Case Report

J Korean Soc Radiol 2021;82(5):1297-1303
https://doi.org/10.3348/jksr.2020.0163
eISSN 2288-2928

대한영상의학회지

Pancreatic Collision Tumor of Desmoid-Type Fibromatosis and Mucinous Cystic Neoplasm: A Case Report

데스모이드 섬유종증과 점액성 낭성 종양으로 이루어진 췌장의 충돌 종양: 증례 보고

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Pancreatic collision tumors are rare neoplasm, and cases consisting of ductal adenocarcinoma with a neuroendocrine tumor, intraductal papillary mucinous neoplasm with a neuroendocrine tumor, and solid pseudopapillary neoplasm with a neuroendocrine tumor have been reported. We report a case of a rapidly growing pancreatic collision tumor consisting of desmoid-type fibromatosis and mucinous cystic neoplasm in a 30-year-old pregnant female. To the best of our knowledge, this is the first reported case of a pancreatic collision tumor consisting of desmoid-type fibromatosis and mucinous cystic neoplasm.

Index terms Tumor; Fibromatosis; Neoplasm; Pancreas; Pregnancy

Received September 9, 2020
Revised October 5, 2020
Accepted December 17, 2020

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INTRODUCTION

Collision tumors are rare tumors characterized by the coexistence of two adjacent but histologically different neoplasms arising in the same organ (1).

Desmoid-type fibromatoses (DFs) are rare mesenchymal tumor which can originate in anywhere in the body. Pancreatic mucinous cystic neoplasms (MCNs) are rare epithelial tumor and almost all cases occur in female. DFs and pancreatic MCNs usually grow slowly (2, 3). However, their growth is relatively rapid in pregnancy, which might be attributed to hormonal effects (4, 5).

To the best of our knowledge, there have been no reported cases of pancreatic collision tumors composed of DF and MCN.

Here, we report the first case of a pancreatic collision tumor of desmoid-type fibromatosis and MCN in a 30-year-old pregnant female.

CASE REPORT

A 30-year-old female presented to our institution with complaints of epigastric discomfort and vomiting for 1 month. Physical examination revealed epigastric tenderness. She had no known medical history, including operation. One month before, laboratory test results had been within the normal ranges. Gastroscopy had revealed diffuse mucosal bulging in the distal portion of the stomach. Suspecting extrinsic compression of the stomach, her physician had recommended abdominal CT to determine the cause, but she had declined and was discharged. She arrived at our emergency center after referral from another institution because of aggravation of symptoms. At the local institution, she denied the possibility of pregnancy and undergone abdominal CT. The outside abdominal CT, including a pre-enhanced phase and a phase starting 2 minutes after contrast injection, revealed a lobulated mass located between the pancreatic tail and the left colon, measuring 22.5 cm × 19.3 cm × 17.5 cm. The mass appeared to be predominantly solid, with a peripheral cystic portion. The solid and cystic portions of the mass measured 14.5 cm and 7.2 cm in the longest diameter, respectively. The solid portion of the mass was homogeneous and had lower attenuation [27–34 Hounsfield unit (HU)] than the skeletal muscle on pre-enhanced CT. On contrast-enhanced CT, the solid portion showed mild homogeneous enhancement (46–53 HU), while the cystic portion showed enhancing thin wall (Fig. 1A). CT also revealed intra-uterine pregnancy, which the patient was unaware of. Obstetric evaluation confirmed a 10-week intrauterine pregnancy. She wanted to continue the pregnancy and was admitted for further evaluation.

We evaluated chest CT performed 7 months before her admission. The mass had mixed solid and cystic components, measuring 3.7 cm × 3.5 cm × 3.3 cm in size, between the pancreatic tail and the left colon (Fig. 1B). The solid and cystic portions of the mass measured 3.2 cm and 2.2 cm in the longest diameter, respectively. It was clear that the mass existed 18 weeks before the pregnancy and had grown from 3.7 cm to 22.5 cm in the longest diameter in 7 months.

To obtain additional information regarding the mass, dynamic MRI was performed. On abdominal MRI, the solid portion of the mass appeared hyperintense to skeletal muscles on T2-weighted images (T2WIs) and hypointense on T1-weighted images (T1WIs). The solid portion showed diffusion restriction on diffusion-weighted imaging/apparent diffusion coefficient map ($b = 800$ s/mm^2) (Fig. 1C). On dynamic contrast-enhanced MRI, the mass showed homogeneous and progressive enhancement. Signal intensity of the cystic portion of the mass followed that of the spinal cerebrospinal fluid on all MRI sequences (Fig. 1D). The mass appeared to originate from the pancreatic tail, but the origin was indistinguishable between the pancreas and the retroperitoneum as the fat plane between the mass and the adjacent colon was lost. The cystic portion of the mass was thought to be necrotic tissue of the tumor.

