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## Case Report

# Pancreatic desmoid tumor: A rare case with radiologic–pathologic correlation

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## ABSTRACT

A 39-year-old female patient was referred to our tertiary oncologic center for additional investigations relating to a suspect pancreatic tail lesion. An abdominal computed tomography scan and magnetic resonance imaging scan showed a solid lesion demonstrating progressive enhancement. Complete resection was obtained and the final diagnosis was that of a desmoid tumor of the pancreas, an exceptionally rare tumor demonstrating overlap with other solid and cystic lesions of the pancreas [1]. Therefore, it is important to recognize the essential role of pathology, particularly immunohistochemistry, in identifying this tumor. The high rate of postsurgical recurrence should prompt repeated follow-ups considering the potential aggressive nature of desmoid tumors.

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## 1. Case report

A 39-year-old female patient who complained of left flank pain was referred to our surgical oncologic team for assessment of a pancreatic tail lesion discovered following an abdominal ultrasound. Patient was known for a history of non-Hodgkin lymphoma treated with chemotherapy 6 years prior. A magnetic resonance imaging was first requested to further characterize the lesion. The report described a large mass (6.7 × 5.2 × 5.7 cm) centered on the tail of the pancreas. This

lesion was hyperintense with a hypointense nodular capsule on T2-weighted images and heterogeneous, predominantly hypointense, on T1-weighted images (Figs. 1a and b). After gadolinium injection, the lesion demonstrated progressive enhancement on the venous and late phase, being maximal on the latest (Figs. 2a and b). The lesion showed mass effect on the splenic vein, which remained permeable, and the pancreatic duct which was dilated upstream. The rest of the pancreas was normal. Diagnostic differential included a neuroendocrine tumor or a pseudopapillary tumor of the pancreas. Characteristics were not typical of a lymphoma and no lymphadenopathy was seen.

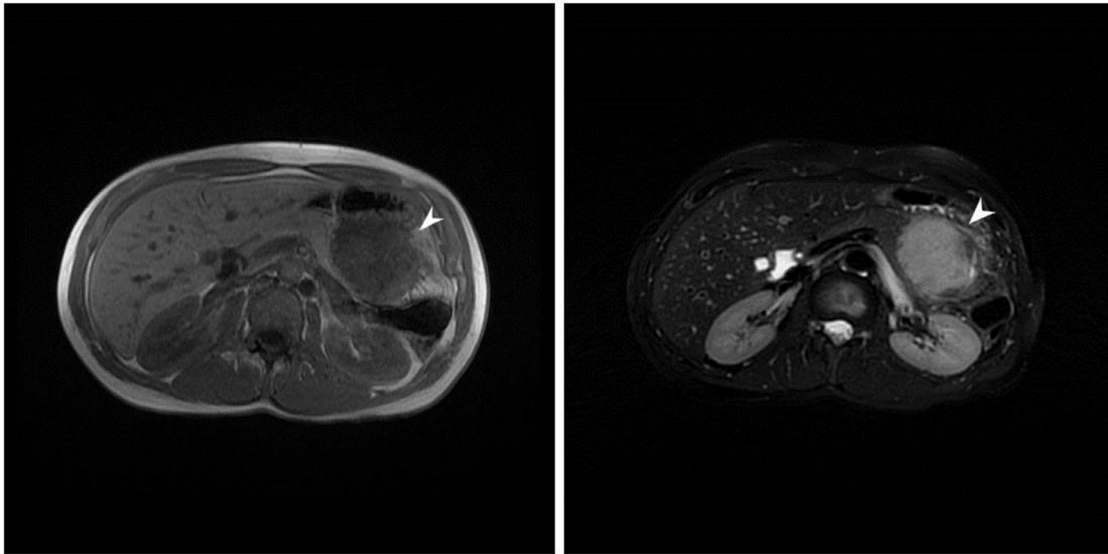
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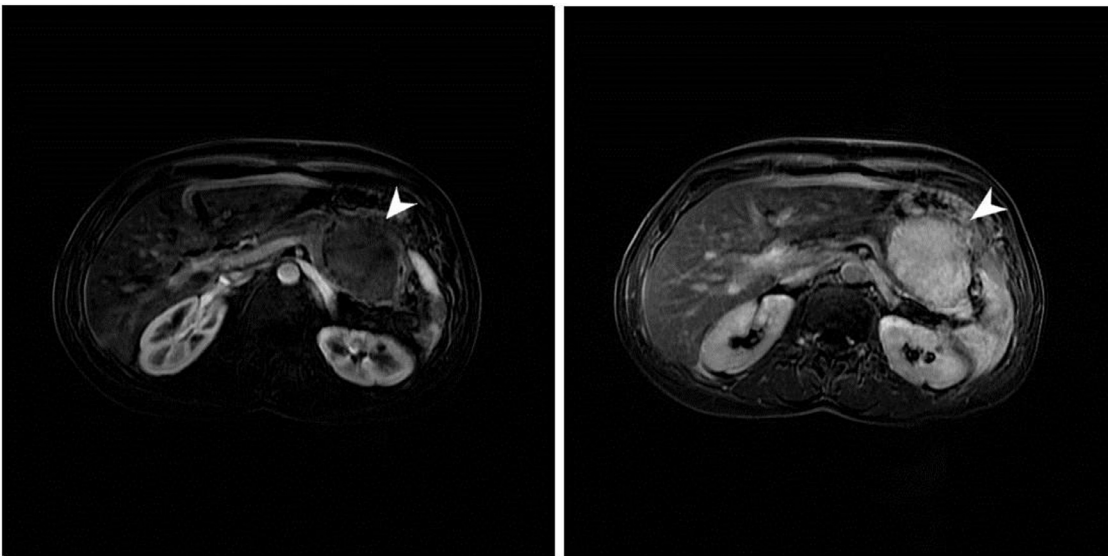
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**Fig. 1 – Axial T1-weighted (a) and fat-suppressed T2-weighted (b) magnetic resonance images.**

