

Letters to the Editor

Benign pancreatic vipoma

Key words: VIPoma. Pancreatic tumor. Vasoactive intestinal polypeptide.

Dear Editor,

VIPoma is a neuroendocrine tumor that secretes vasoactive intestinal polypeptide (VIP) and produces a well-defined clinical syndrome characterized by watery diarrhea, hypokalemia, hypochlorhydria and metabolic acidosis. With low incidence, in 90% of cases the VIPoma arise from the pancreas. We present a new case of pancreatic VIPoma with successful resection and benign histology.

Case report

We report a case of a 33-year-old woman studied in the outpatient gastroenterology clinic due to a one year history of diarrhea of 5-6 depositions a day without pathological products in feces, accompanied of weakness, anorexia and weight loss of 15 kg. In the last 2 months, the patient recounts nausea and frequent vomits as well as weakness and progressive muscle cramps until in the last 24 hours she presents muscular weakness with inability to walk. Due to this situation the patient was admitted to the emergency department. At physical examination, the patient had a normal abdomen and neurological exploration revealed paresis of arms and legs.

Laboratory examinations of blood revealed anemia (hemoglobin 11.1 g/dL), hypokalemia (K^+ 2.4) and acidosis (pH 7.32).

Fecal culture and intestinal parasites study were negative. Gastroscopy, colonoscopy, intestinal transit and capsule enteroscopy showed no abnormalities. Hormonal determination revealed markedly elevated VIP levels (119.4 pmol/L, normal 0-30). Abdominal computed tomography showed a pancreatic head tumor of 3.2 cm and a hepatic nodule of 1.3 cm in the VII hepatic segment suggestive of metastasis.

With diagnosis of pancreatic VIPoma the patient was taken to surgery and a pylorus preserving cephalic pancreatoduodenectomy and resection of hepatic nodule were made (Fig. 1).

Histological examination confirmed a pancreatic islet cell tumor with benign histology, without lymph node invasion, and a hepatic nodule compatible with focal nodular hyperplasia. The patient remains asymptomatic and disease-free at 18 months of follow up.



Fig. 1. Surgical specimen of pylorus preserving cephalic pancreatoduodenectomy.

Discussion

VIPoma is a neuroendocrine tumor that secretes VIP described the first time by Verner and Morrison in 1958 (1). The tumor produces a well-defined syndrome characterized by watery diarrhea, hypokalemia, hypochlorhydria and metabolic acidosis. The estimated incidence is 0.2 to 0.5 per million per year (2). Ninety percent of VIPoma in adults are primary tumors of the pancreas, found primarily in the body and tail, and represents 3-8% of all endocrine pancreatic tumors. They have been described also in the colon, bronchus, adrenals and liver. In children, however, these tumors are most commonly found in the adrenal glands and sympathetic ganglia (3). These tumors are almost always solitary, with less than 5% of cases being multicentric. Approximately 50-60% of VIPomas are malignant and have metastasized at the time of diagnosis. Metastasis occurs most frequently in the liver, but it may also occur in lymph nodes, lung or kidneys (4).

The dominant symptom is profuse watery diarrhea despite fasting; stool volumes are typically profound with volumes greater than 3 L per day in 70% of cases. Diagnosis is made by determination of VIP serum levels and localization of primary tumor with imaging studies like abdominal ultrasonography, computed tomography, magnetic resonance, endoscopic ultrasonography and somatostatin receptor scintigraphy (5).

The only curative treatment is the surgical resection that will change depending on the location of the tumor, generally distal pancreatectomy for tumors of body and tail of pancreas and cephalic pancreatoduodenectomy for the located ones in the head. Sometimes a multivisceral resection can be necessary to

increase the survival that must be realized in experienced centers to obtain minimal morbidity and no mortality (6).

Systemic chemotherapy, somatostatin analogs, interferon alpha, embolization and radiofrequency can be in use in patients with unresectable tumors (5).

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