Diagnosis of polypoid portal hypertensive enteropathy due to superior mesenteric vein thrombosis by capsule endoscopy and double-balloon enteroscopy

Fig. 1 Abdominal computed tomographic axial contrasted scan reveals a desmoplastic-like reaction in the jejunal mesentery (red cross), consisting of dilated superior mesenteric vein branches before a superior mesenteric vein occlusion more cranially. Multiple engorged and thick-walled jejunal loops are seen (yellow arrow).

Fig. 2 Multiple pseudopolyps in the jejunum of a patient with polypoid portal hypertensive enteropathy (small-bowel capsule endoscopy).

Fig. 3 Polypoid jejunal mucosa with lymphatic oozing (underwater view at double-balloon enteroscopy).

Fig. 4 Multiple polypoid lesions in a patient with polypoid portal hypertensive enteropathy at double-balloon enteroscopy.

A 46-year-old man with recently controlled retroviral infection was referred to our Neuroendocrine Tumour Unit for further investigation of recent onset, refractory ascites and a long-standing but stable mesenteric mass of indeterminate cause.

There was no evidence of chronic liver disease. Analysis of the ascitic fluid revealed chylous ascites. Computed tomography of the abdomen showed a 2.6-cm mesenteric mass with associated desmoplasia encasing the superior mesenteric

vein, ascites, and edema of the small bowel (● Fig. 1). Although the radiologic appearances supported the presence of a neuroendocrine tumor, the results of biomarker (chromogranin A) and molecular imaging studies (octreotide scan, fluoro-deoxyglucose positron emission tomography, and gallium Ga 68 octreotate positron emission tomography) were all negative. Small-bowel capsule endoscopy demonstrated features of segmental portal hypertension with mucosal congestion and multiple pseudopolyps (● Fig. 2) but no primary small-bowel tumor. Anterograde double-balloon enteroscopy confirmed the presence of a congested jejunal mucosa with lymphatic oozing (● Fig. 3). Multiple polypoid lesions were observed (● Fig. 4, ● Fig. 5) within parts of the jejunum, and histologic examination of one of these lesions, retrieved at double-balloon enteroscopy, showed villous thickening and the proliferation of small blood vessels with edema of the lamina propria, in keeping with polypoid portal hypertension enteropathy (● Fig. 6). Radiologic and surgical interventions (superior mesenteric vein stent insertion and portosystemic anastomosis, respectively) were not technically feasible, and the patient's refractory chylous ascites was therefore successfully managed with a trial of octreotide (Sandostatin; Novartis Pharmaceuticals, Basel, Switzerland).

Fig. 5 Multiple polypoid lesions in the jejunum of a patient with polypoid portal hypertensive enteropathy at double-balloon enteroscopy.

Fig. 6 Histologic examination of a polypoid lesion demonstrating villous thickening and proliferation of small blood vessels with edema of the lamina propria. These appearances are in keeping with portal hypertensive enteropathy.

Polypoid portal hypertensive enteropathy is a very rare entity which has previously been reported in only a few cases to date: in the duodenum in four patients [1,2] and in the jejunum in another two patients [3,4]. The causes of portal hypertension in the previously reported cases

were alcohol-related liver cirrhosis in adults [1,3,4] and extrahepatic portal venous obstruction in children [2].

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