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Mucinous Cystic Neoplasm of the Pancreas With Extensive Decidualized Ovarian-Type Stroma During Pregnancy: A Case Report and Review of Literature

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Abstract

Mucinous cystic neoplasms (MCNs) of the pancreas are rare cyst-forming epithelial neoplasms that occur predominantly in women and are defined by ovarian-type stroma. MCNs arising during pregnancy are uncommon and present diagnostic and management challenges because of interval growth, uncertain malignant potential, and the need to balance maternal and fetal safety. The overt decidualization of ovarian-type stroma in pancreatic MCN appears to be exceptionally rare.

We report a 20-30-year-old pregnant woman with a large pancreatic tail cystic lesion identified before conception that enlarged during gestation. Pre-pregnancy imaging measured the lesion at 9.2-10.6 cm, and follow-up magnetic resonance cholangiopancreatography (MRCP) at 16 weeks showed growth to 12.5 cm without definite pancreatic duct communication or a solid nodular component. Given interval enlargement and uncertain malignant potential, she underwent open distal pancreatectomy with splenectomy because of dense adhesions and tumor size. Gross examination revealed a 14.0 cm cystic lesion. Histology showed MCN with low-grade dysplasia and extensive decidualized ovarian-type stroma. Immunohistochemistry supported findings of ovarian-type stroma. We performed a literature review of previously reported pregnancy-associated MCNs to contextualize the clinicopathologic features, management, and outcomes of this case. The postoperative course was uncomplicated, and the patient delivered a healthy infant at term.

This case expands the histopathologic spectrum of pregnancy-associated pancreatic MCN and, to the best of our knowledge, is the first report of extensive stromal decidualization in MCN. In enlarging or symptomatic lesions during pregnancy, multidisciplinary evaluation and second-trimester resection can result in favorable maternal and fetal outcomes.

Categories: General Surgery, Pathology, Oncology
Keywords: decidualization, first and second trimester, mucinous cystic neoplasm (mcn), ovarian-type stroma, pregnant females, risk of pancreatic cancer

Introduction

Mucinous cystic neoplasms (MCNs) of the pancreas are uncommon cystic epithelial tumors that account for roughly 5% of all pancreatic neoplasms [1,2]. These lesions occur almost exclusively in women (female-to-male ratio of 20:1) and are characterized by the presence of ovarian-type subepithelial stroma, which distinguishes them from other pancreatic cystic lesions [1,3]. Distinguishing MCNs from other pancreatic cystic lesions, including pseudocysts and intraductal papillary mucinous neoplasms, is clinically important because their malignant potential and management strategies differ. In MCNs, the ovarian-type stroma is a defining feature that expresses estrogen receptor (ER) and progesterone receptor (PR), suggesting hormonal responsiveness [4]. Histologically, MCNs consist of mucin-producing columnar epithelium overlying a cellular, spindle-cell ovarian-type stroma [2]. Although the origin of this stroma remains uncertain, the embryonic incorporation of ectopic ovarian tissue has been proposed, and its hormonal responsiveness may explain pregnancy-associated decidualization, which does not itself indicate malignant transformation [4].

The occurrence of MCNs during pregnancy is extraordinarily rare, and since 1986, there have only been 37 reported cases [5]. These pregnancy-associated MCNs present unique management challenges due to their potential for rapid growth, the risk of malignant transformation, and the need to balance maternal and fetal safety [5]. Some pregnancy-associated MCNs may exhibit accelerated growth during gestation, with the mean size of MCNs in pregnancy being 13.5 cm, compared with 6 cm in the general population [5]. Thus, this raises concern for malignant transformation and the need for heightened clinical management.

The decidualization of ovarian-type stroma in pancreatic MCNs can occur during pregnancy secondary to

the effects of progesterone. However, overt decidualization in MCN is a rare phenomenon that, to the best of our knowledge, has not been well-described or documented histologically. Decidualization represents a morphologic transformation of stromal cells under hormonal influence, particularly progesterone, and is typically characterized by enlarged, rounded epithelioid cells with abundant cytoplasm. This process is well-recognized in reproductive organs during pregnancy but has not been reported or observed in ectopic ovarian-type tissue with regard to MCNs [6].

