Pancreatology xxx (2016) 1–5

Contents lists available at ScienceDirect

Pancreatology

journal homepage: www.elsevier.com/locate/pan

Case report

A distinctive myoepithelial hamartoma of the pancreas histologically confirmed in the mother of a previously reported patient

Takashi Murakami [a, *], Masato Yamazaki [a], Kazuto Yamazaki [b], Kenichi Matsuo [a], Atsushi Hirano [a], Yukihiko Hiroshima [a], Daisuke Kawaguchi [a], Yasuo Ishida [b], Yutaka Suzuki [c], Masanori Sugiyama [c], Keiji Koda [a], Kuniya Tanaka [a]

[a] Department of Surgery, Teikyo University Chiba Medical Center, 3426-3, Anesaki, Ichihara, Chiba, 299-0011, Japan
[b] Department of Pathology, Teikyo University Chiba Medical Center, 3426-3, Anesaki, Ichihara, Chiba, 299-0011, Japan
[c] Department of Surgery, Kyorin University Hospital, 6-20-2, Shinkawa, Mitaka, Tokyo, 181-8611, Japan

ARTICLE INFO

Article history:
Available online xxx

Keywords:
Congenital abnormality
Hamartoma
Pancreas
Pancreatitis
Pancreatoduodenectomy

ABSTRACT

We encountered a 62-year-old female patient with a distinctive pancreatic myoepithelial hamartoma characterized by dilated loops formed by pancreatic branch ducts. The patient, who experienced recurrent acute pancreatitis caused by pancreatic juice stasis, underwent subtotal stomach-preserving pancreatoduodenectomy, achieving remission of pancreatitis. Computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) demonstrated a honeycomb appearance of the pancreatic head, consisting largely of loop-forming dilated pancreatic branch ducts. Radiography of resected specimens demonstrated a tortuous main pancreatic duct that narrowed in the head, but communicated with the pancreatic branch ducts forming intricate loops within the lesion. Histologic examination showed dilated pancreatic ducts embedded in thick layers of smooth muscle, leading to a diagnosis of myoepithelial hamartoma. Her son shared a similar clinical course, radiologic findings, and histopathologic findings with his mother. MRCP demonstrated a honeycomb appearance of the pancreatic head in her daughter, who complained of persistent diarrhea. To our knowledge, this is the first English-language reports of such a myoepithelial hamartoma of the pancreas showing familial occurrence.

Introduction

Myoepithelial hamartoma (MEH) of the pancreas is a developmental abnormality thought to result from displacement of the pancreatic anlage during embryogenesis [1]. When a displaced anlage differentiates into normal pancreatic tissue with acini and mucous glands, the aberrant structure usually is termed heterotopic pancreas. Less differentiated forms consisting predominantly of pancreatobiliary ducts surrounded by smooth muscle have been assigned various names, including adenomyosis, foregut choristomas, or MEH [2,3]. MEH occurs most commonly in the duodenum, stomach, and small intestine, while a mass-forming bulky lesion in the pancreatic head is extremely rare. Here we report a patient affected by MEH involving the pancreatic head. Pancreatography showed distinctive loops formed by dilated pancreatic branch ducts. Familial occurrence suggested an underlying genetic disorder.

Case report

A 62-year-old woman without a history of alcohol intake was treated conservatively for recurrent attacks of pancreatitis beginning at the age of 32 years. In May 2013 she was referred to our hospital for surgical treatment. A tender mass was palpable in the right upper abdominal quadrant. Abnormalities in serum included elevated alkaline phosphatase (341 U/L) and amylase (226 U/L). Computed tomography (CT) disclosed a cystic mass in the head of the pancreas that measured 8 cm in maximum diameter and contained multiple opacities (Fig. 1A). Magnetic resonance

