Acute Pancreatitis with a Mucinous Cystoadenoma of the Pancreas in Pregnancy

STEFANIA ASCIUTTI¹, TOMI T KANNINEN², GRAZIANO CLERICI², ELISABETTA NARDI¹, DANilo CASTELLANI¹, GIAN CARLO DI RENZO² and CARLO CLERICI¹

Departments of ¹Gastroenterology and ²Obstetrics and Gynecology, University of Perugia, Italy

Abstract. Pregnancy complicated by pancreatitis is a rare and difficult clinical situation. Gallstones are the most frequent cause of pancreatitis in pregnancy. Non-gallstone pancreatitis in pregnancy has been shown to be significantly more prone to premature delivery and pseudocyst formation. Cystic lesions as a cause of pancreatitis in pregnancy have not, to our knowledge, been observed. Pancreatic cystic lesions in general are rare, but are difficult to treat given problems in clarifying their malignancy. Mucinous cystic neoplasms are considered premalignant lesions and resection is recommended. Receptors for estrogen and progesterone receptors in these cysts may cause cystic growth during pregnancy. Treatment recommendations for pancreatitis in pregnancy are not well defined; this applies as well to treatment protocols for cystic lesions. In this case report we describe a new potential cause of acute pancreatitis in pregnancy due to compression of the principal pancreatic duct by a mucinous cystoadenoma.

Pregnancy complicated by pancreatitis is a rare and difficult clinical situation to manage. The occurrence of pancreatitis in pregnancy has been reported to be between one in 1,000 to 12,000 patients (1-3).

Gallstones are the most frequent cause of pancreatitis in pregnancy, accounting for 66% of the total cases in one large multi-institution review (2). Non-gallstone pancreatitis in pregnancy has been shown to be significantly more prone to premature delivery and pseudocyst formation (2). Cystic lesions as a cause of pancreatitis in pregnancy have not, to our knowledge, been observed.

Pancreatic cystic lesions in general are rare and include pseudocysts, cystic neoplasms and congenital cysts. Cystic neoplasms are divided into four types of neoplasm: serous, mucinous, intraductal and papillary cystic. The variable malignancy and difficulty in forming a clear diagnosis poses clear problems in treatment selection (4).

Mucinous cystic neoplasms (MCNs) are considered premalignant lesions and resection is recommended. Importantly, the presence of estrogen and progesterone receptors in the stroma of these cysts may cause cystic growth during pregnancy (5).

Pancreatitis in pregnancy is difficult to treat and studies have not been able to draw concrete conclusions on patient treatment recommendations, especially in patients with non-gallstone pancreatitis. Additionally, the treatment of cystic lesions in pregnancy has not been well defined.

In this case report, we describe a case of acute pancreatitis caused by compression of the principal pancreatic duct by a mucinous cystoadenoma in pregnancy.

A 31-year-old female patient, II gravida 1 para, at 23 weeks and 6 days of gestation was transferred from a peripheral hospital with suspected acute pancreatitis after having epigastric pain of the abdomen, a palpable mass in the epigastric region of the abdomen and generalized bloating without nausea, vomiting or fever. The patient history showed that a 1.5 cm round, thick-walled, anechogenic lesion had been found in the tail of the pancreas during a follow-up abdominal ultrasound examination following a resection of a parachordoma. Given the lack of malignant signs on imaging and symptomatology, the patient was conservatively managed with a follow-up in 6 months (6). The patient became pregnant before the scheduled follow-up, which she did not attend. Biochemical evaluation, at admission, was normal except for an elevated amylase of 1633 UI/l, with normal calcium, alkaline phosphatase (ALP), gamma-glutamyl transpeptidase (G-GT), aspartate aminotransferase (AST), alanine aminotransferase (ALT) and triglycerides. The patient had no history of alcohol abuse and had immunity to mumps. Abdominal ultrasound showed a normal gallbladder, devoid of stones, with nondilated bile ducts. The head and body of the
pancreas appeared normal; the tail seemed completely replaced by an anechoogenous 83×62 mm lesion, with hyperechoic spots and septa (data not shown). An abdominal magnetic resonance imaging (MRI) was then performed which showed a round, liquid-filled, thick-walled 80×50 mm lesion, with an eccentric solid component and septa within, giving a polychambered aspect (Figure 1). This lesion was in continuity with a similar 19 mm lesion. The main pancreatic duct and the parenchyma of the tail of the pancreas were not visualized. The suspected diagnosis was of an MCN. Endoscopic ultrasound confirmed the presence of the lesion, which compressed the tail end of the main pancreatic duct (Figure 2). At three days from admission, C-reactive protein was 9.5 mg/dl, amylase 2265 UI/l, lipase 928 UI/l and CA 19.9 213.7 U/ml (normal <35 U/ml). The patient was conservatively managed for pancreatitis and was released after all biochemical examinations returned to normal, with a scheduled follow-up in one month.

