Case 882

Eurorad

Focal Nodular Hyperplasia

Published on 01.07.2001

DOI: 10.1594/EURORAD/CASE.882
ISSN: 1563-4086
Section: Abdominal imaging
Imaging Technique: CT
Imaging Technique: MR
Case Type: Clinical Cases
Authors: J.L. Turkenburg, E.L. van Persijn van Meerten
Patient: 36 years, female

Clinical History:

presented with a 3-year history of progressive malaise, watery diarrhea, muscle weakness and a 10 kg weight loss. On admission, she weighted 46 kg (body length 1.67m). She was afebrile, but slightly dehydrated with persistent diarrhea also during fasting. The fecal potassium was elevated (97mmol/L). Laboratory examination of the serum revealed severe hypokalemia (2.2mmol/L), mild hyperglycemia (6.2mmol/L) and a reduction of glucose tolerance. Coloscopy disclosed no abnormalities. CT scan and MRI of the abdomen were performed.

Imaging Findings:

A 36-year-old woman presented with a 3-year history of progressive malaise, watery diarrhea, muscle weakness and a 10 kg weight loss. On admission, she weighted 46 kg (body length 1.67m). She was afebrile, but slightly dehydrated with persistent diarrhea also during fasting. The fecal potassium was elevated (97mmol/L). Laboratory examination of the serum revealed severe hypokalemia (2.2mmol/L), mild hyperglycemia (6.2mmol/L) and a reduction of glucose tolerance. Coloscopy disclosed no abnormalities. CT scan and MRI of the abdomen were performed.

Discussion:

The clinical syndrome of our patient is known as the Verner-Morrison syndrome, which represents a triad of signs: watery diarrhea, hypokalemia and dehydration. This syndrome is characteristic for vasoactive intestinal peptide (VIP)-producing tumors. The secretory diarrhea is usually large in volume (> 1L/day), and persists also during fasting. Hypokalemia is the result of important potassium loss into the diarrhea fluid. Ninety percent of the VIP-producing tumors are nonbetaislet cell tumors of the pancreas, the so-called VIPomas. In 10%, these tumors are extra-pancreatic in location. Most of them are neutral crest tumors, originating from neural, adrenal or periganglionic tissue. VIP-producing adrenal tumors composed of ganglioneuroblastoma and pheochromocytoma elements are very rare. This type of tumor in our patient produced both VIP and catecholamines, but the clinical presentation was mainly determined by VIP secretion. Pheochromocytomas and ganglioneuroblastomas are hypervascular and show inhomogeneous enhancement after intravenous injection of contrast medium with areas of hypoattenuation, indicating necrosis or cystic degeneration. On MRI these tumors are characterized by a low to intermediate signal intensity on T1-weighted sequences and a high signal intensity on T2-weighted sequences. Additional findings on CT and MRI are distended fluid-filled bowel loops because of VIP induced inhibition of bowel motility and excessive secretion of fluid and electrolytes into the small bowel. In the majority of the patients the VIP-producing tumor is malignant. Metastases located preferentially in the liver are found in half of the patients at the time of diagnosis. The lesion in the liver of our patient however proved to be FNH.

