

IMAGE | PANCREAS

Solid-Cystic Pancreatic Tail Desmoid Tumor with Beta-Catenin Positivity

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CASE REPORT

A 19-year-old Hispanic woman with a history of polycystic ovarian syndrome presented with worsening abdominal pain, nausea, and vomiting. She had stable vitals and lower/suprapubic abdominal pain on exam, as well as a positive urinalysis. She was initially admitted for treatment of a urinary tract infection. An abdominal computed tomography scan, performed due to persistent abdominal pain, revealed a cystic lesion anterior to the pancreatic tail adjoining the splenic flexure and colonic diverticulosis, which was concerning for phlegmon formation (Figure 1). Magnetic resonance imaging showed a 4.1-cm pancreatic tail soft tissue lesion with a 3.4-cm unilocular nonenhancing cystic lesion. Carcinoembryonic antigen and CA19-9 antigen levels were normal. The patient underwent endoscopic ultrasound with fine-needle aspiration of the pancreatic lesion, and histopathology showed uniform spindle-cell proliferation (Figure 2). A colonoscopy, performed to exclude colonic involvement, had unremarkable gross findings. Laparoscopic distal pancreatectomy, splenectomy, partial gastrectomy, and partial segmental colectomy were performed. Gross examination revealed an infiltrative poorly circumscribed 4-cm mass

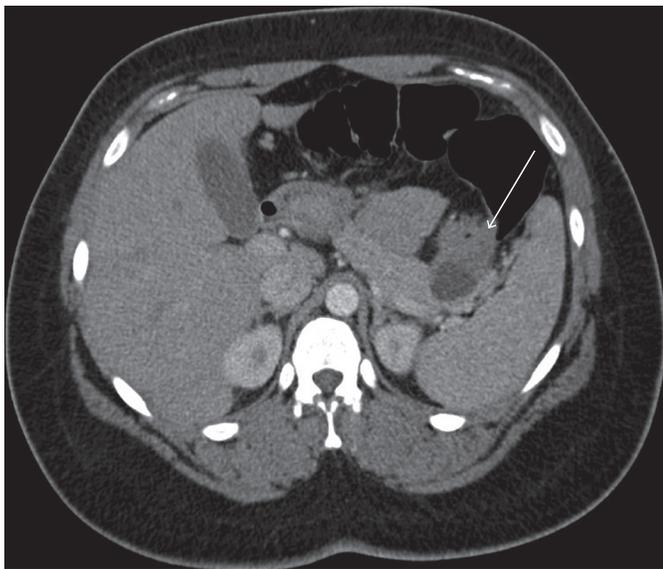


Figure 1. Abdominal computed tomography scan of a cystic lesion anterior to the pancreatic tail adjoining the splenic flexure and colonic diverticulosis.

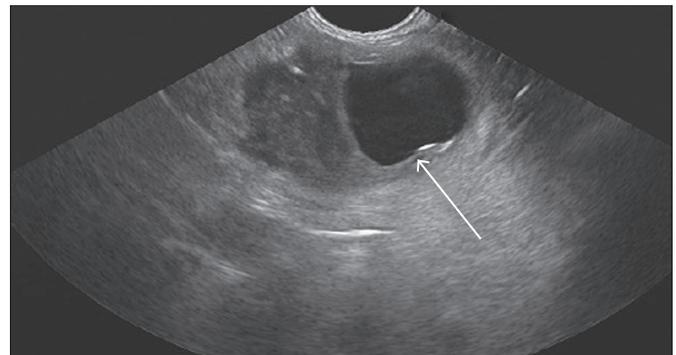


Figure 2. Endoscopic ultrasound showing a solid-cystic pancreatic mass.

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Figure 3. Gross image of an infiltrative, poorly circumscribed 4-cm mass with a unilocular cystic area measuring 2.5 cm.

with a 2.5-cm unilocular cystic area (Figure 3). Immunohistological evaluation led to a diagnosis of desmoid tumor primarily involving the pancreas, with cells showing nuclear positivity to beta-catenin. The patient recovered well postoperatively and was discharged with follow-up with a genetic counselor.

Desmoid tumors make up 0.03% of all neoplasms with an incidence of 2–3 per million population each year, arising largely sporadically, with 5–15% linked to familial adenomatous polyposis (FAP).^{1,2} With no potential for metastasis or dedifferentiation, desmoid tumors can cause complications through compression or obstruction of surrounding structures. FAP-associated tumors occur primarily intraabdominally, while non-FAP associated tumors occur extraabdominally and on the pelvic/abdominal wall.³ Immunohistochemical testing is required to establish a definitive diagnosis, with nuclear beta-catenin staining supporting the diagnosis with 80% and 67% positivity in sporadic and FAP-associated cases, respectively.⁴ Pancreatic desmoid tumors are particularly rare and require pathological evaluation for differentiation from other

pancreatic lesions. In a literature review, there have been 12 reported cases of such lesions, primarily occurring sporadically and within the pancreatic tail.

The generally indolent nature of these tumors allows treatment to be dictated by location, risk to surrounding structures, and the extent of infiltration. With extraabdominal or abdominal wall tumors, surgical resection with negative margins is the treatment of choice. Surgical intervention becomes more complex with intraabdominal tumors, as does the inherent risk of recurrence. While the decision was made for surgical resection in this case given concern for multi-organ infiltration, further exploration on the role of multimodal treatment involving radiation and systemic therapy is needed.

DISCLOSURES

Author contributions: HD Patel wrote the manuscript and searched the literature. NR Desai wrote the manuscript and obtained the images. A. Som searched the literature. SK Shah reviewed the manuscript. NC Thosani wrote and edited the manuscript and is the article guarantor.

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