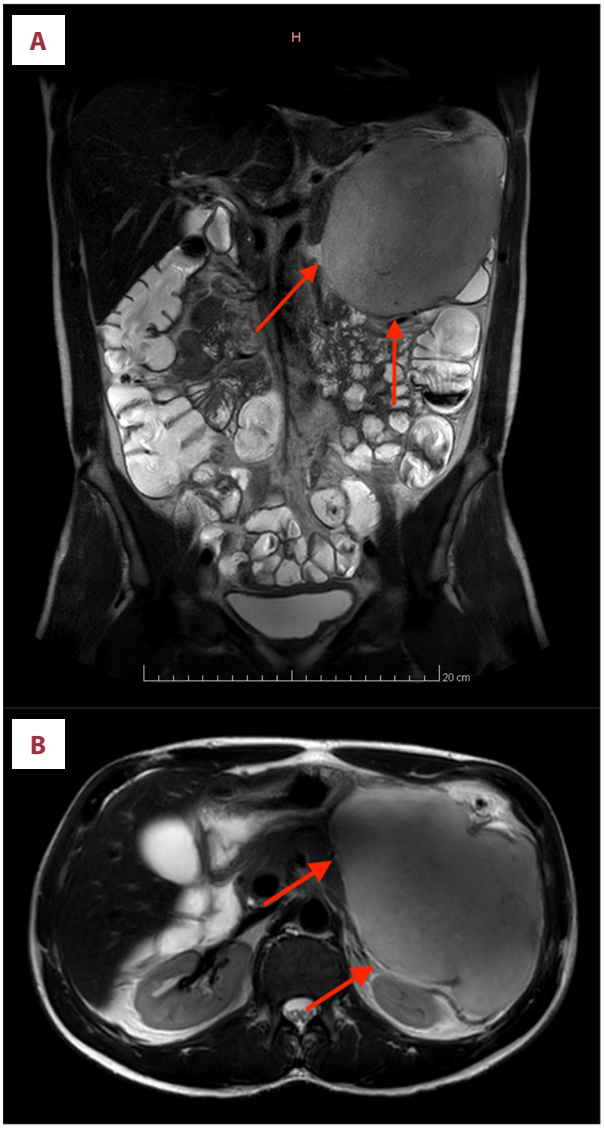

Figure 2. First MRI images in T2 sequence showing the pancreatic mass compressing the left colic flexure. Red arrows indicates the tumor. (**A**) Frontal view. (**B**) Axial view.

of intra-abdominal free fluid and suspected the tumor to be a gastrointestinal stromal tumor. We then performed an MRI (**Figure 2**), which revealed that the left hypochondrium mass was mildly heterogeneous with T2 hypersignal and T1 hyposignal of unclear origin, with restricted diffusion, mild enhancement after contrast injection, and encapsulated, and was merging with the tail of the pancreas and compressing the left colic flexure. Colonoscopy was normal apart from a severe extrinsic compression localized in the left colic flexure. To explore the potential malignancy and metastases of the tumor, a whole-body PET/CT scan was performed (**Figure 3**). It revealed a hypermetabolic tumor of more than 20 cm extending from the left pancreas and invading the left colic flexure, and no other