Differential Diagnosis List: Focal Nodular Hyperplasia

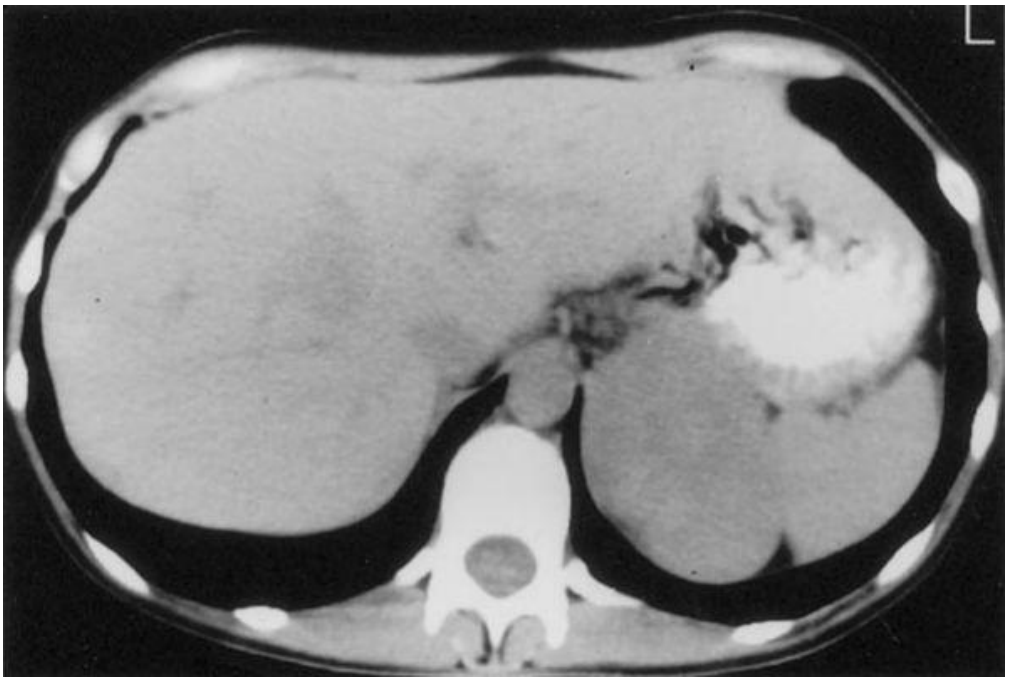
Final Diagnosis: Focal Nodular Hyperplasia

References:

Krejs GJ. VIPoma syndrome. Am J Med 1987; 82: 37-46. (PMID: 3035922)
Mekhjian HS, O'Doriso TM. VIPoma syndrome. Seminars in Oncology 1987; 14: 282-291. (PMID: 2820063)
Tjon A Tham RT, Jansen JBMJ, Falke THM et al. MR, CT and ultrasound findings of metastatic vipoma in pancreas. JCAT 1989; 13: 142-144. (PMID: 2778140)