The mass was rapidly growing, solid, with possible necrosis and local invasion, suggesting malignancy but specific diagnosis were not confidently made based on imaging features.

Surgical resection was planned based on the uncertainty of malignant potential and aggravation of the patient's symptoms. Distal pancreatectomy with wedge resection of the colon was performed.

On surgery, the gross appearance of the mass was pinkish, firm, with mixed solid and cys-

Fig. 1. Pancreatic collision tumor of desmoid-type fibromatosis and mucinous cystic neoplasm in a 30-year-old pregnant female.
A. Axial pre-enhanced abdominal CT reveals a mass, with mixed solid and cystic components, located in the pancreatic tail. The solid portion of the mass shows homogeneously lower attenuation than that of skeletal muscles. Axial contrast-enhanced abdominal CT, 2 minutes after contrast injection reveals that the solid portion of the mass shows homogeneous enhancement and the cystic portion shows thin walls.
B. Axial chest CT, 7 months prior to admission reveals a smaller mass at the same location, with mixed solid and cystic components (arrow).
C. Axial T2WIs/T1WIs reveals a mass, with mixed solid and cystic components, located in the pancreatic tail. The solid portion of the mass appears hyperintense compared to skeletal muscles on a T2WI and hypointense on a T1WI. Axial DWI/ADC map reveals diffusion restriction of the solid component.
ADC = apparent diffusion coefficient, DWI = diffusion-weighted imaging, T1WI = T1-weighted image, T2WI = T2-weighted image