Case Presentation

Patient demographics and history

A pregnant woman aged 20-30 years (gravida 1, para 0) was evaluated at 16 weeks' gestation for an enlarging pancreatic tail cystic lesion initially identified before pregnancy. Her medical history was significant for sickle cell trait, with no family history of pancreatic or ovarian malignancies. She had no prior history of pancreatitis or pancreatic disorders.

Clinical presentation

The patient was initially asymptomatic, with the pancreatic lesion discovered incidentally during prenatal imaging. She reported mild nausea during early pregnancy, which improved with standard prenatal care. Physical examination revealed a soft abdomen with fullness consistent with a gravid uterus and no palpable masses or tenderness. Tumor markers were not performed at this time.

Imaging findings

MRI, 2023 (Prior to Pregnancy)

MRI demonstrated a large cystic lesion in the pancreatic tail measuring up to 10.6 cm, with peripheral nodules suggestive of prior rupture or higher risk of transformation. There was no definite communication with the pancreatic duct, and the lesion was favored to represent a mucinous cystic neoplasm (MCN).

CT, 2023 (Prior to Pregnancy)

A contrast-enhanced CT of the abdomen and pelvis (pancreatic protocol) demonstrated a 10 cm low-attenuation, nonenhancing lesion between the pancreatic tail, spleen, and left kidney. Differential considerations included pseudocyst, MCN, and solid pseudopapillary epithelial neoplasm (SPEN) (Figure *1A*, *1B*).

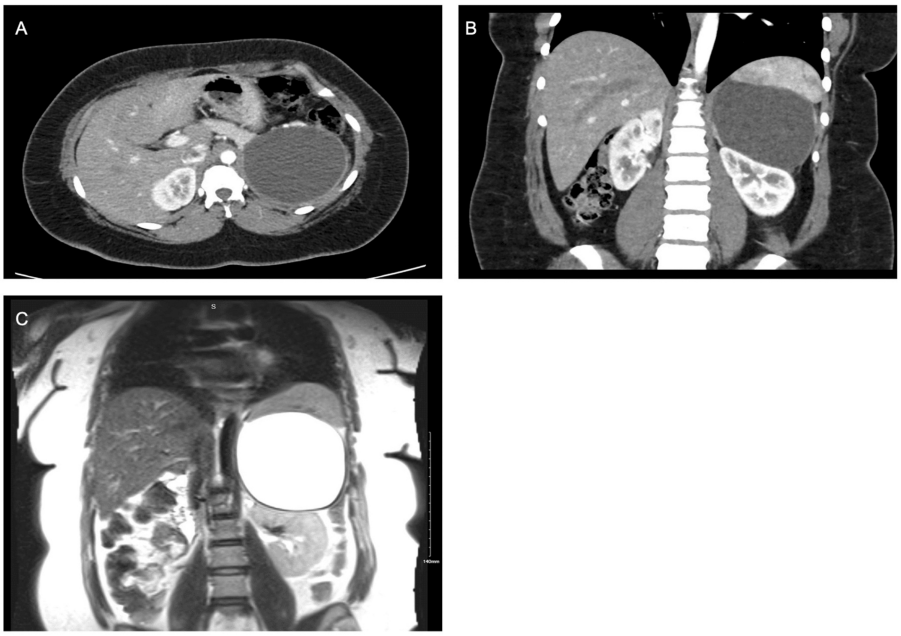


FIGURE 1: Cross-sectional imaging of pancreatic mucinous cystic neoplasm.

(A) Axial contrast-enhanced CT of the abdomen and pelvis (CTA of the pancreas) performed prior to pregnancy, demonstrating a well-circumscribed exophytic cystic lesion measuring approximately 9.2 × 9.7 × 8.2 cm arising from the pancreatic tail, displacing adjacent organs without evidence of invasion. (B) Coronal CT reconstruction from the same pre-pregnancy study, showing the large pancreatic cystic lesion with thin peripheral septations, abutting the spleen, stomach, and left kidney. (C) Coronal T2-weighted MRCP performed in early 2024 during pregnancy (gestational age approximately 15-16 weeks), demonstrating mild interval growth of the cyst to 12.5 × 11.2 × 12.5 cm with peripheral septations, without solid mural nodules or pancreatic duct dilation.

