



Giant pancreatic solid cystic desmoid tumor with two ectopic adrenal tissues

*Neşe Ekinci¹, *Fikret Dirilenoğlu¹, Arzu Avcı¹, Oğuzhan Özsay²

¹Department of Pathology, İzmir Katip Çelebi University Atatürk Training and Research Hospital, İzmir, Turkey

²Department of General Surgery, İzmir Katip Çelebi University Atatürk Training and Research Hospital, İzmir, Turkey

Cite this article as: Ekinci N, Dirilenoğlu F, Avcı A, Özsay O. Giant pancreatic solid cystic desmoid tumor with two ectopic adrenal tissues. Turk J Gastroenterol 2017; 28: 401-4.

ABSTRACT

A 19-year-old woman presented with painless swelling of the abdomen. During surgery, a giant mass measuring 37 cm×26 cm×12 cm within the distal pancreas invading the spleen was noted. The clinical diagnosis of a solid cystic pseudopapillary tumor of the pancreas was suspected. Distal pancreatectomy, splenectomy, and debulking surgery were performed. Histological examination showed that the tumor infiltrated the spleen and pancreatic parenchyma, and sections of the solid areas revealed a proliferation of spindle-shaped or stellate cells growing in fascicular and storiform patterns within a myxoid intercellular matrix. Cystic areas were representing the entrapped excretory pancreatic ductules. Interestingly, there were two ectopic adrenal tissues found incidentally in the peripheral portion of the tumor. The histopathologic and immunohistochemical features were consistent with a solid cystic desmoid tumor of the pancreas. Desmoid tumors of the pancreas are very rare, and if they present as a solid cystic lesion, their diagnosis may be difficult. We report the case for its rarity and huge size and to emphasize a regular follow-up because the long-term prognosis is currently unknown.

Keywords: Desmoid tumor, pancreas, solid cystic, ectopic adrenal, aggressive fibromatosis

INTRODUCTION

Desmoid tumors (DTs) are a rare type of benign soft-tissue tumors, and intraabdominal forms are associated with familial adenomatous polyposis (FAP) and Gardner syndrome. They may also occur sporadically or subsequent to a surgical or nonsurgical trauma. They are characterized by dense fibroblastic proliferation in an abundant collagenous extra-cellular matrix (1,2). Although they have no metastatic potential, they are locally aggressive with a high recurrence rate. DTs are usually asymptomatic for a long period of time. Diagnosis can be made if symptoms or mechanical complications develop.

Desmoid tumors occur very rarely in the pancreas and only a few cases have been reported in the literature since 1956. Here we report a sporadic giant solid cystic DT of the pancreas in a 19-year-old woman presenting with an abdominal mass.

CASE PRESENTATION

A 19-year-old woman presented with complaints of painless swelling of the abdomen. There was no history of weight loss, jaundice, vomiting, or fever, and no family history of a genetic disease. Abdominal computed tomography delineated a mass measuring 37 cm×25 cm in the left side of the abdomen with cystic and solid areas and septations, as well as locally invading the spleen and porta hepatis. During surgery, a huge mass of 37 cm×25 cm×12 cm within the distal pancreas invading the spleen and the adjacent portion of the diaphragm was noted. The clinical diagnosis of a solid cystic pseudopapillary tumor of the pancreas was suspected. Distal pancreatectomy, splenectomy, and debulking surgery were performed. An informed consent was obtained from the patient before the surgery and for this publication.

Gross examination revealed a 16 cm×12 cm×3.5 cm spleen, a 14 cm×8 cm×1 cm mesentery, and a 6 cm×4.5

This study was presented as a poster at the 27th European Congress of Pathology, 9 September 2016, Belgrade, Serbia.

*These authors have contributed equally to this work.

Address for Correspondence: Fikret Dirilenoğlu E-mail: fikretdirilenoglu@gmail.com

Received: May 8, 2017

Accepted: May 16, 2017

Available Online Date: August 9, 2017

© Copyright 2017 by The Turkish Society of Gastroenterology • Available online at www.turkjgastroenterol.org • DOI: 10.5152/tjg.2017.17208