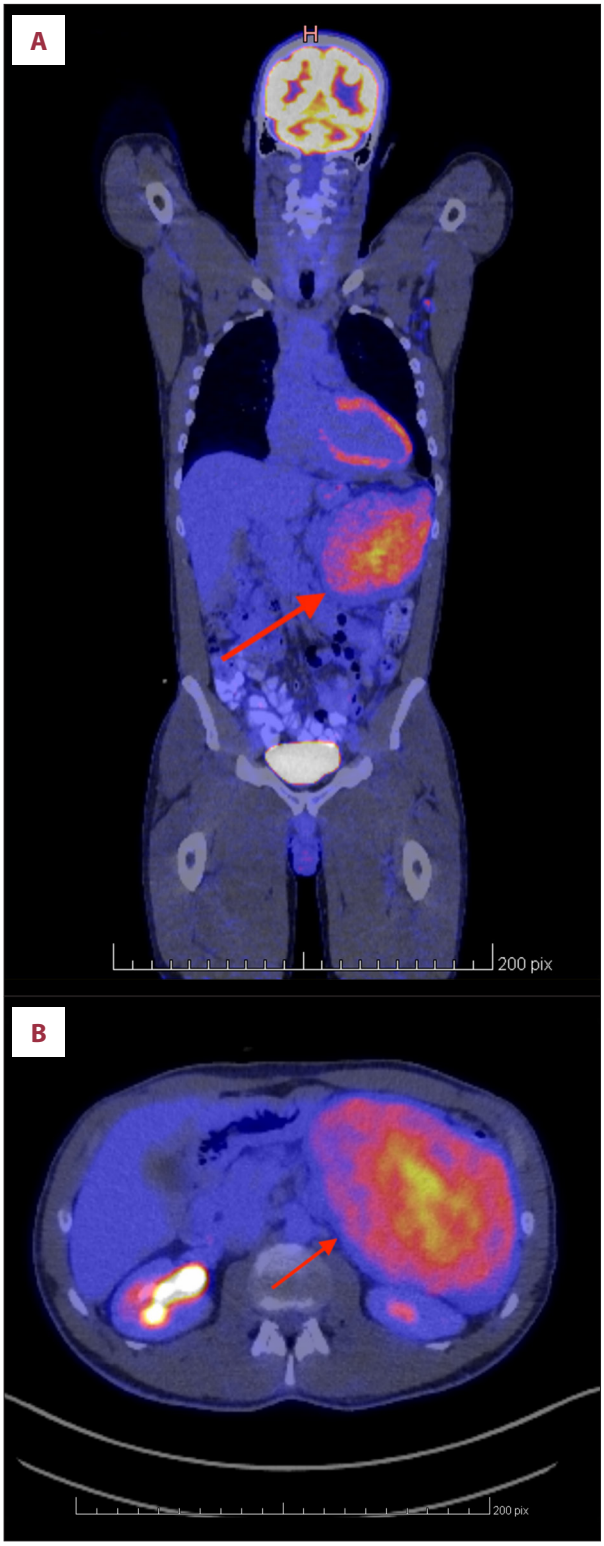

Figure 3. PET/CT scan showing hypermetabolic signal of the pancreatic mass and absence of secondary tumors. Red arrows indicate the hypermetabolic tumor. (**A**) Frontal view. (**B**) Axial view.

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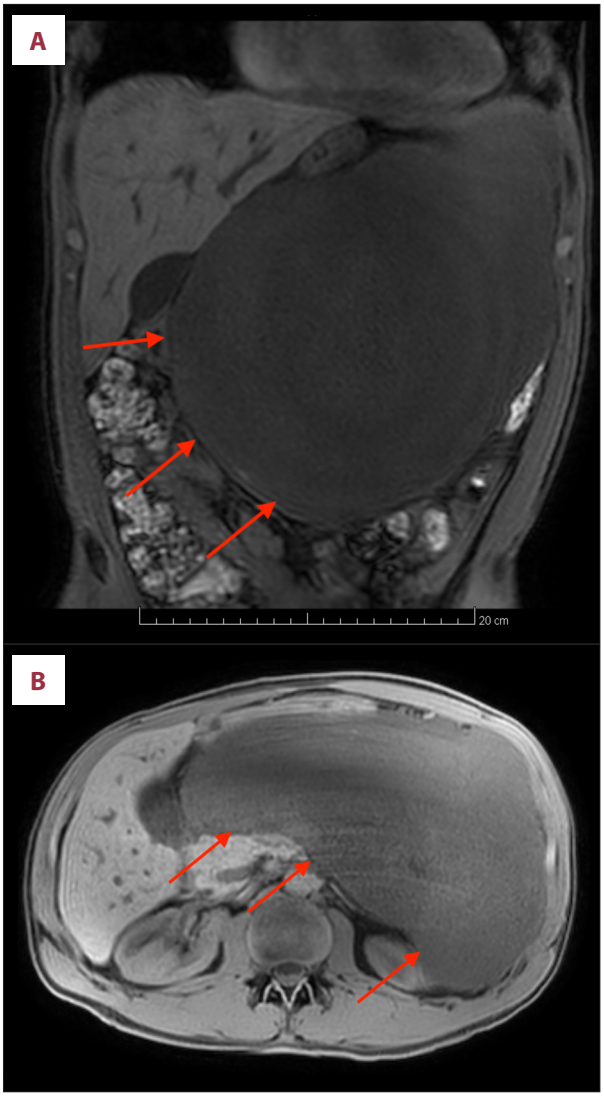

Figure 4. Second MRI image showing the expansion of the pancreatic mass. (**A**) Frontal view in T1 sequence. (**B**) Axial view contrast enhancement.

suspect hypermetabolic lesions. Histological and immunohistochemical images of a percutaneous biopsy were compatible with an active DT.

