Somatostatinoma of the minor duodenal papilla associated with pancreas divisum treated by endoscopic papillectomy

Somatostatinomas are found in the major duodenal papilla (MaDP), but their diagnosis is difficult because of their absence of symptoms and small size, and because the tumor emerges from the submucosa [1]. Somatostatinomas in the minor duodenal papilla (MiDP) are extremely rare [2]. Most MiDP tumors are asymptomatic, while those of the MaDP more often cause jaundice and pain [3].

Pancreas divisum is a common congenital anatomic variant of the pancreas and occurs when the ventral pancreatic duct (VPD) and the dorsal pancreatic duct (DPD) do not fuse during embryogenesis [4]. This disunity causes the exocrine pancreatic secretions to drain via the MiDP, increasing the pressure within the DPD. Pancreatoduodenectomy is the option of choice for curative treatment of neuroendocrine tumors (NETs), but the morbidity and mortality rates are 50% and 2%, respectively [5]. As a result treatment by endoscopic papillectomy has become more attractive owing to its lower morbidity and mortality rates. We present a rare case of somatostatinoma in the MiDP, associated with pancreas divisum, which was treated by endoscopic papillectomy without the insertion of a pancreatic stent even though there was pancreas divisum.

A 60-year-old woman presented with epigastric pain, and esophagogastroduodenoscopy (EGD) showed bulging of the MiDP. A biopsy was taken; histology of this specimen revealed a NET (grade I according to the World Health Organization [WHO] classification). Her amylase level was 92 IU/dL; the results of chromogranin A and all other standard tests were normal. Computed tomography (CT) scanning showed the prominent MiDP, associated with pancreas divisum, which was treated by endoscopic papillectomy without the insertion of a pancreatic stent even though there was pancreas divisum.

Video 1

Imaging showing pancreas divisum (PD) with predominance of the dorsal pancreatic duct (DPD) over the ventral pancreatic duct (VPD) and a somatostatinoma of the minor duodenal papilla (MiDP) that was resected en bloc by endoscopic papillectomy (EP).

NET, neuroendocrine tumor; WHO, World Health Organization; MRCP, magnetic resonance cholangiopancreatography; CT, computed tomography; CBD, common bile duct; EUS, endoscopic ultrasound; FICE, Fuji Intelligent Chromoendoscopy.
showed a rounded hypoechoic nodule of more than 2.0 cm in size, which was restricted to the MiDP with no invasion of the DPD, which was 35mm in diameter, and without communication with the VPD (Fig. 3).

Endoscopic papillectomy removed the tumor en bloc and insertion of a pancreatic stent was not required because of the dilatation of the DPD (Fig. 4). Hematochezia occurred 28 hours after the resection and was treated with clips and injection of 1:10 adrenaline and glucose (Fig. 5). The patient progressed well and was discharged 6 days after the endoscopic papillectomy.

Histology showed a grade I somatostatinoma (WHO classification) with angio-lymphatic infiltration and the following immunohistochemistry results: somatostatin (+), neuron-specific enolase (+), synaptophysin (+), chromogranin (+), and a Ki-67/mitotic proliferation index of <2%.

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