MRCP, magnetic resonance cholangiopancreatography; CTA, computed tomography angiography

Endoscopic Ultrasound (EUS), Late 2023 (Prior to Pregnancy)

EUS demonstrated a large cystic lesion measuring 9.9 × 7.9 cm without definite communication with the pancreatic duct. No nodules or solid mass were identified, although multiple septations were present. The cyst was not sampled because of concern for introducing infection.

Computed Tomography Angiography (CTA), Late 2023 (Prior to Pregnancy)

CTA again demonstrated a similar 9.2 cm cystic lesion along the pancreatic tail, possibly representing SPEN or a mucinous neoplasm.

Magnetic Resonance Cholangiopancreatography (MRCP), Early 2024 (~16 Weeks' Gestation)

Follow-up MRCP performed at 16 weeks' gestation demonstrated interval growth to 12.5 cm (12.0 × 11.2 × 12.5 cm), with thick walls and peripheral septations but no definite solid nodular component (Figure *1C*). Minor wall calcification was better seen on prior CT. The lesion displaced the spleen, splenic vessels, splenic flexure, and left kidney, without clear evidence of invasion. The overall findings suggested mild interval increase in the size of the pancreatic tail cystic lesion, now measuring up to 12.5 cm, with peripheral septations and no definite solid nodular component on noncontrast MRI. The lesion displaced adjacent structures without evidence of invasion.

Multidisciplinary management

Following consultation with maternal-fetal medicine specialists, the decision was made to proceed with surgical resection during the second trimester given the rapid growth pattern and uncertain malignant potential. The timing was chosen to minimize anesthetic risks to the developing fetus while allowing adequate time for fetal maturation.

Operative course

The patient underwent an attempted robotic-assisted distal pancreatectomy at 19 weeks' gestation, which was converted to an open procedure due to markedly dense inflammatory changes worrisome for either malignant transformation or contained rupture and the large size of the lesion yielding limited working space. The procedure included distal pancreatectomy, splenectomy, partial left adrenalectomy, and wedge gastrectomy due to adherent inflammatory tissue. It was not possible to dissect the pancreas or cystic lesion away from adjacent organs without disrupting it. Intraoperative fetal monitoring was maintained throughout the procedure.

Gross (macroscopic) pathological findings

The surgical specimen consisted of a 14.0 × 14.0 × 9.2 cm cyst containing yellow-brown fluid (Figure *2A*-*2D*). The cyst wall measured up to 0.8 cm in thickness with multiple areas of excrescences. The lesion was located 0.2 cm from the posterior pancreatic surface and 2.3 cm from the pancreatic margin, without the involvement of the main pancreatic duct.

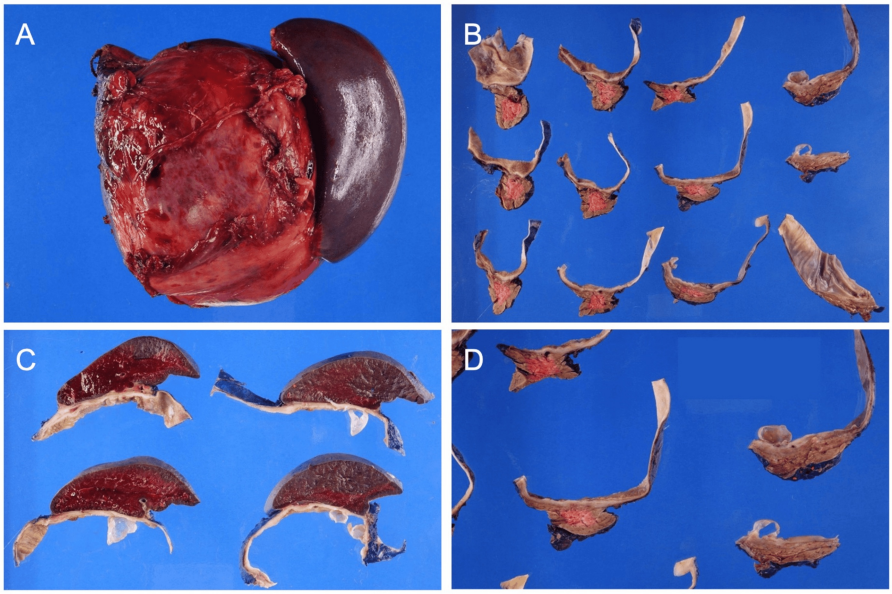


FIGURE 2: Gross examination of the pancreatic mucinous cystic neoplasm.