**A large mass is seen centered on the tail of the pancreas (arrowhead). This lesion was hyperintense with a hypointense nodular capsule on T2-weighted images (a) and heterogeneous, predominantly hypointense, on T1-weighted images (b).**



**Fig. 2 – Axial T1-weighted fat-suppressed subtraction magnetic resonance images after contrast administration during the arterial (a) and late (b) phases.**

**After gadolinium injection, the lesion (arrowhead) demonstrated progressive enhancement on the venous (a) and late phase (b), being maximal on the latest.**

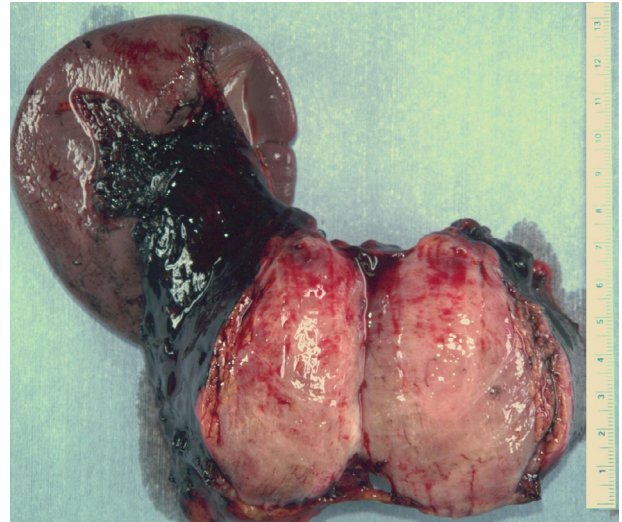
An octreotide radionuclide scan was recommended following the diagnostic hypotheses, which showed no uptake by the pancreatic mass or at a distant site, thus making the diagnosis of a neuroendocrine tumor less probable. A thoraco-abdominal computed tomography (CT) scan was performed to complete the staging. No signs of metastases were found. The tumor was described as a solid homogenous mass, slightly hypodense compared to the muscles, with no calcifi-

cations or cystic components (Fig. 3). After contrast administration, it showed a heterogeneous, predominantly hypovascular enhancement pattern, with an enhancing nodular capsule (Fig. 4a). The venous and late phases showed a progressive homogenous enhancement pattern (Fig. 4b). The mass was in close proximity with the greater curvature of the stomach, with no obvious signs of invasion.