Three weeks after being released, the patient was admitted again after developing epigastric abdominal pain with nausea and vomiting. Blood examinations were normal except for amylase of 2180 UI/l and lipase of 1550 UI/l. The patient developed fever two days after being admitted. Given the worsening clinical situation of the patient, the risk to the fetus and the potential of malignancy of the cystic lesion, a cesarean section was performed at 33 weeks and 5 days of gestation after induction of fetal pulmonary maturity (betamethasone 12 mg q24h for 2 doses) 3 days before cesarean section. The newborn was a male of 2030 g and had an Apgar of 6 at 1 minute and 10 at 5 minutes; there were no pathologies or abnormal examinations at follow-up one year after birth.

At one month post cesarean section, the patient was scheduled for an ultrasound, which confirmed the previous findings. Laparotomic distal splenopancreasectomy was performed to resect the lesions. The histological examination of the resected lesions confirmed the presence of an MCN, without atypia. Subsequent follow-ups were negative for recurrence.

Discussion

Gallstones are responsible for the majority of cases of pancreatitis in pregnancy. Outcome with gallstone pancreatitis is better with respect to preterm delivery, recurrence, maternal morbidity and pseudocyst formation when compared to other causes. Non-gallstone pancreatitis has been shown to have poorer outcomes than gallstone pancreatitis. This is amplified in traumatic, hyperlipidemic and alcohol-induced pancreatitis. Idiopathic causes have been reported to be the second most frequent cause of pancreatitis in pregnancy, with a frequency around 16.8% (2). Cystic lesions in the absence of gallstones as a cause of pancreatitis in pregnancy have not, to our knowledge, been observed.

Treatment for pancreatitis usually consists of conservative treatment in non-pregnant patients, with rehydration and antibiotic treatment, if necessary. Treatment of pancreatitis in pregnancy presents unique problems, given the effects of the maternal clinical situation on the fetus. Currently, surgical intervention has been supported by various authors in cases of gallstone pancreatitis, being that patients in their studies with conservative treatment in pregnancy had worse outcomes (3, 7). Other studies have not been able to draw concrete conclusions on patient treatment decisions, especially in patients with non-gallstone pancreatitis (2). Additionally, the treatment of cystic lesions is more complicated given the additional physiological hormonal changes occurring during pregnancy (8).

Pancreatic cystic lesions, in general, are rare and include pseudocysts, cystic neoplasms and congenital cysts. Pseudocysts make up the majority of cystic lesions and can usually be treated conservatively in the absence of symptoms. Conservative treatment is also recommended for congenital cysts (4). The variable malignancy and difficulty in forming a clear diagnosis of cystic neoplasms poses clear problems in treatment selection. The problem has been amplified by the greater availability of imaging techniques that has increased the frequency of incidental cyst diagnosis. This creates a situation where the identification of a low malignancy sub-group would be advantageous in order to avoid unnecessary interventions. Currently, cross imaging (computer tomography and MRI) combined with fine-needle aspiration are the diagnostic tests of choice. However, imaging is not always capable of forming a definite diagnosis, which causes problems in choosing conservative or surgical treatment options, and fine-needle aspiration, though useful, can potentially spread malignant cells at the point of insertion (9-10). Moreover, in most studies the sensitivity of cytology obtained during fine-needle aspiration is reported to be between 27% to 64% given biopsy specimens are often falsely negative because of sampling error (11). Complications (infections, bleeding, and perforation), occurring in up to 4% of patients, may also limit the use of this technique (12).

MCNs make up 40-50% of the cystic neoplasms of the pancreas (13). MCNs tend to arise exclusively in women between the age of 35-90 years and have an increased malignant potential when present in older patients. Symptoms are generally non-specific such as abdominal pain, weight loss, back pain, jaundice, postprandial fullness and a palpable mass. The presence of symptoms increases the chance of an MCN developing malignancy (13). MCNs are considered premalignant lesions and resection is recommended. MCNs are characterized by large thick-walled, septated cysts, with ovarian type stroma and the absence of connections to the ductal systems.
Of particular importance is the presence of estrogen and progesterone receptors in the stroma of these cysts (8). This may cause augmented cystic growth during the physiological hormonal increase during pregnancy. The association between mucinous cystoadenomas and pregnancy has been reported before and the increase in the mass of tumors with estrogen and progesterone receptors has been documented in other studies (8, 14-16).

We believe this is the first description of acute pancreatitis caused by the growth of an MCN during pregnancy. The growth of the cyst seemed directly responsible for the development of the acute pancreatitis, following the compression of the main pancreatic duct. Clinicians should be aware of the potential growth and compressive nature of these types of cysts. This is important in asserting the possible causes of pancreatitis during pregnancy, especially in an individual with known cysts.

As highlighted above, currently the treatment protocol for pancreatitis in pregnancy is not well defined, especially for patients with non-gallstone pancreatitis. Clarity in identifying correct treatment options is an important step in improving maternal-fetal outcomes in these cases. In particular, treatment of certain cystic lesions associated with pancreatitis in pregnancy has not been well established, and this case highlights the need for specific guidelines.

References


