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 and that the DPD was predominant, but there was no evidence of metastases (Fig. 1) and this was confirmed by magnetic resonance imaging (Fig. 2). Endoscopic ultrasound (EUS), performed in planning for the endoscopic papillectomy,
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 had a diameter of 35 mm; the dominance of the DPD in the head of the pancreas; the dominance of the DPD in the body of the pancreas; the peridiverticular major duodenal papilla (DIV).

Endoscopic ultrasound (EUS) images showing: a hypoechoic homogeneous nodule with clear limits (1.3 × 0.8 cm) without invasion of the dorsal pancreatic duct (DPD), which had a diameter of 35 mm; the dominance of the DPD in the head of the pancreas; the dominance of the DPD in the body of the pancreas; the peridiverticular major duodenal papilla (DIV).

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

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