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A case of gastrinoma associated with ectopic Cushing’s syndrome.

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A 36-year-old woman with recurrent epigastric pain underwent laparotomy due to gastric ulcer perforation two days after labor. After surgery inflammatory markers remained elevated. In ultrasound liver tumor and abdominal abscesses were observed and patient was reoperated. Liver tumor histopathology revealed metastasis of neuroendocrine tumor (NET) G2. Thus, the patient was admitted to the Endocrinology Department.

In abdomen CT and MRI scans in the left liver lobe a tumor size 80x70x75 mm with central necrosis and pancreatic tail tumor size 41x54 mm were described, peritoneal and extraperitoneal lymph nodes were enlarged. In retroperitoneum abscesses size up to 115x60 mm were observed (Fig 1). Laboratory tests revealed increased gastrin (2888 pg/ml), chromogranin A (699 µg/l), serotonin levels (158 ng/ml) and elevated 24-hour urinary serotonin excretion (1386 µg/24 h). No additional endocrinopathies were found and gastrinoma was diagnosed. In PET/CT 18FDG slightly increased metabolism in pancreatic tail tumor, liver metastasis in segment 2, retroperitoneal lymph nodes and left supraclavicular lymph node were observed. Somatostatin receptors expression in SPECT/CT 99mTc-DOTA-TOC was found in the same locations and additionally in two more liver metastases (segments 4,5). The T3N1M1 stage IV NET was diagnosed. Because of numerous surgical complications developed before the diagnosis, clinical status, and advanced disease stage, the patient was disqualified from surgery. Systemic therapy with somatostatin analogs and also radionuclide therapy (177Lu-DOTA-TATE and 90Y-DOTA-TATE 3.7 GBq) were applied. The disease was stable.

Seven months after the diagnosis appeared clinical hypercortisolism features, hypertension, hypokalemia and diabetes requiring insulin. Cortisol and ACTH levels were increased, without typical circadian rhythms (cortisol: 30 µg/dl and 29.7 µg/dl; ACTH: 161 pg/ml and 170 pg/ml at 8.00 and 24.00, respectively), 24-hour urinary free cortisol (UFC) excretion was increased (1626 µg/24h, normal<75). Serum cortisol and UFC remained elevated after high-
dose dexamethasone suppression (28.5 µg/dl, 1532 µg/24h, respectively). ACTH and cortisol concentrations did not respond to the administration of CRH and desmopressin. In pituitary MRI no adenoma was found. Ectopic Cushing’s syndrome was confirmed. After administration of ketoconazole 600 mg daily the UFC has halved but remained elevated (293.7 µg/24h). It was not possible to increase ketoconazole dose due to liver dysfunction. At the same time, radiological disease progression was observed and second radionuclide therapy was given (177Lu-DOTA-TATE 7.4 GBq). A month later, one year after the gastrinoma diagnosis, the patient died probably because of pulmonary embolism, regardless of thromboembolic prophylaxis administration. Gastrinomas are rare (0.5–21.5 cases/10⁶/year) functional NET localized mostly in pancreas or duodenum. Occur sporadically in about 75-80% cases, or in association with multiple endocrine neoplasia type 1. The gastrin secretion stimulates excessive gastric acid production and causes Zollinger-Ellison syndrome [1]. Additional ectopic ACTH secretion by malignant gastrinoma was extremely rare described, but according to some authors may occur in up to 5% cases [2]. The established diagnosis of the tumor secretion does not exclude the occurrence of a new functional tumor activity, which affects the prognosis, significantly worsening it, and requires additional treatment (steroidogenesis inhibitors) and prophylaxis of opportunistic infections and thromboembolic events [2-5].
References:


Figure 1 a.

BLADE sequence, T2-weighted fat-saturated transverse image: 1 - Left liver lobe metastasis with features of central scarring/retrograde lesions. 2 - Subcutaneous bile reservoir.
Figure 1 b.

EPI DWI sequence b 800 s/mm²: 1 - Pancreas tumor. 2 - Enlargement of lymph nodes.
Figure 1 c.
HASTE sequence, T2-weighted coronal image. Left liver lobe metastasis.
Figure 1 d.

T1-weighted transverse image, 1.5h post intravenous contrast injection: 1 - Gallbladder fistula in intra-abdominal fat and in the abdominal muscles. 2 – The next metastasis - in the right liver lobe. 3 – Bile collection under the right armpit.