The case was presented to our multidisciplinary care team, who opted for a conservative initial management with sulindac (NSAID) and close surveillance. One month after this decision, the patient presented with an increased abdominal circumference and alternating diarrhea and constipation. A new MRI showed that the tumor had rapidly grown from 7 to more than 18 cm (**Figure 4**), with T2 hypersignal and T1 hyposignal. The case was once again presented to our multidisciplinary care team, who suggested a surgical "en-bloc" resection.

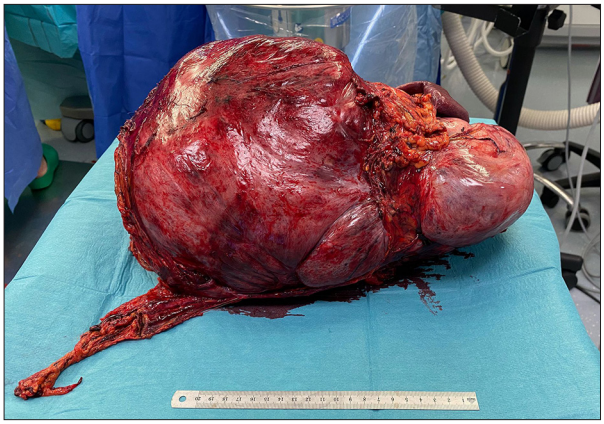

Figure 5. Intraoperative picture of the resected tumor.

The patient underwent surgery 3 months after the initial CT scan. He underwent midline laparotomy and exploratory laparotomy, which revealed no suspect secondary lesions. The mass was bulging from the left hypochondrium and infiltrated the left colic flexure and the spleen. We performed a meticulous adhesiolysis to isolate the whole tumoral mass, a left pancreatectomy, division of the pancreas with a 60-mm stapler Echelon™ (Johnson & Johnson), with splenectomy and segment resection of the left colic flexure with direct latero-lateral anastomosis. The mass was resected "en-bloc" (**Figure 5**) and weighed 7.057 g (**Figure 6**). The surgery lasted 264 min, blood loss was estimated to be 300 cc, and the patient did not receive any transfusion. Postoperative care was uneventful, with a standard application of the ERAS® protocol of our institution [14] and the patient was discharged on day 5.

Pathology and immunohistochemistry examination of the resected mass revealed a DT measuring 40 cm. It consisted of elongated, thin, tapered cells of uniform appearance with pale cytoplasm, embedded in a collagenous stroma. There was no nuclear hyperchromasia, cytological atypia, or mitosis. Blood vessels were thin-walled, with perivascular edema and vascular microhemorrhages (**Figure 7A**).

We observed nuclear β-catenin staining (**Figure 7B**). Smooth muscle actin (SMA) was weakly positive and desmin was negative. DNA sequencing revealed that the CTNNB1 (β-catenin) gene carried the p.T41A (c.121A>G) mutation. The tumor invaded almost the whole pancreatic parenchyma and infiltrated the colonic wall, but circumferential resection margins and organ resection margins were negative.

We proposed active close surveillance with MRI every 3-6 months, because even with the negative resection margins the risk of recurrence seems to be proportional to the initial size of the tumor. The patient had 2 follow-up MRI scans at 6 and 12 months after surgery, without any sign of recurrence and without hepatic, pancreatic, or colic nodules.

