Primary signet-ring cell carcinoma of the pancreas diagnosed by endoscopic retrograde pancreatic duct biopsy: a case report with an immunohistochemical study

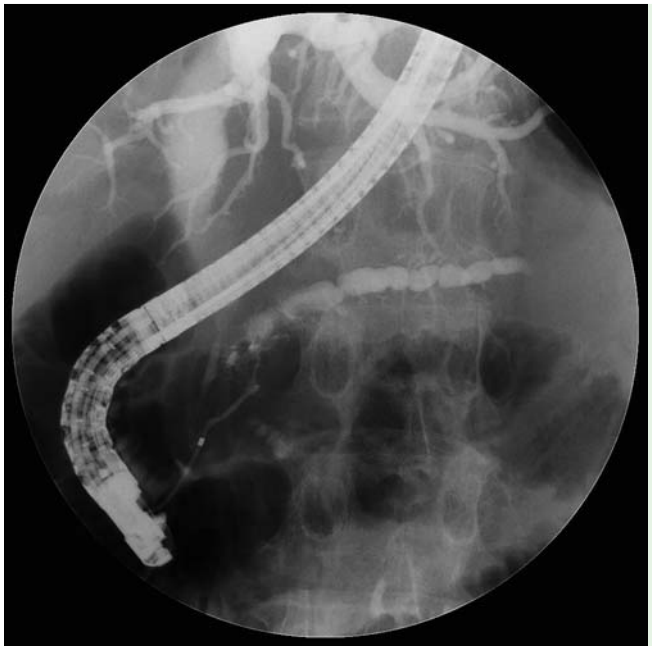

Fig. 1 Endoscopic retrograde cholangiopancreatography in a patient with primary signet-ring cell carcinoma of the pancreas. The main pancreatic ducts and common bile duct in the pancreatic head portion show irregular occluded contours.

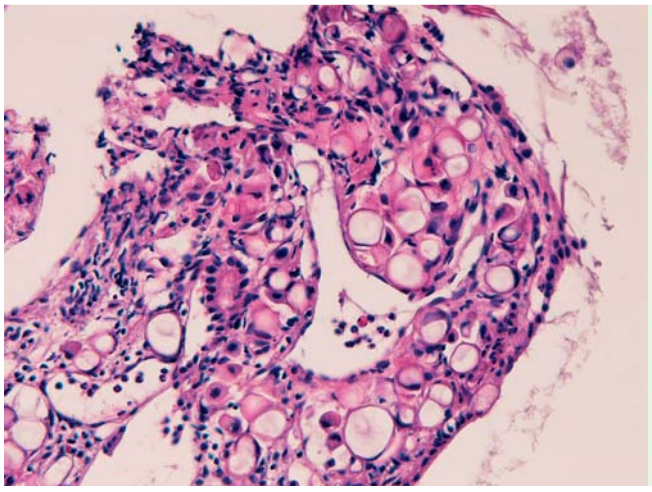

Fig. 2 Pancreatic biopsy taken during endoscopic retrograde cholangiopancreatography. The biopsy shows pancreatic signet-ring cell carcinoma. Hematoxylin and eosin, × 100.

Primary signet-ring cell carcinoma (SRCC) of the pancreas is extremely rare [1]; only three cases have been reported in the literature [2–4].

A 61-year-old man presented with abdominal pain. Laboratory blood testing revealed hyperamylasemia (626 IU/L). Tumor marker concentrations were within normal ranges. Upper gastrointestinal endoscopy showed no significant changes, as did CT including the pancreas. Pancreatitis was suspected and the patient underwent endoscopic retrograde cholangiopancreatography (ERCP) and endoscopic retrograde pancreatic duct biopsy. The ERCP showed irregular occluded pancreatic ducts at the pancreatic head (**Fig. 1**). The pancreatic duct biopsy revealed SRCC (**Fig. 2**).

The carcinoma cells were positive for mucins on alcian blue/periodic acid–Schiff staining. An immunohistochemical study was performed using Dako's EnVision method, as previously described [5,6]. The SRCC cells were positive for cytokeratin AE1/3, cytokeratin CAM5.2, cytokeratin 7 (CK7), cytokeratin 8 (CK8), cytokeratin 18 (CK18) (**Fig. 3a**), cytokeratin 19 (CK19), carcinoembryonic antigen (CEA) (**Fig. 3b**), epithelial membrane antigen (EMA), p53, Ki67 (labeling = 15 %), mucin 1 (MUC1), and mucin 5AC (MUC5AC) (**Fig. 3c**).

They were negative for cytokeratin 34BE12, cytokeratin 5/6 (CK5/6), cytokeratin 14 (CK14), cytokeratin 20 (CK20), p63, carbohydrate antigen 19–9 (CA19–9), vimentin, thyroid transcription factor 1 (TTF-1), homeobox protein CDX-2 (CDX2), mucin 2 (MUC2), and mucin 6 (MUC6). The patient underwent pancreatoduodenectomy at another hospital. The resected surgical specimen showed infiltrating SRCC at the pancreatic head. The patient is now alive 6 months after the operation.