(A) En bloc resection specimen showing the large cystic mass arising from the body/tail of the pancreas, closely associated with the spleen. (B) Representative thin sections of the cyst wall demonstrating collapsed fibrotic cystic structures with smooth internal surfaces with minimal fibrous septations. (C) Cross-sections of the spleen demonstrating no gross evidence of involvement by the cystic lesion. (D) Higher magnification of the cyst wall sections highlighting the variably thickened fibrotic wall and delicate internal septations; portions of the cyst wall demonstrate variable thickness.

Histopathologic findings

Microscopic examination revealed a mucinous cystic neoplasm with low-grade dysplasia (Figure *3A*-*3D*), characterized by a simple mucinous epithelial lining with areas of flattening and erosion. The most striking feature was extensive decidualized ovarian-type stroma, which comprised most of the specimen and expanded throughout the tumor (Figure *3D*). The decidualized stromal cells were large, polygonal, and epithelioid, with abundant eosinophilic cytoplasm and enlarged nuclei. Areas of stromal hyalinization with focal calcification and ossification were also present.

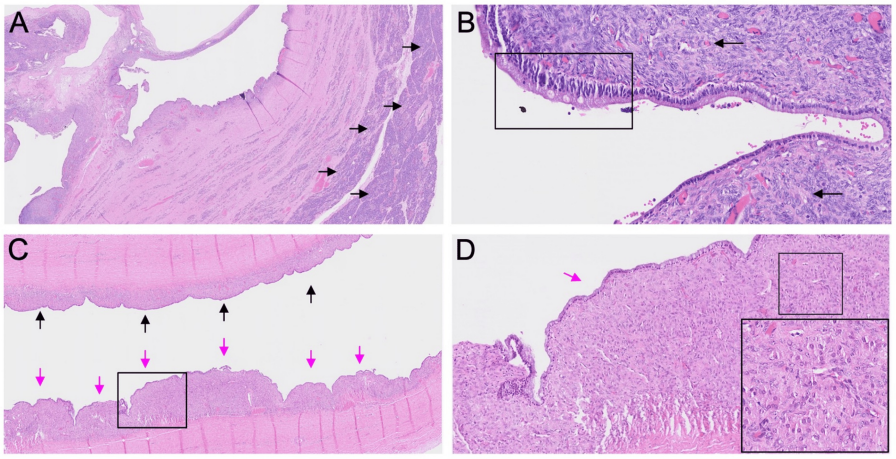


FIGURE 3: Histological sections of mucinous cystic neoplasm of the pancreas.

(A) Low-power architecture view demonstrating cystic spaces with characteristic ovarian-type stroma beneath. The surrounding pancreatic parenchyma is seen on the right (black arrows) (10×). (B) The cyst lining epithelium shows focal low-grade dysplasia (box), characterized by mild nuclear enlargement, nuclear stratification limited to the basal half, and preserved polarity. The underlying classic ovarian-type stroma (black arrows) is composed of bland spindle cells with scant cytoplasm and uniform nuclei (200×). (C) The upper portion (black arrows) shows non-decidualized ovarian-type spindle-cell stroma with bland nuclei and scant cytoplasm. The lower portion (pink arrows) shows the diffuse decidualization of the stroma with enlarged polygonal cells, abundant eosinophilic cytoplasm, and round nuclei. Most of the lesion demonstrates extensive decidualization (20×). (D) Higher-power view (of the black-boxed area in C) showing mucinous epithelium overlying expansile decidualized stroma (pink arrow) composed of large polygonal cells with abundant eosinophilic cytoplasm and bland round nuclei. Decidualization extends to the subepithelial stroma and into deeper stromal layers and comprises the majority of the neoplasm (100×).