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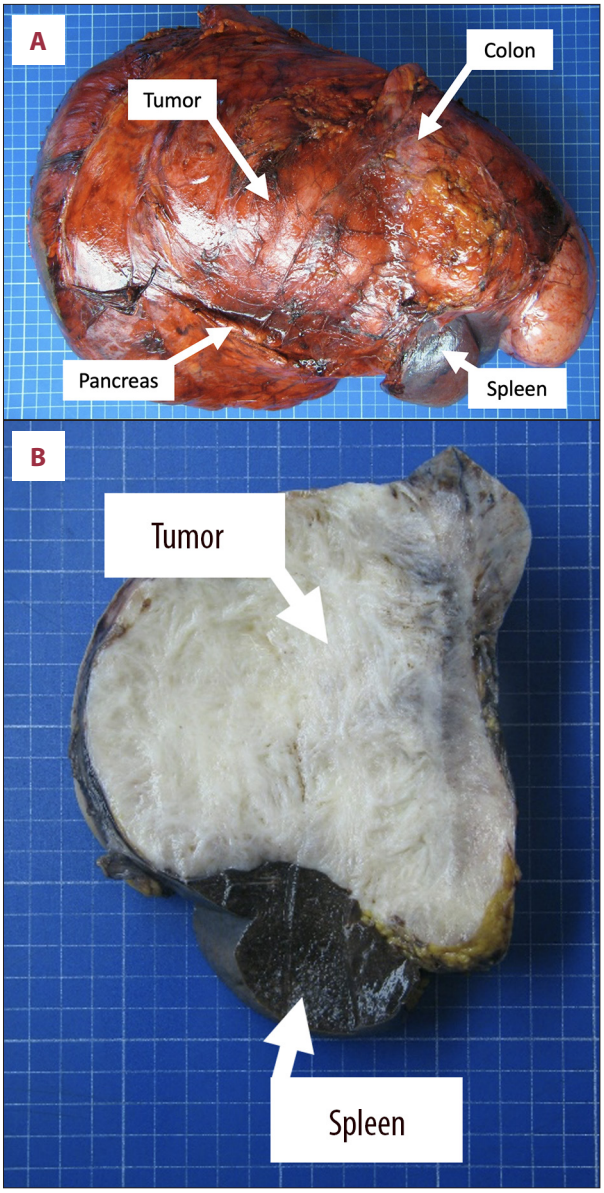

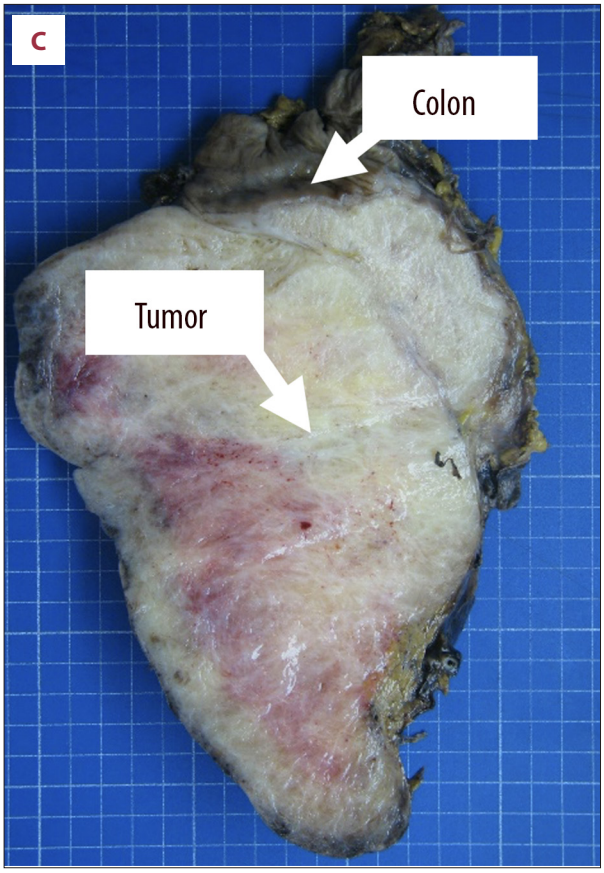

Figure 6. Macroscopy of the resected tumor. (**A**) Resected tissue weighing 7.057 g, consisting of a large tumor measuring 40×31 cm and a spleen segment measuring 12×10 cm, a colonic segment measuring 25×3 cm, and a pancreatic segment measuring 25×3 cm. (**B, C**). The mass protruding through the colonic wall.