* Corresponding author. Department of Surgery, Teikyo University Chiba Medical Center, 3426-3, Anesaki, Ichihara, Chiba, 299-0011, Japan. Tel.: +81 436 62 1211; fax: +81 436 61 3961.
E-mail address: impressor@hotmail.co.jp (T. Murakami).

http://dx.doi.org/10.1016/j.pan.2015.12.178

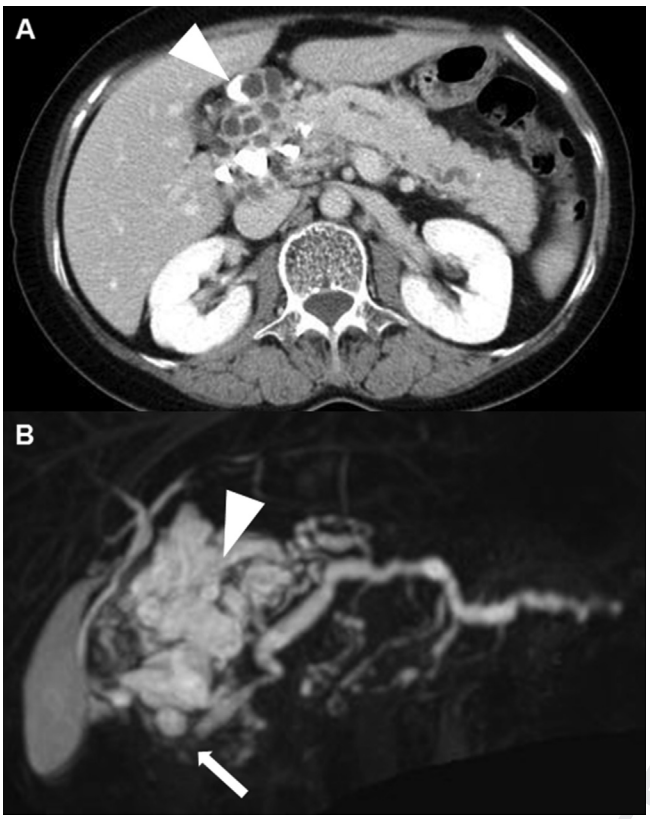

Fig. 1. CT shows a cystic mass in the head of the pancreas measuring 8 cm in maximum diameter that contains multiple opacities (A, arrowhead). The main pancreatic duct in the pancreatic body is dilated. MRCP shows a mass composed of loops formed by dilated pancreatic ducts (B, arrowhead). The main pancreatic duct shows stenosis in the head of the pancreas (arrow).

cholangiopancreatography (MRCP) visualized a mass consisting of intricate loops formed by dilated pancreatic ducts in the head and body of the pancreas (Fig. 1B). The dilated, tortuous main pancreatic duct showed focal stenosis in the head of the pancreas. The patient was considered to have a pancreatic malformation but was not diagnosed definitely. Nonetheless, the patient's recurrent pancreatitis was attributable to the tortuous pancreatic duct, so, subtotal stomach-preserving pancreatoduodenectomy was performed. On gross examination of the resected specimen, the lesion measured 7.0 × 5.7 × 4.0 cm and occupied the pancreatic head, extending into the duodenal wall (Fig. 2A). The pancreatic branch ducts were greatly dilated, imparting a honeycomb appearance. Overall, the border between the lesion and non-lesional pancreatic tissue was well defined, except where obscured focally by a chronic inflammatory reaction (Fig. 2B). Specimen radiography with contrast injection via the main pancreatic duct from the pancreatic stump side demonstrated narrowing of the tortuous main pancreatic duct in the head of the pancreas, but the duct communicated with the pancreatic branch ducts forming intricate loops within the lesion (Fig. 2C). The pancreatobiliary junction and biliary tract showed no abnormality. Microscopically, the lesion consisted mainly of pancreatic ducts encircled by markedly thickened smooth muscle bundles (Fig. 3). The surrounding pancreatic parenchyma showed inflammatory changes and fibrosis, without evidence of neoplasia. The patient was diagnosed with MEH of the pancreas. The postoperative course was uneventful, and pancreatitis has not recurred since surgery.