Immunohistochemical results

Immunohistochemical staining further characterized the stromal component and supported ovarian-type stroma with decidual change (Figure *4A*-*4H*). CD10 (clone 56C6, product ID: PA0131; vendor/platform: Leica) highlighted only rare scattered stromal cells. WT1 (clone WT1, product ID: IR055; vendor/platform: Dako) showed strong nuclear staining in the ovarian-type stroma. Inhibin (clone MXR1, product ID: IR513; vendor/platform: Dako) demonstrated strong cytoplasmic positivity in the decidualized stromal cells, whereas the classic spindle-cell stroma showed only rare positive cells. PR (clone 636, product ID: IR068; vendor/platform: Dako) showed moderate to focal strong nuclear positivity, and ER (clone SP1, product ID: IR151; vendor/platform: Dako) showed moderate nuclear positivity, in both the classic spindle-cell stroma and the decidualized stromal areas. Calretinin (clone DAK-Calret-1, product ID: IR627; vendor/platform: Dako) was strongly positive in the decidualized stromal areas but largely negative in the classic spindle-cell stroma. The overlying mucinous epithelium showed no significant CD10 or calretinin expression. All antibodies were performed using high-pH antigen retrieval on their respective platforms.

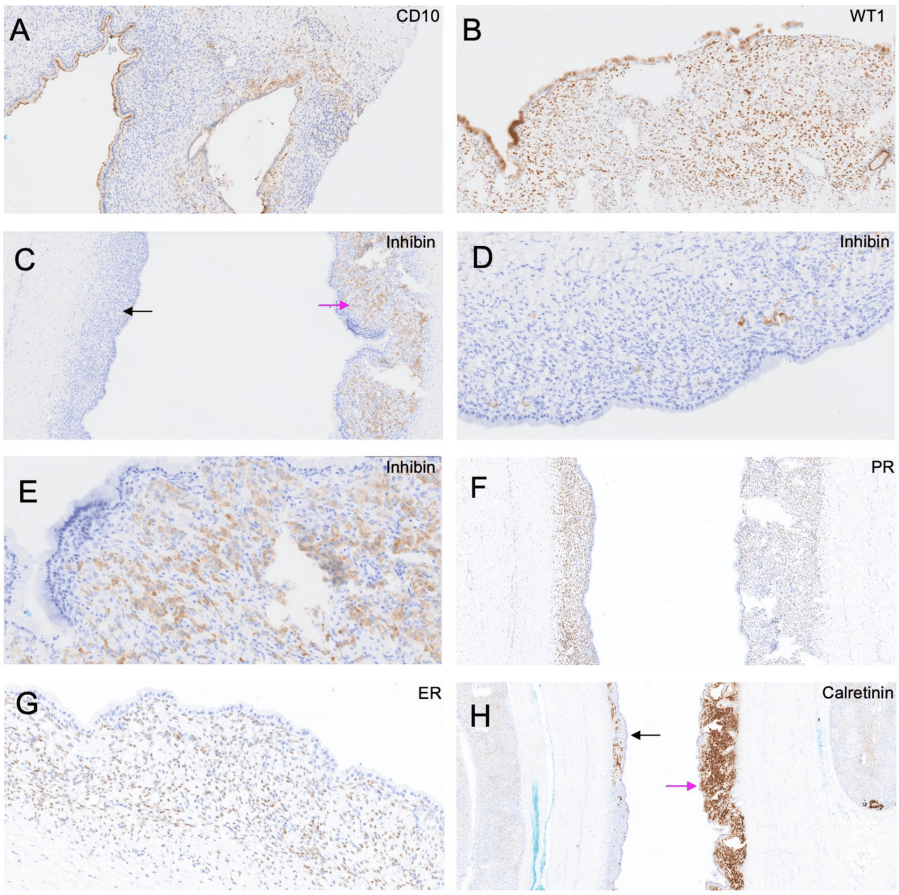


FIGURE 4: Immunohistochemical staining in MCN highlighting decidualized stromal areas.