Figure 1. Gross appearance of the giant tumor with the spleen on the right side

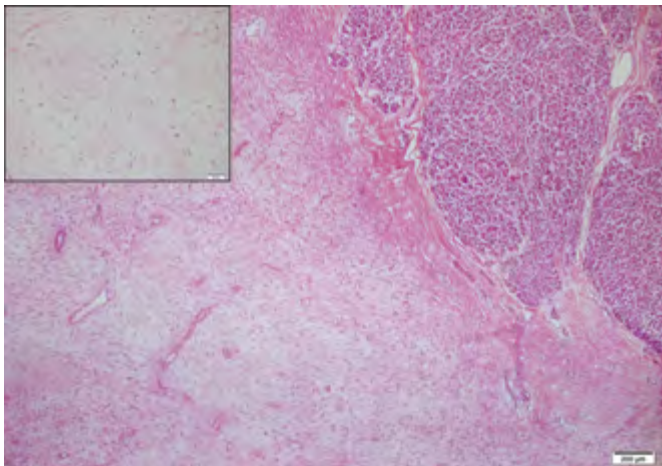


Figure 2. Pancreatic infiltration by the tumor, H&E 200x; inlet, hypocellular areas, H&E 40x

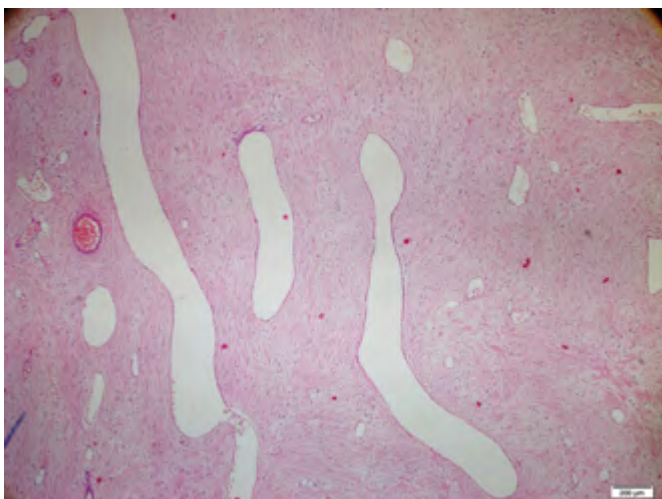


Figure 3. Dilated vascular channels, H&E 200x

cm \times 1.5 cm pancreas with a huge, partially well circumscribed, non-encapsulated tumor measuring 37 cm \times 26 cm \times 12 cm (Figure 1). It was invasive to the hilus of the spleen and surrounding fat tissue, and the cut surface of the tumor showed several multicystic cavities and solid-myxoid areas. Histological analysis showed a tumor arising from the pancreas, and sections of the solid areas revealed a proliferation of spindle-shaped or stellate cells growing in fascicular and storiform patterns within a myxoid intercellular matrix (Figure 2,3). Glassy, hyalinized, and keloid-like collagen fibers were seen in the peripheral portions. Spindle-shaped cells had regular nuclei, and their mitotic activity was very low (only 2 per 50 high-power fields). Tumor necrosis was not seen. Cystic areas within the solid component were made up of retentional cysts and entrapped excretory pancreatic ductules with dilation (Figure 4). Interestingly, there were two ectopic adrenal tissues found incidentally, measuring 2 mm each, which were located within the peripheral portion of the tumor (Figure 5).

Immunohistochemical analysis showed nuclear positivity for β -catenin, while estrogen receptor was negative (Figure 6). The Ki-67 proliferation index was <1%. Entrapped dilated excretory pancreatic ductules were highlighted with keratins and EMA among neoplastic cells. Other markers used in differential diagnosis, such as CD117, DOG-1, CD34, desmin, smooth muscle actin, and S100, were all negative. The histopathologic and immunohistochemical features were consistent with a solid cystic DT of the pancreas.

DISCUSSION

Desmoid tumors comprise 3% of soft-tissue tumors, but intraabdominal forms are more rarely seen (3). The incidence of sporadic intraabdominal DT is very low (5%), and the ones originating from the pancreas are the rarest with only 20 cases having been described in the English literature since the 1980s (4,5). While abdominal DTs are associated with FAP and Gardner syndrome, so far only one case had this association (4). There was no history of any syndromes or genetic abnormalities in our case.

Although the etiology is not known, previous trauma, surgery, pregnancy, hormonal factors, and irradiation are among the risk factors. In our case, there were no known risk factors. While female patients are at risk for intraabdominal DTs, there is a slight male predominance in pancreatic DTs (M/F=11/10) (6).

Symptoms depend on the location and occur with local invasion to adjacent tissues (7). The patients with intraabdominal DT generally have no symptoms and are encountered incidentally during laparotomy. The most common symptom is epigastric pain, while weight loss, nausea, abdominal fullness, or painless jaundice may be seen. Our patient presented with painless abdominal swelling.