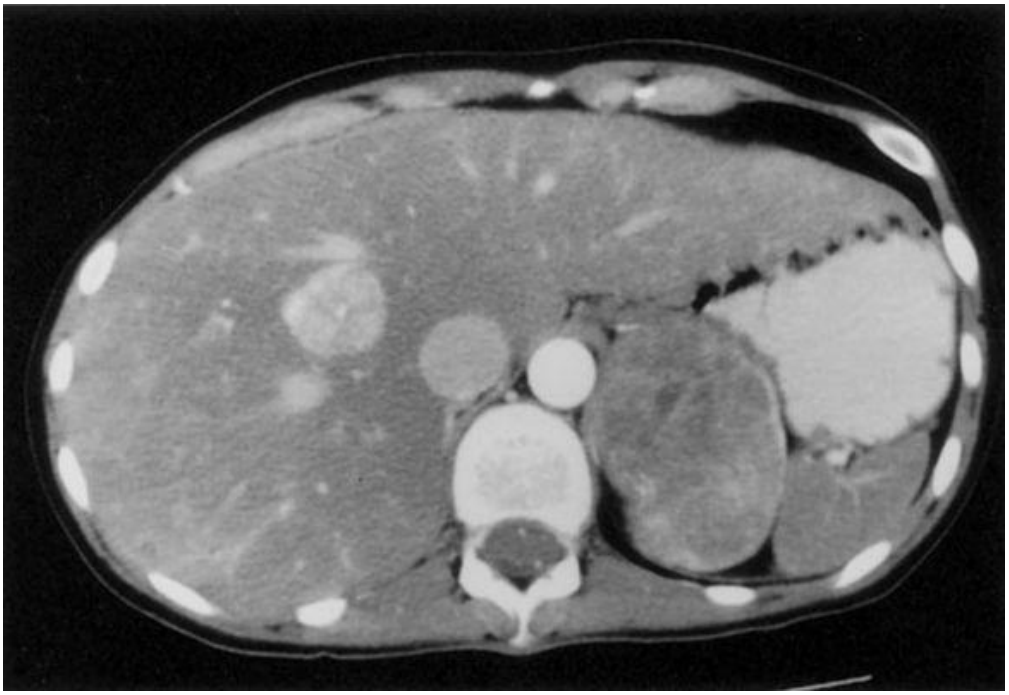
Figure 1

a



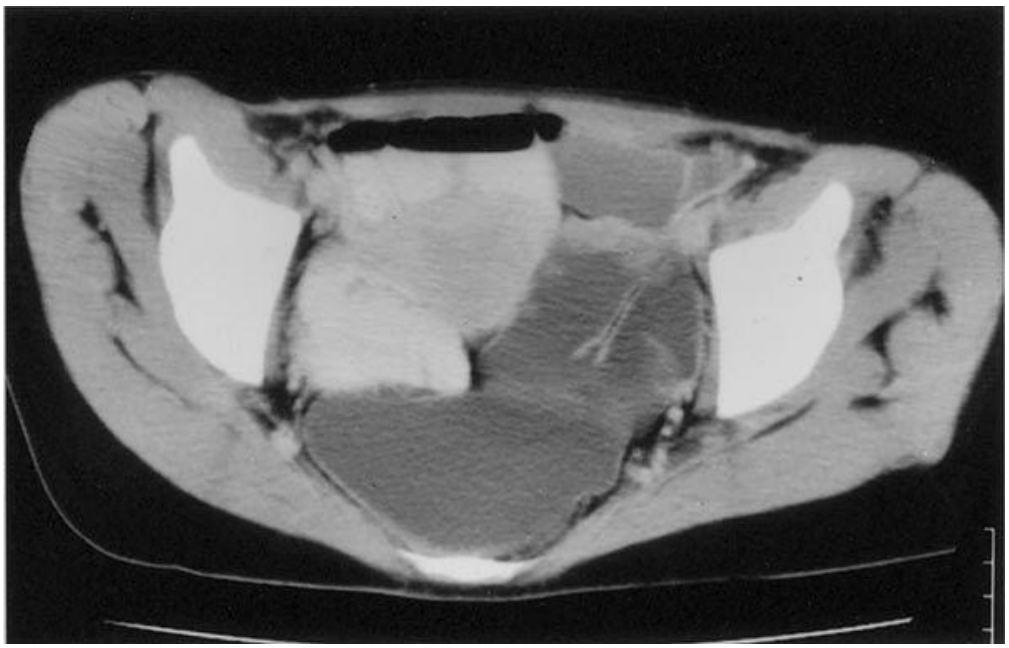
Description: Native CT scan at the level of the upper abdomen shows a large, hypodense tumor (7x5x6 cm) in the left adrenal fossa and a rounded hypodense mass (3x3x3 cm) in segment eight of the liver. **Origin:**

b



Description: Enhanced CT scan at the same level demonstrates an inhomogeneous enhancement of the adrenal tumor. The lesion in the liver shows high attenuation and a hyperdense dot in the center. **Origin:**