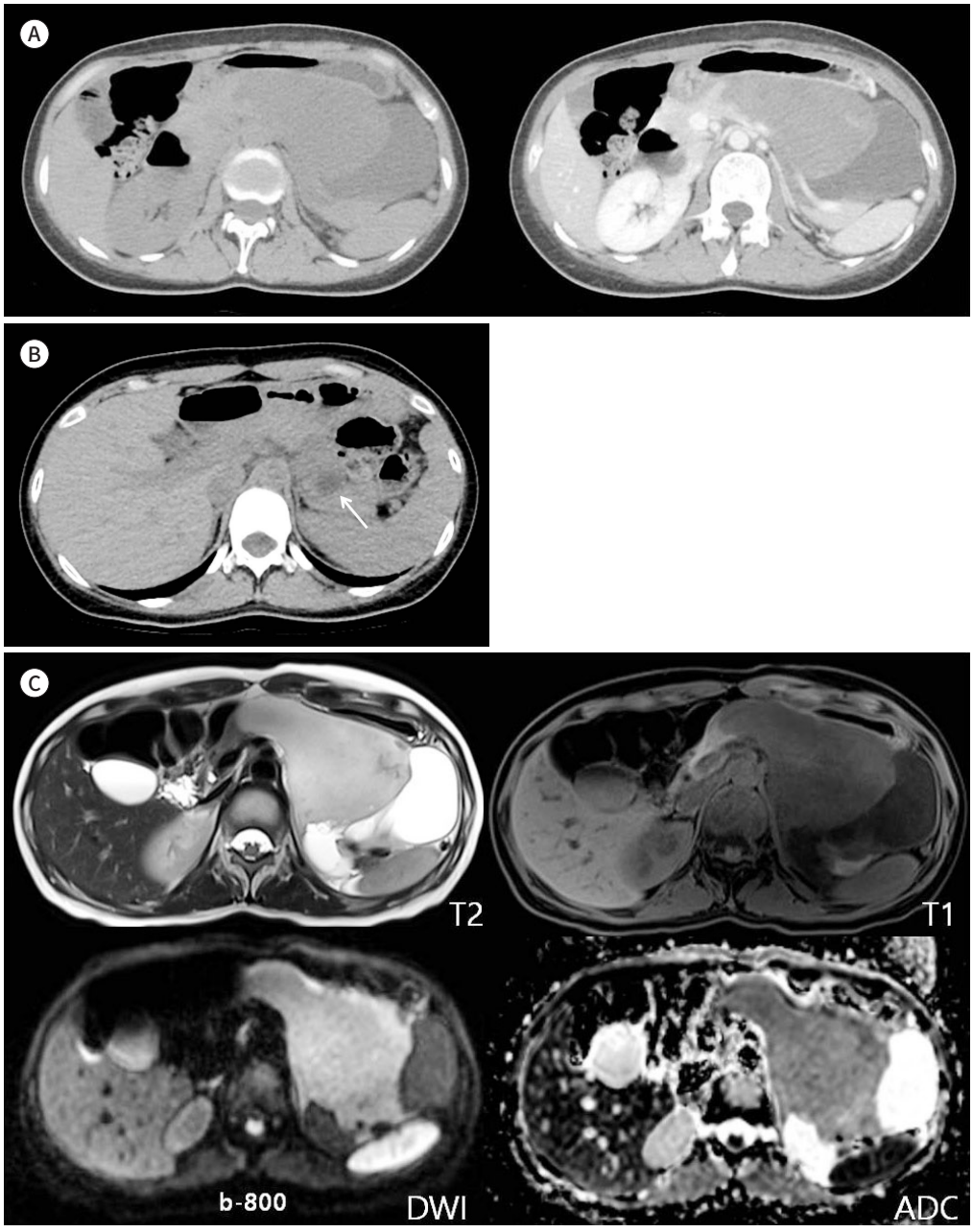
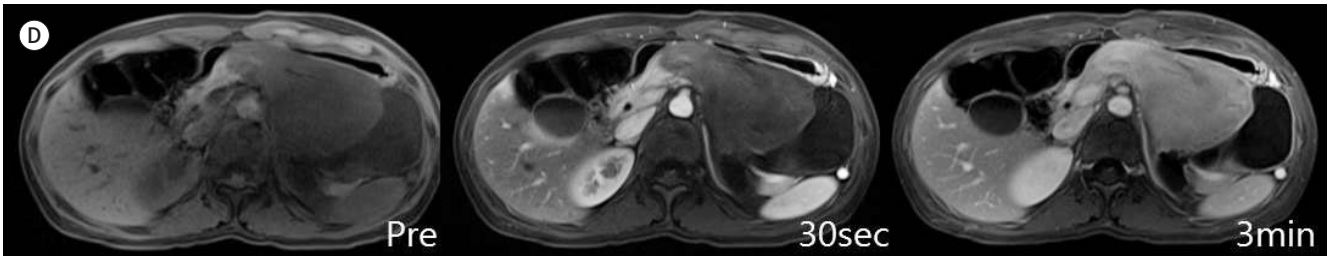
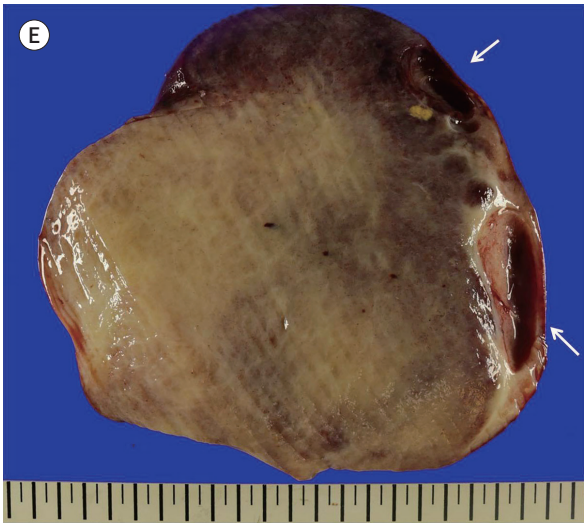
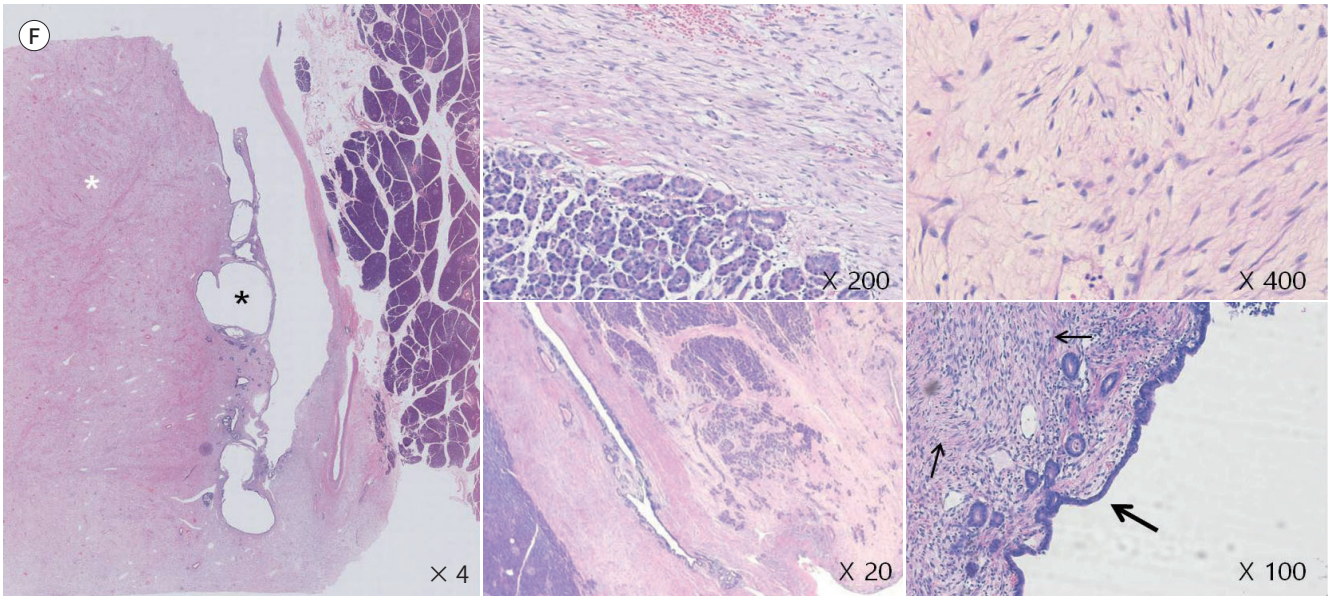