Somatic mutation of the Wnt/ β -catenin gene and abnormal myofibroblastic proliferation are observed in DTs. This mutation

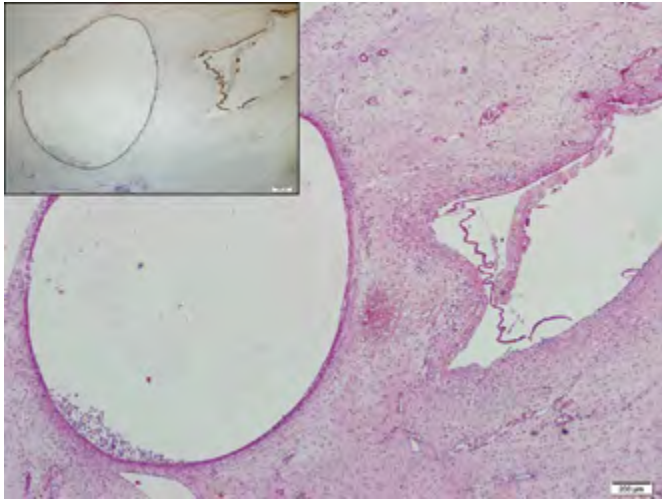


Figure 4. Dilated entrapped excretory pancreatic ducts, H&E 200x; inlet, cytokeratin 7 positivity, 200x

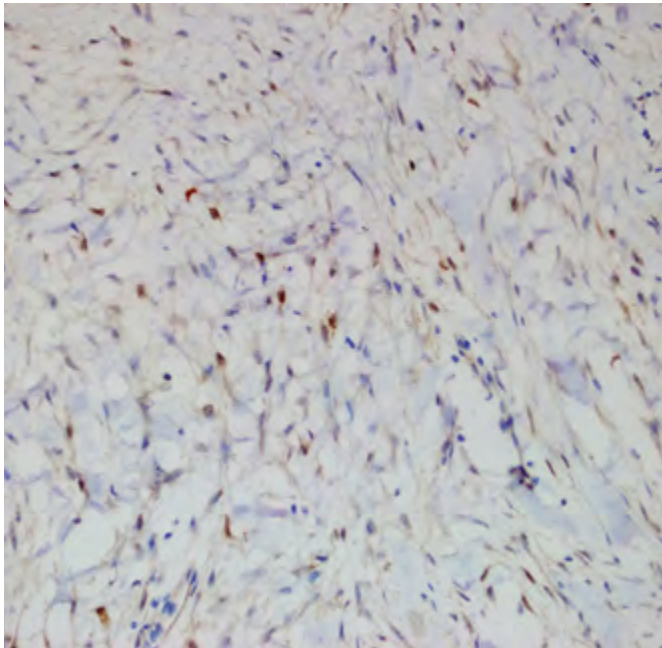


Figure 5. β -catenin nuclear positivity, 40x

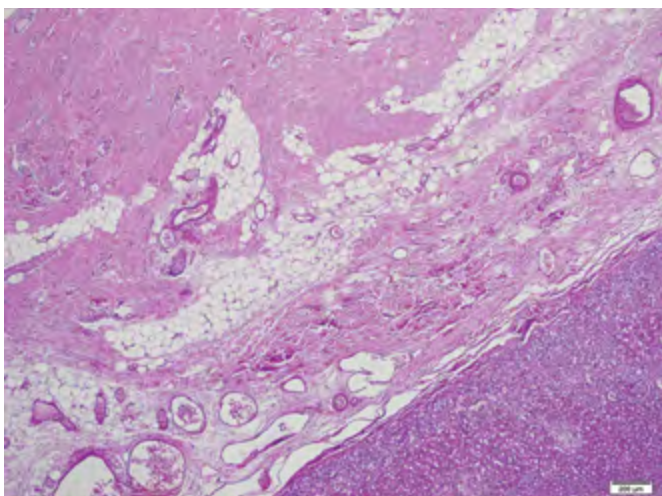


Figure 6. Ectopic adrenal tissue at the tumor periphery, H&E 200x

causes the accumulation of β -catenin in the nucleus. It is helpful to show intranuclear β -catenin by immunohistochemistry in the differential diagnosis from other benign and malignant proliferations (8).

Preoperative diagnosis of sporadic intraabdominal DT is not usual due to the asymptomatic growth of the tumor. Because of local invasion and mechanical complications in the long-term, surgery is needed (3). Radiologic studies such as ultrasonography (US), computerized tomography, and magnetic resonance imaging are essential to determine tumor characteristics, invasion, and resectability. Endoscopic US helps image the tumors localized within the pancreatic head and provides examination of the superficial tumors with fine needle aspiration (9).

If clear margins are obtained, radical surgery is curative for pancreatic DT (10). Most of these patients so far had distal pancreatectomy or pancreaticoduodenectomy, while a biopsy was done only in four cases (3). In our case, due to local invasion of the tumor, distal pancreatectomy, splenectomy, and debulking surgery were performed. Other effective treatment choices used in DTs are NSAIDs, chemotherapy, and radiotherapy (11). No other treatments were used in our case besides surgery. After 27 months follow-up, no recurrence has been observed.