The present case is the first one of primary SRCC diagnosed by endoscopic retrograde biopsy of the pancreatic duct. Thus, this procedure may be stated to be useful in the diagnosis of pancreatic SRCC when tumor cells have penetrated the lumen of the pancreatic duct. The present case is also the first one in which the immunoprofile of pancreatic SRCC was extensively examined. The cytokeratin profile has been demonstrated. The expression of CK7, CK8, CK18, and CK19 – pancreatic cytokeratins – is compatible with the cytokeratin profile of pancreatic ductal carcinoma [7,8]. The cytokeratin profile indicates that high-molecular-weight cytokeratins (CK34BE12 and CK5/6), CK14, and CK20 were absent from pancreatic SRCC. Pancytokeratins (AE1/3 and CAM5.2) and EMA were present. The positive p53 result and relatively high Ki67 labeling index show p53 mutations and high proliferative activity. The positive expression of CEA indicates the malignant nature of the tumor [2]. However, the serum CEA level was normal. The MUC apomucin profile is compatible with the pancreatic duct MUC mucin profile [7,8]. The negative results for expression of p63, CA19–9, vimentin, TTF-1, and CDX2 indicate that these substances are absent in pancreatic SRCC.

In conclusion, the author has presented an extremely rare case of primary pancreatic SRCC diagnosed by endoscopic retrograde biopsy of the pancreatic duct. In addition, he performed an extensive immunohistochemical study of primary pancreatic SRCC.

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Competing interests: None

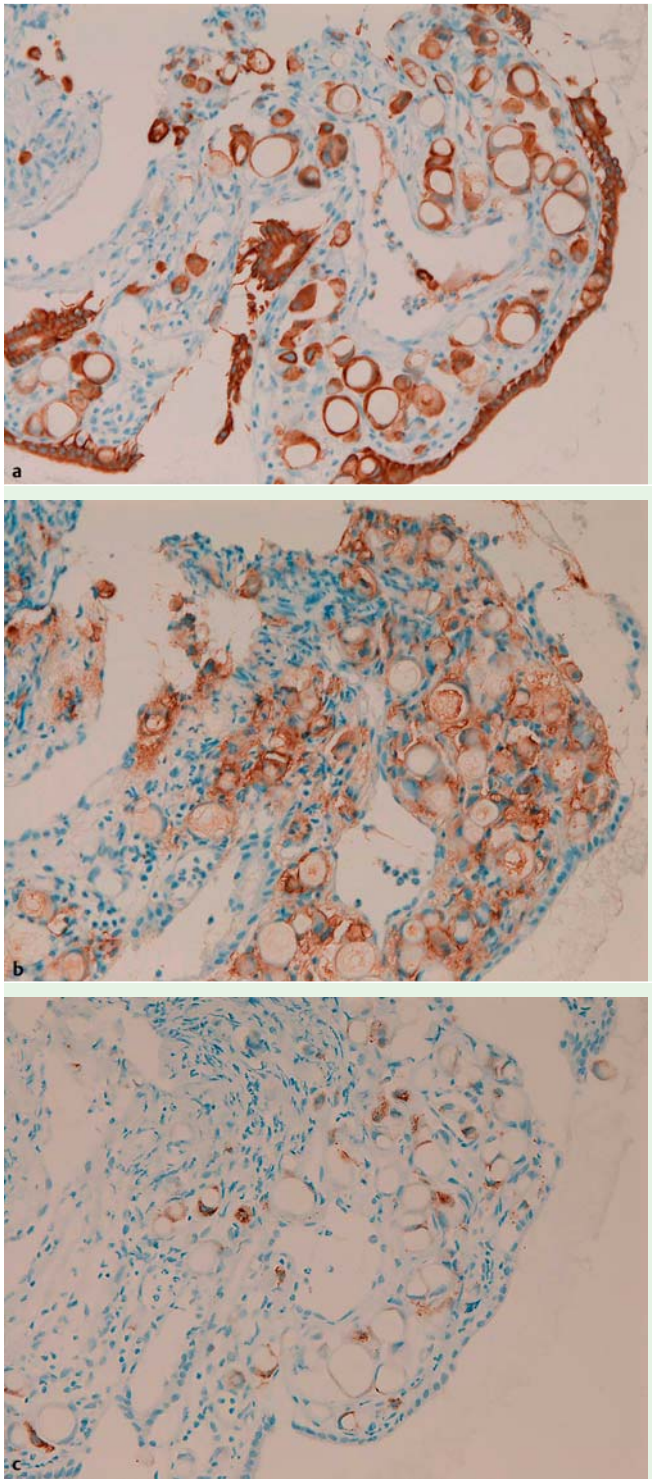

Fig. 3 The pancreatic signet-ring cell carcinoma cells stain positive for cytokeratin 18 (**a**), carcinoembryonic antigen (**b**), and mucin 5AC (**c**). Immunostaining, × 100.

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Bibliography

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