Discussion

A search of the English-language scientific literature yielded 30 reports of DTs which developed at the expense of the pancreas (**Table 1**) [7,15-39]. The mean age at diagnosis was 42.1 years old [0.4-75] and 61.3% of patients were male. The mean tumor size was 7.99 cm and, except for Bruce et al [16] and Alghamdi et al [39], none of the reports specified the weight of the tumor; 26% of tumors were totally asymptomatic, and 58% of patients had unspecific epigastric pain or discomfort. This case is, to our knowledge, the heaviest DT of the pancreas ever reported in the English-language scientific literature.

The road to clear diagnosis of DT is often long and winding – from clinical examination of unspecific symptoms through various imaging techniques to histological examination. As DTs are rare in routine practice, it is important to remember their frequent association with FAP, attenuated FAP, or Gardner syndrome. Quintini et al [40] showed that 10-25% of patients with mutations in the APC gene develop DTs and that these tumors are an important cause of mortality after total colectomy. We recommend that patients diagnosed with DTs be referred to an experienced multidisciplinary team. Even if the guidelines are unclear, we strongly recommend a surgical approach for tumors larger than 10 cm, grow rapidly, or, most importantly, are symptomatic. Surgeons should always strive to achieve negative resection margins, but even then, the recurrence of DTs is frequent (25-60%) [11]. DTs can also be challenging to treat surgically, especially when located in tight spaces such as around the pancreas.

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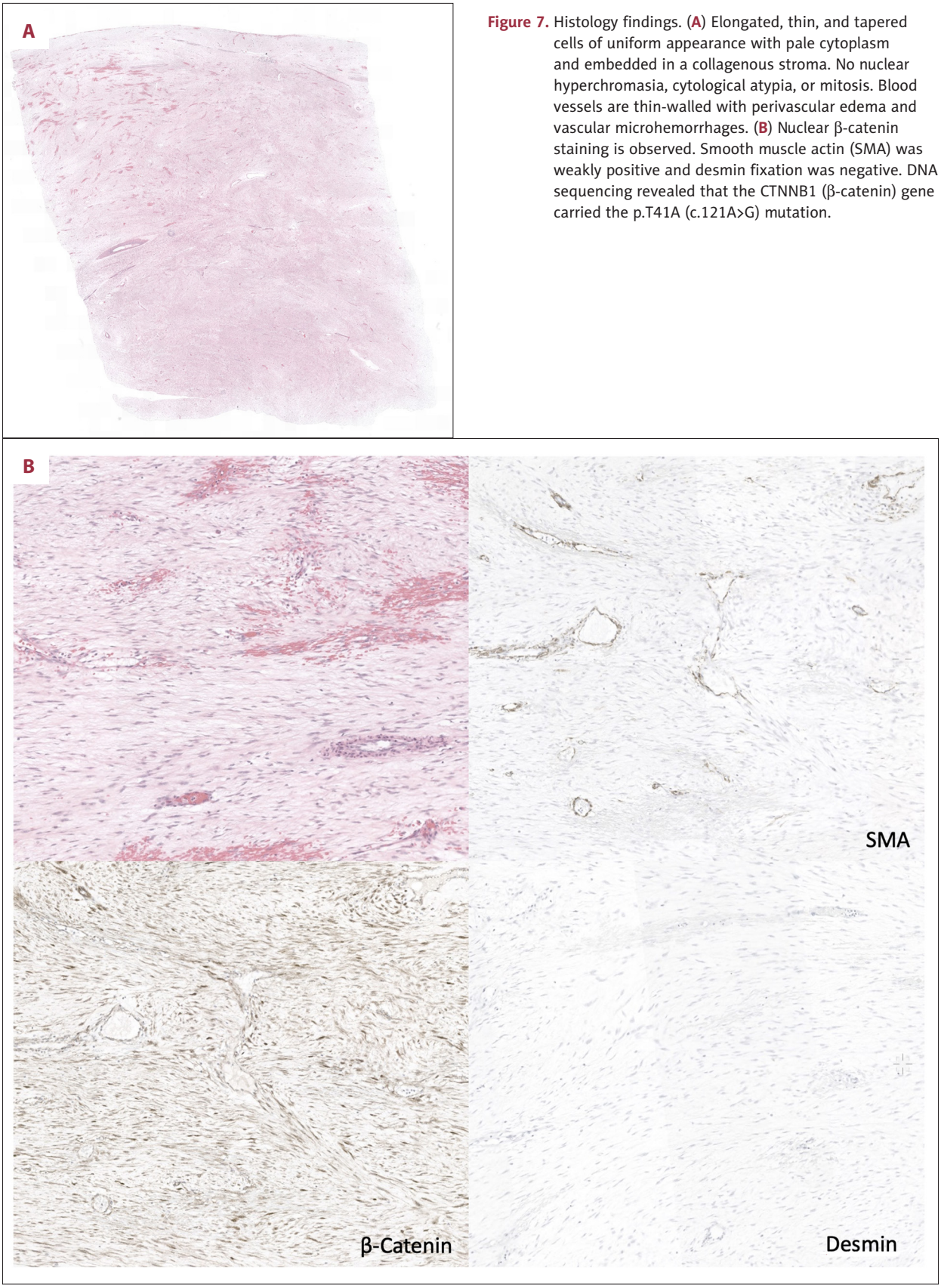

