Intraductal papillary mucinous neoplasm of the Santorini duct in a patient with pancreas divisum diagnosed by transpapillary biopsy

A 76-year-old patient presented with pancreatic duct dilatation on magnetic resonance imaging (MRI). The patient had no symptoms and no previous history of hepatobiliary or pancreatic disease. Endosonography showed pancreatic duct dilatation (12 mm) in the pancreatic body and tail. The pancreatic duct in the pancreatic head was only moderately dilated (5 mm). Endoscopic retrograde cholangiopancreatography (ERCP) showed a normal major papilla and normal biliary system. Pancreatography revealed a blind-ending main pancreatic duct of only 35 mm in length. The minor papilla was found in the typical position, 2 cm above the major papilla. Pancreatography confirmed the dilated main pancreatic duct in the pancreatic body and tail, with a narrow Santorini duct (Fig. 1a), so a plastic drain was inserted. Chronic pancreatitis because of pancreas divisum was assumed to be the reason for the duct dilatation.
During follow-up ERCP 10 weeks later, pancreatography showed the typical sign of mucus in the Santorini duct, with shallow contrast passage (▶ Fig. 1 b). After the extraction of a large amount of mucus, a mucinous tumor measuring 14 mm was detected in the Santorini duct (▶ Fig. 1 c). Transpapillary biopsy was performed using a one-sided opening-cup biopsy forceps (Olympus Corporation, Tokyo, Japan) and pathological results showed an intrapapillary mucinous neoplasm (IPMN). Because of the location of the neoplasm within the Santorini duct in a pancreas divisum, this was classified as a main duct IPMN (▶ Video 1).

In line with the revised Fukuoka guidelines [1], surgery was considered; however, after discussion with the patient, surgery was not pursued owing to the patient’s cardiological co-morbidities. Further surveillance was performed by direct pancreatoscopy (SpyGlass DS System, Boston Scientific Corporation, Marlborough, Massachusetts, USA) and the typical papillary structure of an IPMN was seen, with re-biopsy performed (▶ Fig. 2).

IPMN of the Santorini duct is a rare finding [2]. This case outlines the difficulty of differentiating between chronic pancreatitis and carcinoma of the pancreas and its precursor lesions.

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Competing interests
The authors declare that they have no conflict of interest.

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