A rare cause of diarrhea: pancreatic VIPoma

A 45-year-old man presented with watery, large-volume diarrhea for 4 years and 13.6 kg (30 lb) weight loss. Multiple admissions and investigations for dehydration, including computed tomography (CT), esophagogastroduodenoscopy and colonoscopy with biopsies, and capsule endoscopy, yielded negative findings. Laboratory workup, including complete blood count, tissue transglutaminase, thyroid-stimulating hormone, cortisol, serum/urine protein electrophoresis, fecal fat, calprotectin, and erythrocyte sedimentation rate (ESR)/C-reactive protein (CRP), was normal [1]. Workup for infectious diseases was negative for parasites, *Clostridium difficile*, and human immunodeficiency virus (HIV). Stool electrolytes were consistent with secretory diarrhea based on an osmolar gap of 40 mOsm/kg. Neurohormonal testing revealed elevated chromogranin A (320 ng/mL, normal range: 0–95) and vasointestinal peptide (VIP) (360 pg/mL, normal range: 0–60), suspicious for neuroendocrine tumor producing VIP (VIPoma). Prior CT scans were inconclusive for neuroendocrine tumor, but octreotide scan showed marked accumulation of tracer in the pancreas and liver (▶ Fig. 1 a,b). Endoscopic ultrasound (EUS) revealed a hypoechoic mass measuring 25×30 mm in the pancreatic neck (▶ Fig. 1 c) and a hypoechoic 7×8 mm lesion in the right hepatic lobe. Fine needle aspiration showed mononuclear cells with granular cytoplasm and stippled chromatin, hyperchromatic nuclei, and stained positive for synaptophysin (▶ Fig. 2) consistent with a neuroendocrine tumor. The patient had symptomatic improvement with administration of octreotide, and the diagnosis of VIPoma was confirmed.

VIPoma is rare, with worldwide incidence of 1 in 10 000 000 [2], and 80% have metastasized before diagnosis [3]. Described as “pancreatic cholera,” symptoms include 3–5 L/day of watery diarrhea, hypokalemia, achlorhydria, and acidosis. Diagnosis is established via octreotide scan (sensitivity 58%–86%) and EUS (sensitivity 87%) [4]. Complete resection is curative but rarely possible due to the extent of disease at diagnosis [2,3]. Symptomatic palliation may be achieved with debulking/chemoembolization and/or administration of octreotide [2,3].

Fig. 1 a,b Computed tomography (CT) and octreotide scanning findings in pancreas suggestive of neuroendocrine tumor in a 45-year-old man presenting with watery, large-volume diarrhea for 4 years and massive weight loss. c Endosonographic findings of tumor in pancreatic neck.

Pancreatic neuroendocrine tumors are rare and can be difficult to diagnose. They should be suspected in those with longstanding secretory diarrhea. EUS has an increasingly recognized role in diagnosis and staging [3,4].

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Fig. 2 Histological features of neuroendocrine tumor. **a** Mononuclear cells with granular cytoplasm and stippled chromatin. **b** Hyperchromatic nuclei. **c** Positive synaptophysin staining.

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