Fig. 1. Pancreatic collision tumor of desmoid-type fibromatosis and mucinous cystic neoplasm in a 30-year-old pregnant female.
D. Axial pre-enhanced and dynamic enhanced MRI performed 30 seconds and 3 minutes after contrast injection, respectively, reveal homogeneous and progressive enhancements of the solid component, respectively.
E. The gross specimen shows two distinct components, a large grayish white-to-brown rubbery solid area and a peripheral cystic area (arrows).
F. Haematoxylin and eosin staining shows desmoid-type fibromatosis (white asterisk) and mucinous cystic neoplasm (black asterisk) presenting as a collision tumor in the pancreas. Photomicrograph reveals that the solid tumor exhibits cytologically bland, uniform fibroblastic/myofibroblastic spindle-shaped cells arranged in mainly short fascicles in a myxoid collagenous matrix. There is no significant atypia or pleomorphism, and mitoses are rare. The cystic tumor shows multiloculated cysts lined by mucinous epithelium (thick arrow). There are focal areas of intermediate-grade dysplasia in the ovarian-like stroma (thin arrows).







tic components. The mass was located at the pancreatic tail, adhered to the colon (Fig. 1E), and measured 20 cm in the greatest diameter.

Pathology revealed two independent masses in the pancreas: one solid and the other cystic. The solid mass, measuring 19.5 cm × 17.2 cm × 15.5 cm, was homogeneous, myxoidal, and invaded the colon. It was composed of cytologically uniform fibroblastic/myofibroblastic cells arranged in mainly short fascicles in an abundant myxoid matrix. Hemorrhage and necrosis were absent. The solid mass was diagnosed to be pancreatic DF. The other mass, thought to be a cystic portion of the mass, was lined by mucinous epithelium with ovarian-like stroma, which is consistent with pancreatic MCN with intermediate-grade dysplasia (Fig. 1F). Finally, a diagnosis of pancreatic collision tumor of DF and MCN was made.

DISCUSSION

Collision tumors are rare neoplasms characterized by the coexistence of two adjacent but histologically distinct tumors in a single organ (1). Pancreatic collision tumors are rare. Reported cases of pancreatic collision tumors consisted of ductal adenocarcinoma with a neuroendocrine tumor, intraductal papillary mucinous neoplasm with a neuroendocrine tumor, and solid pseudopapillary neoplasm with a neuroendocrine tumor (1, 6, 7).

DF, also known as aggressive fibromatosis, is a rare mesenchymal tumor, histologically characterized by fibroblastic proliferation with intercellular collagen. It has a tendency of local invasion and recurrence but without metastasis. It can originate anywhere in the body (2). Histologically, it consists of uniform fibroblasts, fascicles, and a dense collagenous stroma and may present with myxoid changes, keloid fiber formation, muscular hyperplasia of the small arteries, dilated thin-walled veins, or microhemorrhages (4).