Differential diagnosis includes other benign or malignant mesenchymal lesions, such as extra-gastrointestinal stromal tumor, solitary fibrous tumor, dedifferentiated liposarcoma, schwannoma, leiomyoma, leiomyosarcoma, pancreatic mucinous cystic tumor, and solid pseudopapillary neoplasm (12-14). In problematic cases, immunohistochemistry and molecular tests would be helpful in diagnosis.

In summary, intraabdominal DTs may reach extreme sizes without demonstrating noticeable symptoms. In cases with large, isolated intraabdominal mass with solid and cystic components originating from the pancreas, DT should be kept in mind.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author contributions: Concept - N.E., F.D.; Design - N.E., F.D.; Supervision - N.E., A.A.; Materials - O.O.; Data Collection and/or Processing - F.D., O.O.; Analysis and/or Interpretation - N.E., A.A., F.D.; Literature Search - F.D., N.E.; Writing - N.E., A.A., F.D.; Critical Reviews - N.E., A.A., F.D., O.O.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Polistina F, Costantin G, D'Amore E, Ambrosino G. Sporadic, non-trauma-related, desmoid tumor of the pancreas: a rare disease-case report and literature review. *Case Rep Med* 2010; 2010: 272760. [\[CrossRef\]](#)
2. Amiot A, Dokmak S, Sauvanet A, et al. Sporadic desmoid tumor. An exceptional cause of cystic pancreatic lesion. *JOP* 2008; 9: 339-45.
3. Clark SK, Phillips RKS. Desmoids in familial adenomatous polyposis. *Br J Surg* 1996; 83: 1494-504. [\[CrossRef\]](#)
4. Tsukamotoa Y, Imakita M, Nishitani A, Ito T, Izukurac M, Hirotaa S. Pancreatic desmoid-type fibromatosis with beta-catenin gene mutation-Report of a case and review of the literature. *Pathol Res Pract* 2016; 212: 484-9. [\[CrossRef\]](#)
5. Patel HD, Desai NR, Som A, Shah SK, Thosani NC. Solid-cystic pancreatic tail desmoid tumor with beta-catenin positivity. *ACG Case Rep J* 2017; 4: e40. [\[CrossRef\]](#)
6. Lazar AJ, Hajibashi S, Lev D. Desmoid tumor: from surgical extirpation to molecular dissection. *Curr Opin Oncol* 2009; 21: 352-9. [\[CrossRef\]](#)
7. Roggli VL, Kim HS, Hawkins E. Congenital generalized fibromatosis with visceral involvement. A case report. *Cancer* 1980; 45: 954-60. [\[CrossRef\]](#)
8. Bhattacharya B, Dilworth HP, Iacobuzio-Donahue C, et al. Nuclear beta-catenin expression distinguishes deep fibromatosis from other benign and malignant fibroblastic and myofibroblastic lesions. *Am J Surg Pathol* 2005; 29: 653-9. [\[CrossRef\]](#)
9. Lips DJ, Barker N, Clevers H, Hennipman A. The role of APC and beta-catenin in the aetiology of aggressive fibromatosis (desmoid tumors). *Eur J Surg Oncol* 2009; 35: 3-10. [\[CrossRef\]](#)
10. Ballo MT, Zagars GK, Pollack A, Pisters PW, Pollack RA. Desmoid tumor: prognostic factors and outcome after surgery, radiation therapy, or combined surgery and radiation therapy. *J Clin Oncol* 1999; 17: 158-67. [\[CrossRef\]](#)
11. Tanaka K, Yoshikawa R, Yanagi H, et al. Regression of sporadic intra-abdominal desmoid tumour following administration of non-steroidal anti-inflammatory drug. *World J Surg Oncol* 2008; 6: 17. [\[CrossRef\]](#)
12. Shimada S, Ishizawa T, Ishizawa K, Matsumura T, Hasegawa T, Hirose T. The value of MDM2 and CDK4 amplification levels using real-time polymerase chain reaction for the differential diagnosis of liposarcomas and their histologic mimickers. *Hum Pathol* 2006; 37: 1123-9. [\[CrossRef\]](#)
13. Miettinen M, Lasota J. Gastrointestinal stromal tumors: review on morphology, molecular pathology, prognosis, and differential diagnosis. *Arch Pathol Lab Med* 2006; 130: 1466-78.
14. Xu B, Zhu LH, Wu JG, Wang XF, Matro E, Ni JJ. Pancreatic solid cystic desmoid tumor: case report and literature review. *World J Gastroenterol*. 2013; 19: 8793-8. [\[CrossRef\]](#)