(A) CD10 highlights rare scattered positive stromal cells, supporting the presence of ovarian-type stroma. The majority of the stroma is negative. The overlying mucinous epithelium shows no significant CD10 expression (100×). (B) WT1 shows strong nuclear staining in the ovarian-type stroma. The overlying mucinous epithelium shows cytoplasmic and weak nuclear staining (100×). (C-E) The classic ovarian-type spindle-cell stroma (C, left side of the image {black arrow}, low power: 20×; D, high power: 200×) shows rare inhibin-positive cells. (E) In contrast, the decidualized stromal areas (also C, right side of the image, pink arrow) show strong cytoplasmic positivity. (F) Progesterone receptor (PR) immunohistochemistry shows that strong nuclear positivity is observed in both the classic ovarian-type spindle-cell stroma and the decidualized stromal areas (50×). (G) Estrogen receptor (ER) immunohistochemistry shows moderate nuclear positivity in both the ovarian-type spindle-cell stroma and decidualized stromal areas (200×). (H) Calretinin immunohistochemistry demonstrates strong cytoplasmic and nuclear positivity in the decidualized stromal areas (pink arrow, right), while the classic ovarian-type spindle-cell stroma (black arrow, left) remains mostly negative with rare positive cells. The overlying mucinous epithelium shows no significant calretinin expression (20×).

MCN: mucinous cystic neoplasm

The combined WT1 nuclear expression and ER/PR positivity support ovarian-type stromal differentiation in the appropriate morphologic context, whereas the limited CD10 staining provides only ancillary support. Compared with the conventional spindle-cell stroma, the decidualized areas demonstrate markedly increased inhibin and calretinin expression, highlighting a distinct stromal immunophenotype. These findings, together with the characteristic cytomorphology and clinical context of pregnancy, support extensive stromal decidualization, although inhibin and calretinin are not specific markers of this process.

Postoperative course and follow-up

The patient experienced an uncomplicated postoperative recovery with the resolution of pregnancy-related nausea and emesis. She was discharged on postoperative day 7 and continued regular obstetric care. She delivered a healthy infant at 39 weeks' gestation via vaginal delivery. At six-month follow-up, the patient remained asymptomatic, with no evidence of recurrence.

Discussion

Mucinous cystic neoplasms (MCNs) are uncommon pancreatic cystic epithelial neoplasms defined by mucinous epithelium and ovarian-type stroma, with marked female predominance and typical body/tail location [1-3]. Pregnancy-associated MCNs are reported mainly as case reports and small series, but available data suggest presentation at younger age, frequent symptoms, large size, and occasional interval growth during gestation [2,3,5]. In the largest systematic review, median age was 32 years, mean tumor size was 13.5 cm, and 27% enlarged during pregnancy, supporting possible hormone-associated growth without proving greater malignant potential [5].

The key feature of this case is the extensive decidualization of the ovarian-type stroma. Ovarian-type stroma is diagnostic of MCN and often expresses progesterone receptor, with variable estrogen receptor positivity, supporting hormonal responsiveness [1,4,5]. Although decidualization is a recognized progesterone-driven stromal change in reproductive tissues, overt decidual change in pancreatic MCN appears uncommon. To our knowledge, this is the first reported pregnancy-associated pancreatic MCN in which decidualized stroma comprised the majority of the lesion.

Our patient shared recognized features of pregnancy-associated MCN, including young age, body/tail location, gestational enlargement, and favorable maternal-fetal outcome after resection. The lesion increased from 10.6 cm on pre-pregnancy MRI to 12.5 cm on MRCP at 16 weeks' gestation, representing a 1.9 cm increase in reported maximum diameter, and measured 14 cm at resection. However, differences in imaging technique and gross specimen measurements limit the precise assessment of growth. Histology showed only focal low-grade dysplasia despite its large size and striking stromal change. As summarized in Table *1*, this combination of rapid pregnancy-associated growth and diffuse decidualized ovarian-type stroma makes this case novel and unique and is not reported in the literature.