The patient's family tree is shown in Fig. 4. Like his mother, the son of the present patient had no history of excessive alcohol intake. His clinicopathologic findings were reported previously by Sugiyama et al. [4] He had undergone pancreatoduodenectomy at age 23 for treatment of recurrent pancreatitis associated with pancreatic juice stasis. Findings of CT, MRCP, pancreatography of the resected specimen, and histopathologic examination demonstrated a hamartomatous lesion in which atrophic pancreatic lobules contained dilated ducts surrounded by fibromuscular stroma. These findings closely resembled those in his mother. The daughter of the present patient underwent evaluation of diarrhea that had persisted for a few years. In October 2015, at age 30, she underwent MRCP (Fig. 5). Multiple loops formed by dilated pancreatic ducts were demonstrated in the head of the pancreas; the findings were similar to those in her mother and brother. Oral pancreatic enzyme replacement has alleviated her symptoms. Considering the risk of acute pancreatitis, the daughter continues to undergo frequent follow-up examinations. The mother, mother-in-law, and father-in-law of our present patient died of gastric cancer. The present patient and her children have not agreed to undergo genetic testing so far, since other family members have not been affected by any pancreatic or congenital disease.

Discussion

Here we report a unique familial form of MEH characterized by dilated loops formed by pancreatic branch ducts. Pathologic diagnosis of MEH was based on abnormally arranged non-neoplastic pancreatic tissues consisting of thickened smooth muscle surrounding looped ductal structures. Islets were absent in the lesions. MEH of the pancreas first was reported by Kwon et al.; their case did not exhibit notably dilated pancreatic ducts [3]. We know of no previous English-language reports of MEH showing dilated loops formed by pancreatic branch ducts.

The radiographic differential diagnosis in our present cases included certain cystic pancreatic lesions such as solid pseudopapillary neoplasm, serous cystic neoplasm (SCN), and intraductal papillary mucinous neoplasm (IPMN) [5–8]. An otherwise solid pseudopapillary neoplasm can include multiple cysts, and a microcystic or mixed-type SCN can exhibit a honeycomb appearance [9,10]. However, these focally or largely cystic neoplasms show no communication with pancreatic ducts, in distinction to our patients' abnormally dilated pancreatic ducts shown to communicate with the main pancreatic duct. Although our patients' lesion resembles branch duct or mixed-type IPMN in terms of dilation of branch ducts, IPMN does not show loop formation by these ducts.

Recurrent acute pancreatitis in the present patient and her son likely arose from stasis of pancreatic juice within tortuous, dilated pancreatic branch ducts caused by focal stenoses of these ducts or from stenosis of the main pancreatic duct [11,12]. Since MEH represents non-neoplastic disease, pancreatoduodenectomy should be avoided if possible considering the highly invasive nature and possible undesired effects of the procedure. However, if recurrent acute pancreatitis cannot be controlled by medical treatment, pancreatoduodenectomy should be considered. The daughter in this family, who has experienced only diarrhea related to exocrine dysfunction, nonetheless will be followed up closely with respect to possible future occurrence of pancreatitis.

Familial occurrence of MEH of the present patient and her children suggests an underlying genetic disorder. No other family members appear to be affected by hamartomas in any organ (Fig. 4). PTEN hamartoma tumor syndrome, an overall term that includes Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome, Proteus syndrome, and Proteus-like syndrome, is a genetic disease causing hamartomas in various organs [13,14]. All of these conditions result from germline mutation of the *PTEN* tumor suppressor gene. Cowden syndrome, characterized by hamartomatous lesions of the

Please cite this article in press as: Murakami T, et al., A distinctive myoepithelial hamartoma of the pancreas histologically confirmed in the mother of a previously reported patient, Pancreatology (2016), http://dx.doi.org/10.1016/j.pan.2015.12.178