Figure 7. Histology findings. (**A**) Elongated, thin, and tapered cells of uniform appearance with pale cytoplasm and embedded in a collagenous stroma. No nuclear hyperchromasia, cytological atypia, or mitosis. Blood vessels are thin-walled with perivascular edema and vascular microhemorrhages. (**B**) Nuclear β-catenin staining is observed. Smooth muscle actin (SMA) was weakly positive and desmin fixation was negative. DNA sequencing revealed that the CTNNB1 (β-catenin) gene carried the p.T41A (c.121A>G) mutation.

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Table 1. Summary of the previous case reports of pancreatic desmoid tumors and the present case.

Publication [reference no]	Year	Age [years]	Gender	Symptoms
Roggli et al [15]	1980	0.33	M	Tachypnea/fever
Bruce et al [16]	1996	38	M	Asymptomatic
Sedivy et al [17]	2002	68	F	Weight loss/nausea
Nursal et al [18]	2003	39	M	Epigastric/back pain
	2003	25	F	Epigastric/back pain
Pho et al [19]	2005	17	M	Weight loss/chest pain
Weiss et al [20]	2006	63	M	Abdominal fullness/epigastric pain
Amiot et al [21]	2008	51	F	Weight loss, epigastric pain with dorsal irradiation and nausea
Polistina et al [22]	2010	68	M	Asymptomatic
Rao et al [23]	2013	11	M	Asymptomatic
Xu et al [24]	2013	17	M	Epigastric pain/nausea
Kim et al [25]	2014	33	F	Abdominal pain
	2014	49	F	Abdominal discomfort
	2014	72	M	Asymptomatic
	2014	40	M	Abdominal pain
Jia et al [26]	2014	41	M	Weight loss/epigastric pain
Gerleman et al [27]	2015	63	F	Abdominal pain
Słowik-Moczydłowska et al [28]	2015	13	M	Abdominal pain in left hypochondrium
Tsukamoto et al [29]	2016	75	F	Asymptomatic
Wang et al [30]	2016	57	F	Weight loss/epigastric pain
Jafri et al. [31]	2017	54	F	Weight loss/dysphagia
Torres et al [7]	2017	15	M	Abdominal pain/nausea
Dallaire et al [32]	2018	39	F	Abdominal pain in left hypochondrium
Khanna et al [33]	2020	45	M	Abdominal pain/nausea
Shayesteh et al [34]	2020	66	M	Asymptomatic
Xuesong et al [35]	2020	17	M	Abdominal pain/melena
Sugimachi et al [36]	2020	52	F	Asymptomatic
Stone et al [37]	2020	72	M	Asymptomatic
Park et al [38]	2021	23	F	Epigastric pain
Alghamdi et al [39]	2021	42	M	Abdominal pain in left hypochondrium
Present case	**2021**	**41**	**M**	**Abdominal pain in left hypochondrium**

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Table 1 continued. Summary of the previous case reports of pancreatic desmoid tumors and the present case.