Number	Author/citation	Age at diagnosis	Gestational age at diagnosis (weeks)	Timing of operation	Clinical presentation/complications	Surgical procedure	Histological diagnosis
1	Olsen et al. [7]	25	5	Week 18	NR	DP	Adenoma
2	Ganepola et al. [8]	37	4	Week 23	NR	DP	Adenoma
3	Kato et al. [9]	33	15	Week 23	IUGR	DP	Adenoma
4	López-Tomassetti Fernández et al. [10]	26	20	Week 26	Episodic epigastric pain	DP	Adenoma
5	Herring et al. [11]	34	3	Week 17	NR	DP	Adenocarcinoma
6	Hakamada et al. [12]	38	2 years before pregnancy	Second trimester	Nausea, hematemesis, and tarry stool	DP + partial stomach resection	Borderline
7	Wiseman et al. [13]	32	11	Week 15	Intractable nausea	DP	Low-grade dysplasia
8	Brown et al. [14]	38	8	Week 8	Gastrointestinal bleeding	DP	Severe dysplasia
9	Martins Filho et al. [15]	20	20	Week 20	No	DP	Adenoma
10	Boyd et al. [16]	21	7 months before pregnancy	Week 20	Abdominal distension and fullness	DP	Moderate dysplasia
11	Tsuda et al. [17]	28	Some years before pregnancy	Week 18	No	DP	Severe dysplasia
12	Veits et al. [18]	28	11	NA	Pancreatitis	DP	Low-grade dysplasia
13	Revoredo et al. [4]	38	17	Week 29	Rupture	DP	Moderate dysplasia
14	Revoredo et al. [4]	30	18	Week 20	No	DP	Adenocarcinoma
15	Alomair et al. [1]	33	9	Week 20	Episodic epigastric pain	DP + S	Mucinous cystadenoma (adenoma)
16	Lee et al. [3]	35	26	Week 26	Fetal distress	DP + S	Low-grade MCN
17	Spinak et al. (present case)	20-30	11 months before pregnancy	Week 19	No	DP + S	Low-grade MCN

TABLE 1: Pregnancy-associated pancreatic mucinous cystic neoplasms with surgical resection before delivery.

This table provides a descriptive summary of selected published cases of pancreatic MCN resected during pregnancy and is not intended as a systematic review.

DP, distal pancreatectomy; S, splenectomy; MCN, mucinous cystic neoplasm; NR, not reported; NA, not available; IUGR, intrauterine growth restriction

Management should balance maternal oncologic risk and fetal safety. Available reports support MRI for evaluation in pregnancy and second-trimester resection for symptomatic, enlarging, or radiologically concerning lesions [2,3,5,19]. In Dhamor et al.'s review, distal pancreatectomy with or without splenectomy was most common, cyst rupture occurred in 10.8%, and no maternal or fetal deaths were reported [5]. Our patient's second-trimester resection, uncomplicated recovery, and term delivery are consistent with this literature. Overall, this case suggests that dramatic stromal decidualization may reflect benign hormone-driven change rather than something more alarming.

This report has several limitations. As a single case with limited postoperative follow-up, the findings cannot be generalized, and the relationship between pregnancy-associated hormonal changes and stromal decidualization cannot be established definitively. Differences in imaging modalities limit the precise assessment of tumor growth. Additionally, the small number of published cases and potential publication bias toward unusual or clinically significant presentations limit conclusions regarding biological behavior and optimal management. Although immunohistochemical findings support the diagnosis, the markers used are not specific for decidualization. The absence of serum tumor markers and cyst fluid analysis further limited preoperative characterization; however, given the lesion's substantial size, interval enlargement, and uncertain malignant potential, additional testing was unlikely to alter the decision for surgical resection.

Conclusions

This rare pregnancy-associated pancreatic MCN expands the histopathologic spectrum of MCN by showing that ovarian-type stroma can undergo extensive decidualization during pregnancy. The recognition of this change is important because it may appear alarming grossly or microscopically despite low-grade epithelial dysplasia. The correlation of imaging, gross findings, histology, and immunohistochemistry is essential. In this case, multidisciplinary evaluation and second-trimester resection were associated with favorable maternal and fetal outcomes, although the findings cannot be generalized from a single case.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Acquisition, analysis, or interpretation of data: Yevgen Chornenkyy, Jacob Spinak, Tara Kent, Jeanne Carbone, Gamze Ayata, Vikram Deshpande, Christopher Felicelli

Drafting of the manuscript: Yevgen Chornenkyy, Jacob Spinak, Tara Kent

Critical review of the manuscript for important intellectual content: Yevgen Chornenkyy, Jacob Spinak, Tara Kent, Jeanne Carbone, Gamze Ayata, Vikram Deshpande, Christopher Felicelli

Supervision: Yevgen Chornenkyy

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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