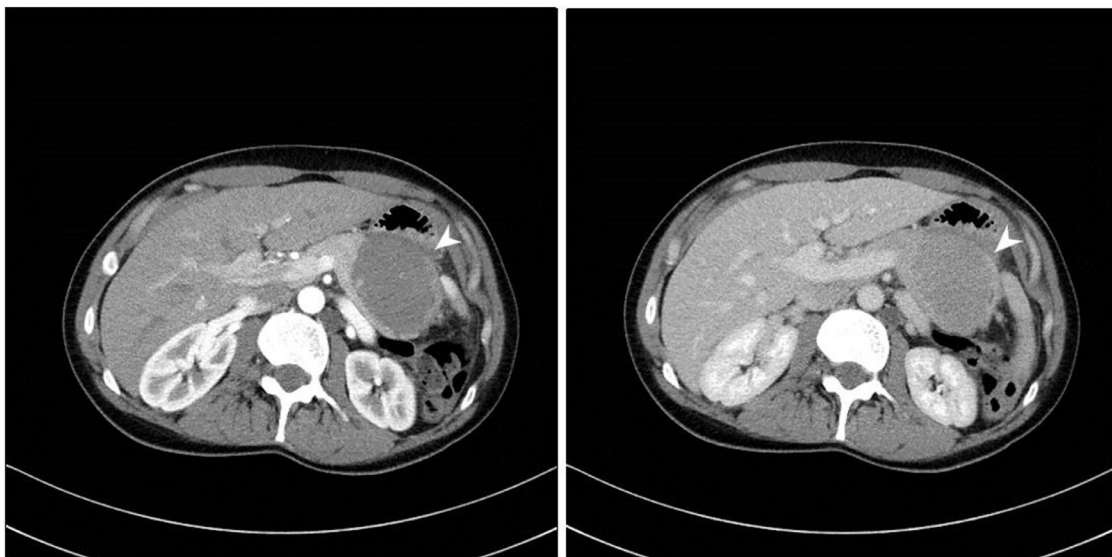
**Fig. 3 – Axial unenhanced computed tomography image.** A solid homogenous mass (arrowhead) is present at the tail of the pancreas. The lesion was slightly hypodense compared to the muscles, with no calcifications or cystic components.

Based on the age of the patient and the working hypotheses, the surgical team decided to proceed to a complete surgical excision of the tumor. The surgeon successfully resected the tumor following a distal pancreatectomy with splenectomy and a partial gastrectomy with negative surgical margins. Gross pathologic specimen showed a firm, white,

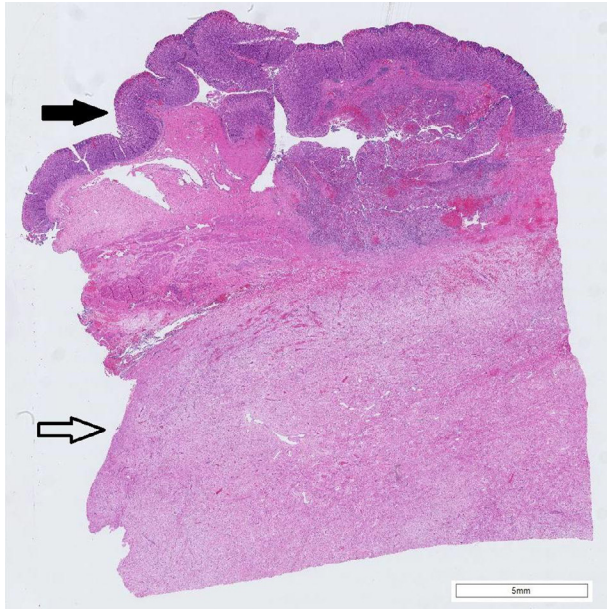


**Fig. 5 – Macroscopy.** Distal pancreatectomy with splenectomy specimen shows a firm, trabeculated 9.2 cm white mass in the pancreatic tail.

