

ESIM 2011

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History

- 39-year old athletic and slim man, married, working as a dentist
- for a few weeks lasting pain in the upper abdomen and meteoristic complaints
- for app. 1 years soft stools, but no diarrhea
- no fever, night sweats or weight loss
- no smoking, no alcohol
- no diseases in history, no drugs
- father died of leukemia
- admitted by GP because of multiple round lesions of the liver in ultrasound examination

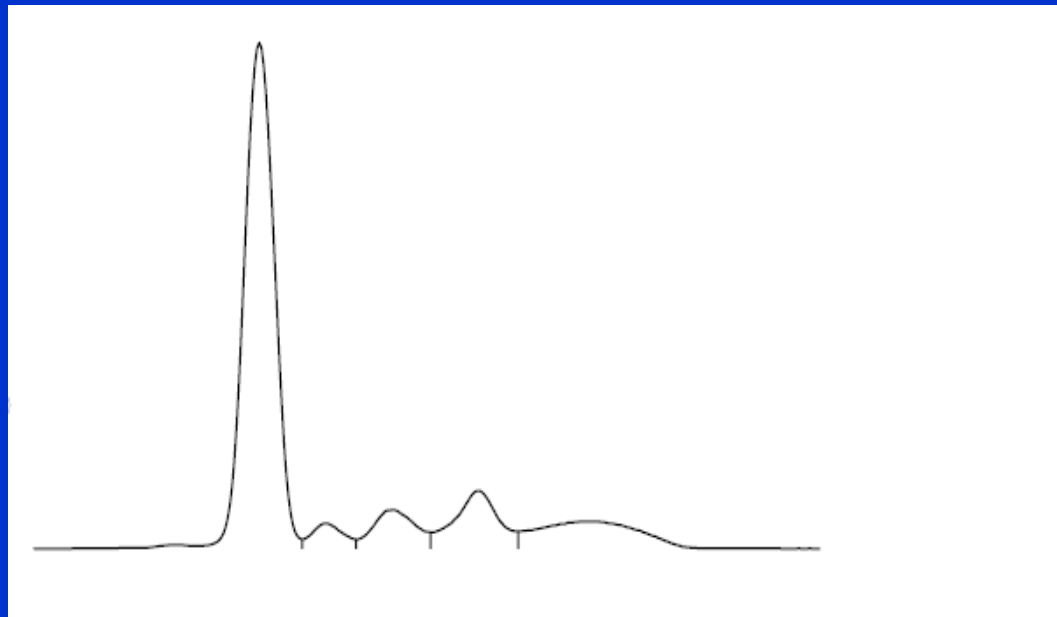
Physical Examination

- good general and nutritional condition
- blood pressure 125/90 mmHg, heart rate 64/min regular
- normal heart sounds
- clear lungs
- abdomen: soft, normal bowel sounds, tenderness of right upper abdomen, slight hepatomegaly
- no lymphadenopathy
- jugular veins not distended, no peripheral edema

Laboratory Findings

| | |
|---------------|----------------------|
| Leuk | 4.6 |
| Hb | 14.9 |
| Thro | 108 (140-345) |
| Cr | 1.1 |
| Urea | 17 |
| Na | 142 |
| K | 4.2 |
| Ca | 2.27 |
| GOT | 87 (< 38) |
| GPT | 289 (< 41) |
| g-GT | 134 (< 60) |
| aP | 102 |
| tot.prot. | 8.2 |
| alb | 4.4 |
| Lipase | 512 (13-60) |
| CRP | 0.61 (<0,5) |
| ESR | 26 (<15) |

Elektrophoresis:



MRI-Abdomen



MRI-Abdomen II



Diagnosics

- liver biopsy: Infiltration by metastases of a malignant tumor with unknown primary
- bronchoskopy, endoskopy: normal
- tumor marker: β -HCG, AFP, CEA, CA19-9, 5-HAA (Urin), Gastrin all normal, Chromogranin A 159 ng/ml (<98 ng/ml)

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Chromogranin A:

- tumor marker of functionally active und inactive neuroendocrine tumors
- high specifity, sensitivity app. 63%

- Octreotid szintigraphy: pathological uptake consistent with lump of pancreatic body/tail
- FDG-PET: no pathological uptake

Summary

- lump of pancreatic body/tail
- widespread metastases of liver
- no pathological uptake in FDG-PET
- pancreatic uptake in octreotide-szintigraphy
- elevated chromogranin A in blood
- histo-pathological and immunohistochemical findings of liver biopsy unclear



Strong suspicion of a functional inactive neuroendocrine carcinoma of the pancreas with liver metastases

Differential Diagnosis

1. Insulinoma (app. 60%)

- solitary, small, nearly always pancreatic
- 90% benign
- 5-7% MEN1
- dizziness, seizures, tremor, sweating, fear, hunger
- hypoglycemia when fasting

2. Gastrinoma

- pancreatic or duodenal, up to 25% MEN1
- mostly malign, very often metastases
- multiple ulcerations of upper gastrointestinal tract, diarrhea (60%), Cushing-Syndrom (6%)
- hypergastrinemia

Differential Diagnosis II

3. Glucagonoma

- mostly in pancreatic tail, 5-17% MEN1
- highly malignant, >80% metastases
- diabetes, weight loss, diarrhea
- erythema necrolyticum migrans (65-80%)

4. VIPoma, carcinoid, functionally inactive tumors

5. exocrine ductal adenocarcinoma, acinar cell carcinoma, small cell carcinoma, metastases

Diagnosis: Acinar cell carcinoma

- rare, app. 1% of exocrine pancreatic cancer
- usually solid, sometimes cystic
- located throughout the pancreas
- predominant age 50-70, sometimes younger
- symptoms: sometimes subcutaneous fat necrosis and polyarthralgia due to elevated lipase
- histologically staining for (chymo-)trypsin, amylase, lipase
- app. 1/3 appearance of neuroendocrine cells
- mixed acinar-endocrine tumors

Therapy/Prognosis

- surgical treatment/radical resection
- no established treatment for unresectable tumors
- radio-chemotherapy (gemcitabine)
- local therapy of liver metastases
- prognosis better than for ductal adenocarcinomas, but worse than pancreatic neuroendocrine tumors
- aggressive surgical resection with negative margins is associated with a better long-term survival
- overall survival dependent on disease stage