Esophageal achalasia presenting during pregnancy

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We report a woman with achalasia cardia who developed dysphagia for the first time during pregnancy. She was initially mistakenly treated elsewhere as hyperemesis gravidarum. The diagnosis and treatment of achalasia during pregnancy is reviewed. [Indian J Gastroenterol 1997; 16: 72-73]

Key words: Balloon dilatation, endoscopic treatment, esophageal motor disorder

Symptoms of achalasia cardia include difficulty in swallowing, regurgitation and weight loss. Though the disorder frequently affects young adults, appearance for the first time during pregnancy is uncommon. We report such a case in which the symptoms were initially attributed to hyperemesis gravidarum.

A 20-year-old, five-month pregnant woman presented with a history of dysphagia for both solids and liquids of two and a half months' duration. She had not noticed any worsening of dysphagia with hot or cold foods. She had frequent regurgitation of food for the same duration and had lost nearly 6 Kg weight during the illness.

Initially, her symptoms were considered to be related to pregnancy (hyperemesis gravidarum) and she was treated with antacids, antispasmodics and H2-receptor blocking agents; when the symptoms did not improve, barium swallow was performed during the fifth month of pregnancy. This showed no evidence of esophageal stricture; the lower end of the esophagus had a beak-like appearance but there was no dilatation of the esophageal body and no evidence of tertiary contractions. She was at this stage referred to our hospital.

On examination, apart from a palpable gravid uterus and evidence of recent weight loss, there was no significant finding.

Upper gastrointestinal endoscopy revealed normal esophagus and gastric fundus. Esophageal manometry showed features classical of achalasia: namely, total spasm of esophageal body, presence of simultaneous contractions, lower esophageal sphincter (LES) pressure of 68 mmHg and incomplete relaxation of the LES on water swallow.

The patient was treated with balloon dilatation of the LES under endoscopic guidance. Owing to her pregnancy, we chose to avoid fluoroscopy and sedation. Following this, she had marked relief in symptoms and was able to swallow both liquids and solids without any difficulty. Regurgitation also disappeared completely. She was well for nearly 10 weeks following the procedure and gained weight. She had an abortion during the seventh month of gestation. Subsequently, she has had a normal pregnancy culminating in normal full-term vaginal delivery; her symptoms of dysphagia and regurgitation have not returned.

Achalasia cardia frequently affects young adults; thus pregnancy is not unusual in patients with achalasia cardia. However, appearance of symptoms for the first time during pregnancy, as occurred in our patient, is unusual. It is in fact believed that since estrogens and progesterone reduce the LES pressure, pregnancy may initially alleviate symptoms of achalasia.

Common symptoms of achalasia include dysphagia, regurgitation and weight loss. Pregnancy leads to a variety of gastrointestinal symptoms, e.g., heartburn, nausea and vomiting, leading to weight loss when these symptoms are pronounced; however, dysphagia is not known to occur with pregnancy. In our patient, symptoms were initially mistakenly attributed to pregnancy: a good history should have led to a suspicion of esophageal disease earlier since dysphagia is not a feature of pregnancy. If achalasia was suspected earlier, esophageal manometry would have been the investigation of choice instead of barium esophagram.

The treatment of achalasia during pregnancy needs a few special considerations. It may be preferable to avoid medical therapy in view of risk to the fetus. Pneumatic dilatation has been performed safely during pregnancy, in one of these cases, endoscopic guidance was used. Premedication and fluoroscopy must be avoided. Cardiomyotomy should be reserved for a rare patient who continues to have severe symptoms despite dilatation.

References
Cystadenoma with mesenchymal stroma mistaken for hepatic hydatid cyst

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Hepatobiliary cystadenoma with mesenchymal stroma occurs uniquely in women, can be associated with other hepatobiliary anomalies, and has the potential for malignant transformation. We describe a case of hepatobiliary cystadenoma with mesenchymal stroma which was initially treated as hepatic hydatid cyst. [Indian J Gastroenterol 1997; 16: 73-74]

Key words: Cystic tumor, liver

Hepatobiliary cystadenoma is a rare biliary tumor that usually arises in the liver and less frequently in the extrahepatic bile ducts. In countries where hydatid disease is endemic, differentiation from echinococcal cysts is important. We describe a case of hepatobiliary cystadenoma with mesenchymal stroma (CMSSM) tumor.

A 35-year-old woman presented with a discharging sinus in the epigastrium. She had undergone surgery for suspected hydatid cyst of the left lobe of the liver about a year back. A continuous discharge of about 200 mL of thin yellowish fluid through the tube drain left in the cyst cavity and a 10 cm × 10 cm residual cavity in the liver on ultrasonography led to re-operation and marsupialization of the cyst. The wound failed to heal and she continued to discharge clear fluid. She was then referred to us.

Ultrasonography revealed a 6.6 cm × 9.3 cm × 10.3 cm multicystic lesion in the left lobe of the liver; she also had cholelithiasis. CT scan revealed the cyst wall to be regular with some solid areas; other findings were similar to those on sonography. Liver function tests were normal and the discharging fluid was sterile. Hydatid serology (ELISA) was marginally positive.

At surgery a multiloculated cavity was found in the left lobe of the liver adherent to the anterior abdominal wall. It contained multiple cysts, 1-3 cm in size, with clear fluid. The cavity was resected along with the left lobe of the liver. A small portion of the cyst wall adjacent to the quadratus lobe was not removed as it was stuck to the porta; it was fulgurated with cautery. Cholecystectomy was performed; the gall bladder was intrathoracic, with anomalous cystic duct.

On gross examination, the resected specimen showed a large cystic area without a definite capsule and multiple cysts of variable size containing clear fluid and smooth lining, densely adherent to the tissues. Microscopically the lesion comprised glandular and cystic structures consisting of an inner layer of cuboidal to columnar epithelial cells with papillary projections into the lumen at some places. The middle stromal layer comprised primitive, spindle-shaped cells with rounded nuclei; the outer layer had collagenous connective tissue with blood vessels and inflammatory cells (Fig). The gall bladder showed evidence of chronic cholecystitis.

The postoperative period was uneventful. She has been advised three-monthly follow-up.

CMS tumor was described by Wheeler and Edmonson in 1985. Fifty three cases of cystadenomas had been reclassified into CMS tumors till 1990. These tumors were found to be different from cystadenoma without mesenchymal stroma with respect to exclusive occurrence in women, presentation at younger age, high rates of recurrence, high chances of malignant transformation, and a distinct histologic picture. Embryologically, these tumors arise from foci of primitive hepatobiliary and mesenchymal cells, in contrast to cystadenoma without mesenchymal stroma which may arise from formed biliary epithelium. Classically, CMS tumors consist of three distinct layers: (1) an epithelial layer of mucin-producing cuboidal to columnar cells lining the cysts; (2) a middle layer of undifferentiated mesenchymal cells usually less than 3 mm in thickness; and (3) an outer layer of collagenous connective tissue, usually 2-3 mm in thickness.

The most successful form of therapy is total excision; however, about 30%-50% of patients undergo incomplete resection at the time of their first surgery. Incomplete excision is associated with a significant risk of recurrence, secondary infection and possible malignant transformation.