Invaginated ampulla of Vater in synchronous malignant intraductal papillary mucinous neoplasm of the pancreas and common bile duct cancer

A 61-year-old woman was admitted for upper abdominal pain and fever. Laboratory findings showed raised neutrophil ratio and amylase and lipase levels. Abdominal computed tomography (CT) scan revealed diffuse swelling of the pancreas with peripancreatic fluid, and mild dilatation of the mid-pancreatic duct. Endoscopic retrograde cholangiopancreatography (ERCP) showed dimpling of the ampullary orifice (Fig. 1). We inserted an endoscopic retrograde pancreatic drain (ERPD) via a guide wire. A pancreatogram showed pancreatic duct dilatation and deformity (Fig. 2) and due to stricture of the pancreatic duct orifice, the ERPD required repositioning. During removal of the ERPD for repositioning, the ampullary orifice, which was invaginated, protruded into the duodenal lumen (Fig. 3). A biopsy specimen was taken from the invaginated ampullary orifice, and confirmed to be adenocarcinoma. The patient underwent pylorus-preserving pancreaticoduodenectomy. The post-operative pathology report confirmed the diagnosis of a double primary tumor: the first tumor being a malignant intraductal papillary mucinous neoplasm (IPMN) of the pancreas and the other an adenocarcinoma of the common bile duct (CBD). Histologic exam documented anomalous union of pancreaticobiliary duct with short segment common channel and separated incidental IPMN of main duct in pancreas head. Protruded mass of ampulla of Vater was confirmed as CBD originated. The ampulla of Vater appears as a hemispherical or oval elevation [1], although presentation varies among individuals. A typology for the shape of ampulla of Vater has not yet been established, but Horiuchi et al. [2] have classified the exophytic type of ampulla of Vater based on shape into “small, large, or swollen.” We did not determine the effect of the distal CBD cancer on the invaginated ampulla. Katsinelos et al. [3] reported a case of carcinoma with site compression at the ampulla of Vater; the cancer had developed in the center with elevated edges. Yoon et al. [4] evaluated mucosal tumor infiltration into the CBD and pancreatic duct in early ampullary cancer and also classified the polypoid or ulcerative lesions. But the particular structural deformation (invagination) seen in our case has not been previously reported. Additionally, recent reports suggest that patients with intraductal papillary mucinous neoplasms are at higher risk of synchronous or metachronous primary cancers arising from various organs. Nevertheless, association with CBD cancer is very rare [5].

Competing interests: None

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