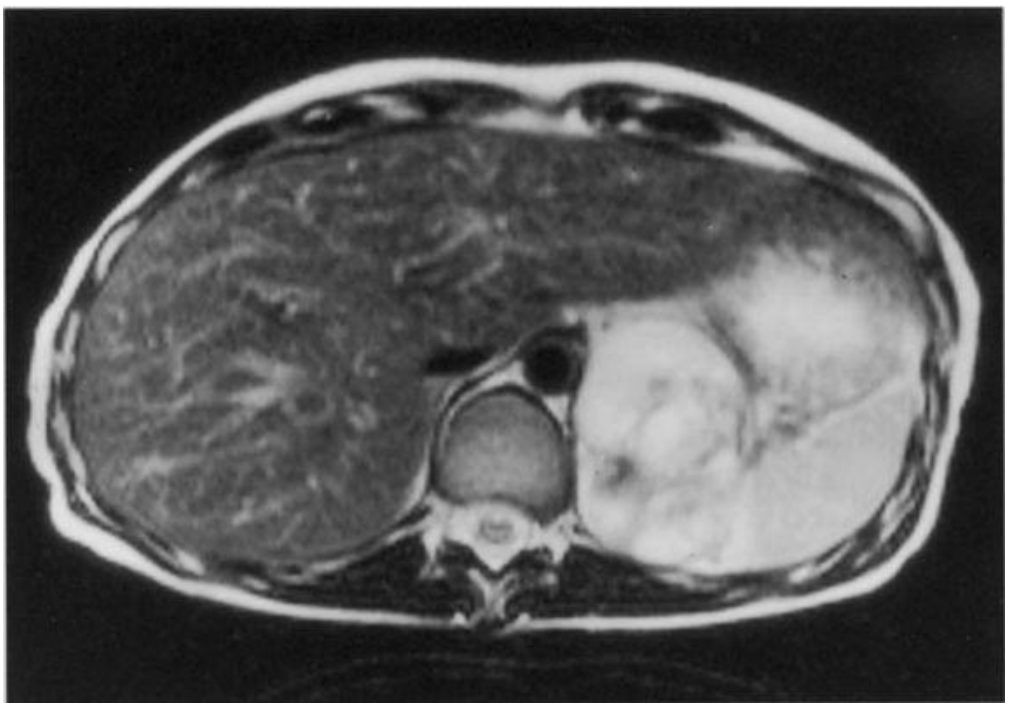
c



Description: CT scan through the pelvis reveals a distended fluid-filled rectosigmoid and other bowel loops. **Origin:**

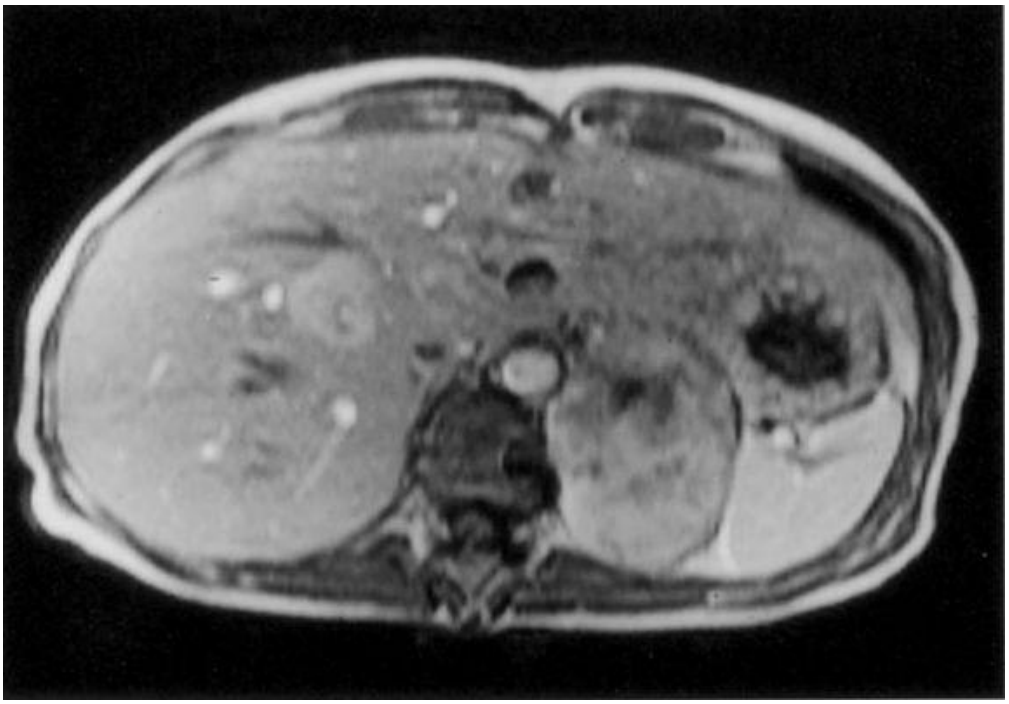
Figure 2

a



Description: Axial SE T2-weighted MR image shows an inhomogeneous adrenal tumor with mainly high signal intensity. No focal mass in the liver is identified. **Origin:**

b



Description: On axial SE T1-weighted MR image, 10 seconds after administration of gadolinium, in this early phase of a dynamic study, the liver mass shows a higher signal intensity compared to the normal liver parenchyma. **Origin:**