Klinisch-Pathologische Konferenz

07.10.2009, 12.15 Uhr
Hörsaal D, Hörsaalzentrum im Innenhof zwischen Chir. Univ.-Klinik und Univ.-Klinik f. Inn. Medizin

Case 138: 39-year-old pregnant woman with jaundice

This 39-year-old woman and mother of 5 healthy children (ages 3 to 18 years) presented to the emergency room (ER) two days before Christmas 2008 with itching and jaundice. She is a housewife and her husband a hospital orderly. As she had experienced morning nausea for the previous 2 weeks, she suspected that she might again be pregnant and a pregnancy test in the ER was in fact positive. She had no abdominal pain, fever, diarrhea or weight loss.

Physical examination revealed scleral icterus and scratch marks on her neck, abdomen and extremities. BP was 120/80 mm Hg, pulse 108/min and regular, cardiopulmonary exam within normal limits, soft abdomen, liver 5 cm below the costal margin, normal bowel sounds, no masses, neurological exam unremarkable.

Laboratory on admission: total bilirubin 10.6 mg/dl (0.1-1.2), alkaline phosphatase 393 U/l (35-105), GGT 654 U/l, AST 175 U/l, ALT 410 U/l (-35), LDH 253 U/l (120-240), prothrombin time (PT) 95% (70-120), PT INR 1.04, APTT 39 sec (26-36), fibrinogen 715 mg/dl (210-400), CRP 5.9 mg/l (-8.0), glucose 131 mg/dl (70-115), serum K+ 2.9 nEq/l (3.5-5.0), within normal limits: Na+, Cl-, creatinin, urea, CPK; IgG4 0.45 g/l (0.08-1.4), WBC 8.7 g/l (4.4-11.3), Ec 4.6 T/l (4.1-5.1), Hb 13.9 g/dl (12.0-15.3)

CA 19-9 was 6 U/ml (-37), CA50 10 U/l (0-23), chromogranin A 35 ng/ml (0-100).

Abdominal ultrasound revealed a large dilated gallbladder, common duct 1.3 cm in diameter, dilated intrahepatic bile ducts and a 4x4 cm mass in the head of the pancreas. ERCP with emphasis on minimal radiation exposure revealed a string-like stenosis 4-5 cm in length immediately above the papilla, and dilated intrahepatic bile ducts. A small papillotomy was performed; the stenosis was very firm and a 10 French stent could only be placed with some difficulty. Cytology obtained from the common duct upon ERCP: red cells, granulocytes and lymphocytes (blood), detritus, fragments of connective tissue, some columnar cells with eccentric nuclei, some goblet cells. Bilirubin started to drop and the patient was discharged to spend Christmas Eve with her family. An outpatient MR on December 29, 2008, for which the radiologist demanded a "vital indication for the mother" because of the pregnancy, again revealed a 4x4 cm mass in the head of the pancreas with encasement of the common duct, the portal vein and the superior mesenteric artery. Dilatation of the main pancreatic duct to 5 mm; with stent in place the common duct was then of normal caliber. Between segments 6 and 7 of the liver there was a 1 cm oval signal enhancement (T2-weighted). The left lobe of the liver contained a lesion 3.4 cm in diameter with inhomogeneous uptake of contrast medium, "possibly due to metastatic spread." Several paraaortal lymphnodes 1 cm in diameter on the left. The consultant surgeon judged the tumor in the head of the pancreas to be inoperable. Biopsy and chemotherapy, possibly neoadjuvant, was recommended. The patient was readmitted on January 7, 2009, for sonographically guided fine-needle (FN) biopsy of the mass. Bilirubin had fallen to a normal value. Obstetrical-gynecological consultation revealed a normal pregnancy at 6 weeks' gestation. FN biopsy showed fibrotic changes with lymphocytic inflammation and some normal pancreatic parenchymal cells. Since no neoplastic tissue was found, another sonographically guided FN biopsy was taken, with the same result as the first biopsy. Pancytokeratin antibody was requested by the gastroenterologists and it was without pathological pattern.

Following the second biopsy, the patient developed severe upper right quadrant pain with poor response to opiates. Hb dropped to 8 g/dl (Ec 2.1, Hct 23). Emergency CT showed a 12x12x26 cm retroperitoneal hematoma, with hemorrhage from a segmental artery of the right kidney and formation of a pseudoaneurysm (2.7 x2.1 cm). An endovascular stent was placed and the leak was successfully sealed; subsequently, arterial perfusion of the entire right kidney was normal. After 2 days in intensive care, the patient was returned to the GI ward.A diagnosis was established.

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Lösung CPC 138

Diagnose: Autoimmunpankreatitis, Typ 2
Diagnost. Text: CT-gezielte Biopsie des Pankreaskopftumors nach Abbruch der Schwangerschaft und Verlaufsbeobachtung (Leberläsion war fokale noduläre Hyperplasie)
Verlauf und Therapie: Nach 9 Monaten immer noch Choledochusstent nötig (3x gewechselt), Cholangioskopie zeigt entzündliche derbe Stenose. Unter zuerst Steroiden, dann Azathioprin weitgehend beschwerdefrei, im MR Größenabnahme der Raumforderung im Pankreaskopf.
Diskutant: Prof. Jamie Barkin, University of Miami, Florida