Publication [reference no]	Type of surgery	Size [cm]	Weight [g]	Follow-up	Outcome
Roggli et al [15]	Autopsy	N/A	N/A	N/A	Death
Bruce et al [16]	Radical resection pancreatic mass	5.0×4.5×2.1	28	2 y.	No recurrence
Sedivy et al [17]	Radical resection pancreatic mass	1.5	N/A	N/A	N/A
Nursal et al [18]	Incisional biopsies of the pancreas	7.5×4.0	N/A	N/A	N/A
	Pancreas/adnexal masses biopsied	8.5×5.0	N/A	N/A	N/A
Pho et al [19]	Left pancreatectomy with spleen preservation	2.8×4.2	N/A	2 y.	Recurrence
Weiss et al [20]	Distal pancreatectomy and splenectomy	6.5	N/A	9 m.	No recurrence
Amiot et al [21]	Distal splenopancreatectomy with en-bloc resection of the splenic colonic flexure	6.0	N/A	18 m.	No recurrence
Polistina et al [22]	Left pancreatectomy with spleen preservation	5.0	N/A	5 y.	No recurrence
Rao et al [23]	Distal pancreatectomy and splenectomy	10.0×8.8×7.5	N/A	10 m.	No recurrence
Xu et al [24]	Central pancreatectomy	8.6×6.0	N/A	40 m	No recurrence
Kim et al [25]	Pancreaticoduodenectomy	2.3	N/A	14 m.	Alive*
	Left pancreatectomy	1.5	N/A	9 m.	Alive*
	Left pancreatectomy	5.0	N/A	4 m.	Alive*
	Biopsies of the pancreas	8.0	N/A	17 m.	Alive*
Jia et al [26]	Pancreaticoduodenectomy	1.9	N/A	2 y.	No recurrence
Gerleman et al [27]	Left pancreatectomy with spleen preservation	4.8×5.8×5	N/A	1 y.	No recurrence
Słowik-Moczydłowska et al [28]	Distal splenopancreatectomy with en-bloc resection of the left colonic flexure	10×10	N/A	19 m.	No recurrence
Tsukamoto et al [29]	Distal pancreatectomy and splenectomy	8	N/A	21 m.	No recurrence
Wang et al [30]	Bypass surgery/COX-2 inhibitor celecoxib	10	N/A	2 y.	No recurrence
Jafri et al. [31]	Pancreaticoduodenectomy	5.2×4.2	N/A	N/A	N/A
Torres et al [7]	Radical resection pancreatic mass	20.0×14.0	N/A	2 m.	No recurrence
Dallaire et al [32]	Distal pancreatectomy and splenectomy	9.2	N/A	N/A	N/A
Khanna et al [33]	Distal pancreatectomy with splenectomy, partial gastrectomy and partial duodenectomy	12.0×10.0×10.0	N/A	N/A	N/A
Shayesteh et al [34]	Distal pancreatectomy and splenectomy	3.0	N/A	8 m.	No recurrence
Xuesong et al [35]	Distal pancreatectomy with splenectomy and a partial gastrectomy	N/A	N/A	6 m.	No recurrence
Sugimachi et al [36]	Left pancreatectomy with spleen preservation	20	N/A	N/A	N/A
Stone et al [37]	Distal pancreatectomy and splenectomy with resection of the posterior gastric wall	5.6	N/A	6 m.	No recurrence

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Table 1 continued. Summary of the previous case reports of pancreatic desmoid tumors and the present case.

Publication [reference no]	Type of surgery	Size [cm]	Weight [g]	Follow-up	Outcome
Park et al [38]	Distal pancreatectomy and splenectomy and partial resection of the stomach, transverse colon and 4th portion of the duodenum	10.0	N/A	3 y.	No recurrence
Alghamdi et al [39]	Extended left pancreatectomy and splenectomy, transverse colectomy and left adrenalectomy	26.0×17.0×9.0	3600	5 y.	No recurrence
Present case	**Extended left pancreatectomy and splenectomy with partial left colectomy**	**40.0×31.0.×11.0**	**7065**	**3 m.**	**No recurrence**

* No mention of recurrence; N/A – non-applicable; m. – months; y. – years.

Conclusions

The presented case is the largest DT ever reported. Its management was successful, with no sign of recurrence at 1-year follow-up. A review of the literature on DTs of the pancreas showed that these rare tumors are still mostly managed surgically. We strongly believe that the management approach for intra-abdominal DTs should be developed by a multidisciplinary panel. New developing techniques, such as high-intensity focused ultrasound [9,10], should also be considered.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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