Imaging features of DF are variable, reflecting the varying proportion of the histologic components: spindle cells, myxoid matrix, and surrounding collagenous stroma. On CT, DF appears as a soft tissue mass with sharp or ill-defined margins. On MRI, it shows variable signal intensity and can be isoattenuated to hyperattenuated compared to skeletal muscles with hyperattenuation and hypoattenuation, likely reflecting the collagenous and myxoid components, respectively. DF shows variable enhancement, with most cases showing mild-to-moderate enhancement. The signal intensity of DF reflects the proportion of histologic components. The most common MRI finding of DF is a heterogeneous tumor showing signal intensity similar to or higher than that of skeletal muscles on T2WIs and similar to skeletal muscles on T1WIs. Low signal intensity on T2WIs reflects dense collagen and hypocellularity; in contrast, high signal intensity results from a high proportion of spindle cells. Most cases of DF demonstrate moderate-to-marked enhancement after injection of a gadolinium-based contrast agent. Enhancement is particularly marked in areas of high cellular and less fibrotic components. Although DF may show areas without enhancement, necrosis is very rare (8).

Pancreatic MCNs are rare tumors lined by mucin-producing columnar epithelium with underlying ovarian-type stroma. Almost all cases of MCN occur in female in the fifth to seventh decades of life, and they are extremely rare in pregnant female. They typically arise in the pancreatic body or tail and usually do not communicate with the pancreatic duct (3).

On CT, pancreatic MCNs appear as unilocular or septated hypoattenuated cystic lesions.

They also appear slightly hyperattenuated because of the presence of mucin or hemorrhage. They often demonstrate thick wall presenting as delayed enhancement (9).

On MRI, pancreatic MCNs appear as unilocular or septated cystic lesions. Although MCNs are typically mucin-filled cystic tumors, the signal intensity of the intratumoral fluid follows that of simple fluid (9).

DFs and pancreatic MCNs are usually slow-growing tumors (2, 3). However, their growth may be relatively rapid in pregnancy, as in our case, which is attributable to hormonal factors. The possibility of an association between hormonal changes during pregnancy and the development of DF is suggested by reports on spontaneous regression of DF after delivery and DF responding to an anti-estrogen agents (4). Moreover, it has been reported that the rapid growth of pancreatic MCNs during pregnancy may be attributable to its responsiveness to sex hormones (5).

Differential diagnosis should include tumors which may appear as mixed solid and cystic masses, such as cystic pancreatic endocrine tumor, cystic change of ductal adenocarcinoma, and solid pseudopapillary tumor. Cystic pancreatic endocrine tumor tends to appear as unilocular cyst, predominantly occupying the tumor with enhancing solid portion. Cystic change of ductal show irregular margin with associated ductal dilatation. The solid component represent poor enhancement. Solid pseudopapillary tumor usually appear as well-marginated mass with fibrous capsule and may represent calcification or hemorrhage (10).

In our patient, DF and pancreatic MCN coexisted as a collision tumor in the pancreatic tail. The two tumors showed typical imaging features on both CT and MRI. The histological finding that the DF had abundant myxoid matrix was well reflected on MRI, considering the fact that most of the myxoid matrix had a high signal intensity on T2WIs. Moreover, the rapid growth of DF and MCN during pregnancy may be attributed to the sex hormones.

However, as they existed as a collision tumor and as pancreatic collision tumors of DF and MCN in pregnant female have not been reported before, it was difficult to make a diagnosis other than that of malignant tumors.

When encountered with solid and cystic mass in the pancreas, radiologist should aware of possibility of collision tumor as differential diagnosis.

Author Contributions

Conceptualization, K.J.W., C.J.H.; data curation, R.M.J., K.J.W.; formal analysis, R.M.J., K.J.W., C.J.H.; funding acquisition, K.J.W.; investigation, R.M.J., L.S.E.; methodology, R.M.J., K.J.W., L.S.E.; project administration, R.M.J., K.J.W., L.S.E.; resources, R.M.J., K.J.W., L.S.E.; software, R.M.J., K.J.W., L.S.E.; supervision, K.J.W.; validation, R.M.J., K.J.W., L.S.E.; visualization, R.M.J., K.J.W., C.J.H.; writing—original draft, R.M.J.; and writing—review & editing, K.J.W., L.S.E.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

None

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데스모이드 섬유종증과 점액성 낭성 종양으로 이루어진 췌장의 충돌 종양: 증례 보고

류민중[1] · 김재운[1*] · 이승은[1] · 최준혁[2]

췌장의 충돌 종양은 매우 드문 종양으로서 췌장선암종과 신경내분비 종양, 췌관내유두상 점액 종양과 신경내분비 종양, 그리고 췌장 고형성 가유두상 종양으로 이루어진 증례들이 보고된 바 있다. 우리는 30세 임신한 여성에서 빠르게 자란, 데스모이드 섬유종증과 점액성 낭성 종양으로 이루어진 췌장의 충돌 종양의 증례를 보고하고자 한다. 저자들이 아는 한, 섬유종증과 점액성 낭성 종양으로 이루어진 췌장의 충돌 종양을 최초로 보고하는 증례이다.

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