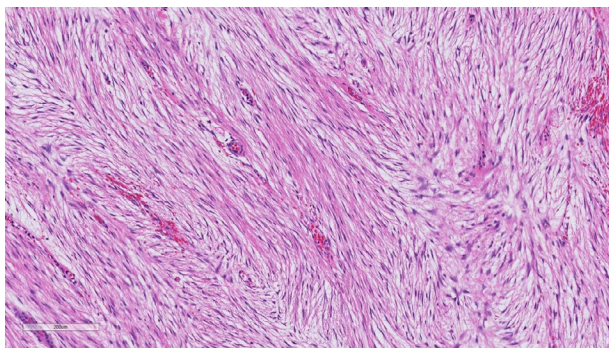
and trabeculated, 9.2 cm mass in the pancreatic tail (Fig. 5). Histologic examination revealed a low to moderately cellular tumor, composed of cytologically bland spindle-shaped cells in a collagenous stroma (Fig. 6), with infiltration of the stomach (Fig. 7). There were no mitoses or atypia. The tumor was consistent with a desmoid tumor. Tumor cells showed focal nuclear immunolabeling for  $\beta$ -catenin and Stat6 (which has been reported in some desmoid tumors) [1,2]. A follow-up with abdominal CT-scans every 6 months for 2 years, then yearly for a total of 5 years was recommended by the surgical team.



**Fig. 4 – Axial enhanced CT images during arterial (a) and venous (b) phases.** After contrast administration, the mass showed a heterogeneous, predominantly hypovascular enhancement pattern, with an enhancing nodular capsule (a). The venous and late phases showed a progressive homogenous enhancement pattern (b). The mass was in close proximity with the greater curvature of the stomach, with no signs of invasion.



**Fig. 6 – Low power view of the desmoid tumor (white arrow) infiltrating the wall of the stomach (black arrow) (photomicrograph, Hemotoxylin and Eosin stain, x 1).**



**Fig. 7 – Desmoid tumor. Diffuse proliferation of cytologically bland spindle cells in a collagenous stroma. Absence of mitosis or nuclear atypia. (photomicrograph, Hemotoxylin and Eosin stain, x 10).**

Considering the patient was not priorly known for a familial adenomatous polyposis, a colonoscopy was recommended to rule-out the diagnosis.

## 2. Discussion

Desmoid tumors are part of a group of disorders called fibromatoses. They are locally aggressive benign soft-tissue tumors [1]. They can occur at any age with a peak incidence in the third decade with a female predominance [1]. Intra-abdominal desmoid tumors arise primarily from mesenteric connective tissue or the retroperitoneum [3]. They are most often found in patients with complicated Gardner's syndrome or FAP [4].

They can also occur sporadically or as a result of trauma or surgery. The incidence of sporadic intra-abdominal desmoid tumors is very low (5%), of which only 10 cases of desmoid tumors of the pancreas have been published in the literature over the last 30 years [1, 5–7]. They have no known metastatic potential [8]. They tend to invade adjacent structures and be locally aggressive with a morbidity of 10% [6]. Patient tends to be asymptomatic or have symptoms related to the mass effect and invasion exerted by the tumor [6].

At pathology, they are homogeneous firm white masses that can have well-defined or infiltrative borders. They are composed of bland proliferative fibroblastic tissue with a collagen matrix. Differentiating desmoid tumors from other tumors is difficult at histology [8]. Immunohistochemistry helps to differentiate them from other fibroblastic and myoblastic tumors (with positive nuclear  $\beta$ -catenin immunostaining) [8].

At imagery, their appearance depends on the composition regarding the relative amounts of fibroblast proliferation, fibrosis, collagen, and vascularity of the tumor [9]. At CT, they may appear as well-defined or ill-defined masses with variable attenuation [9]. At magnetic resonance, they demonstrate low signal intensity compared to muscle on T1-weighted images [9]. T2 signal is variable (some desmoid tumors have been reported to be cystic) [8,9]. Enhancement is variable after contrast administration on CT and magnetic resonance imaging [9].

Surgical resection with clear margins is the mainstay of treatment. The likelihood of recurrence is high (19%-77%) [7].

Considering the rarity of this entity and the overlapping appearance at imagery with other benign primary nonepithelial neoplasms of the pancreas, it is important to recognize the essential role of pathology, particularly immunohistochemistry, in identifying this tumor. The high rate of postsurgical recurrence should prompt repeated follow-ups considering the potential aggressive nature of desmoid tumors.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2018.04.025](https://doi.org/10.1016/j.radcr.2018.04.025).

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