Mucinous cystic neoplasm of the pancreas in a male patient

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Abstract

Mucinous cystic neoplasms (MCNs) make up a morphologic family of similar appearing tumors arising in the ovary and various extra-ovarian organs such as pancreas, hepatobiliary tract and mesentery. MCNs of the pancreas occur almost exclusively in women. Here, we report a rare case of MCN in a male patient. A 39-year-old man was admitted to our hospital with the chief complaint of back pain. Abdominal computed tomography revealed a multilocular cystic mass 6.3 cm in diameter in the pancreatic tail. In addition, the outer wall and septa with calcification were demonstrated in the cystic lesion. On magnetic resonance imaging, the cystic fluid had low intensity on T1-weighted imaging and high intensity on T2-weighted imaging. Endoscopic retrograde cholangio-pancreatography (ERCP) showed neither communication between the cystic lesion and the main pancreatic duct nor encasement of the main pancreatic duct. Endoscopic ultrasonography revealed neither solid component nor thickness of the septa in the cystic lesion. Consequently, we performed distal pancreatectomy with splenectomy under the diagnosis of cystic neoplasia of the pancreas. Histopathologically, the cystic lesion showed two distinct components: an inner epithelial layer and an outer densely cellular ovarian-type stromal layer. Based on these findings, the cystic lesion was diagnosed as MCN.

Introduction

Mucinous cystic neoplasms (MCNs) have been defined as large, separated, thick-walled cysts without connection to the pancreas duct system. Based on the WHO criteria in 1996, the study for 130 cases of MCN with ovarian type stroma (OS) indicated to be female patients appearances in whole cases and bodytail location in almost. Therefore, as a rough rule for pancreas cystic neoplasms, in male and in the head of the pancreas are likely to be IPMNs, whereas cystic lesions in the bodytail in female may be either an MCN or IPMN. And, in the past several years, mucinous cystic neoplasms of the pancreas have been diagnosed more and more frequently. Then it has become crucial for physicians working in this field to have a clear understanding of the biology on these tumors. From recent our experience for a male case with MCN, which is quite rare, diagnostic evaluation and therapeutic procedures including surgical indication will be argued with the references.

Case Report

A 39-year-old man was admitted to our hospital with the chief complaint of back pain. There was no history of previous abdominal imaging examinations, and another abdominal episode, such as sudden abdominal pain, abdominal trauma, or abdominal operation. He was just social drinker of alcohol and was not a smoker. On physical examination, abdomen was soft and flat, and no tenderness was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed. Laboratory tests showed an elevation of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), of both AST, up to 39 IU/L (normal ≤ 55 IU/L), and ALT, up to 56 IU/L (normal ≤ 35 IU/L), was noticed.

Magnetic resonance imaging showed that the cystic lesion was hypointense in T1-weighted imaging (a) and hyperintense in T2-weighted imaging with low intense capsule and septum (b).
Mucin-producing cystic neoplasms of the pancreas have developed a well-recognized entity. In the two decades, due to the utility of high-resolution abdominal imaging techniques, similar cystic lesions of the pancreas are increasingly identified incidentally, and a large number of patients have undergone surgical resection. In 1996, under the aim to describe and categorize the cystic lesions of the pancreas, the WHO classification defined MCN as cystic epithelial neoplasms composed of columnar, mucin-producing epithelium, supported by OS. The OS is known for forming a band of densely packed spindle cells beneath the epithelium; its presence has become a critical requirement as MCN. Then, the MCN was estimated for the different concept from IPMN in the past categories for cystic lesion. And, the Armed Forces Institute of Pathology (AFIP) classification also added the finding for no communication with the pancreatic ductal system. Taken together, no doubt to diagnose the present cystic lesion as MCN was detected. Although the developmental process of MCN has not been well understood, it is indicated to originate from remnant primordial gonadal cells that migrated to the pancreas, because the left primordial gonad and dorsal pancreas anlage lie side by side during embryogenesis. The dorsal anlage develops the body and tail of the pancreas; therefore the MCN was frequently raised in the dorsal pancreas and detected in female. Indeed, from the past reports for 130 cases and 56 cases with MCN, no male case has been demonstrated. According to the recent research for pancreas cystic tumors by the Japan Pancreas Society (1992-2001), detected 179 cases with MCN were female in whole. To diagnose the cystic lesion for MCN, the presence of OS pattern should be important; if limiting the tumor with OS but not without OS, all male cases were excluded. Therefore, it is likely that many of the MCN cases reported in male in the early literature were IPMN or other cystic lesion. To the best of our knowledge, 2 male cases with WHO criteria-mediated MCN have been reported for a 43-year-old man in 1998 and a 25-year-old man in 2005. Both of them, the cystic lesions were located in pancreas tail, 2.5 cm and 5 cm in size, and the expressions of estrogen/progesterone receptor were demonstrated in stromal component immunohistochemically. By contrast, the present MCN was found to have neither estrogen nor progesterone receptors. These rare patterns become the originality of MCNs to be unclear, and then further investigations should be continued.

Similar to IPMNs, MCNs have been divided into those with low-grade, moderate grade and high grade dysplasia, and invasive adenocarcinoma. The prevalence of invasive carcinoma reported in MCNs has varied widely in the range of 6-36%. The MCNs are commonly solitary and the recurrence does not occur after complete resection. The biological and clinical behavior of invasive adenocarcinoma arising from the background of cystic neoplasms might be different from the common invasive ductal adenocarcinoma. Infiltrating carcinoma associated with MCNs, and also IPMNs, appears to grow less aggressively and to have a lower incidence of peritoneal and vascular invasion. Then, the patients suffering from malignant cystic neoplasms of the pancreas are likely to get a better for survival after resection than patients with ductal adenocarcinoma. The patients with ductal pancreatic cancer show around 20% for 5-years survival even after curative resection, while the patients with malignant MCNs demonstrate over 50% for 5-years survival rate. According to the Japan Pancreas Society also, the 5-year survival rate of MCN patients was 100% in adenoma to minimally invasive carcinoma cases, and 57.2% in invasive carcinoma cases. In spite of low grade malignant potential, a common recommendation should be removal for all mucin-producing neoplasms because of the fear of ultimately developing cancer. For the surgical procedures, the consideration about not only operative indications but also care for safety might be argued.

References

1. Compagno J, Oertel JE. Mucinous cystic neoplasms of the pancreas with overt and latent malignancy (cystadenocarcinoma and cystadenoma). A clinicopathologic study of 41 cases. Am J Clin Pathol 1978;69:573-80.
2. Thompson LD, Becker RC, Przygodzki RM, et al. Mucinous cystic neoplasm (mucinous cystadenocarcinoma of low-grade malignant potential) of the pancreas: a clinicopathologic study of 130 cases. Am J Surg Pathol 1999;23:1-16.
3. Tanaka M, Chari S, Adsay V, et al. International consensus guidelines for management of intraductal papillary mucinous neoplasms and mucinous cystic neoplasms of the pancreas. Panreatology 2006;6:17-32.

4. Fernández-del Castillo C, Targarona J, Thayer SP, et al. Incidental pancreatic cysts; Clinicopathological characteristics and comparison with symptomatic patients. Arch Surg 2003;138:427-33.

5. Balcom JH, Rattner DW, Warshow AL, et al. Ten-years experience with 733 pancreatic resections: Changing indications, older patients, and decreasing length of hospitalization. Arch Surg 2001;136:391-8.

6. Zamboni G, Klöppel G, Hruban RH, et al. World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of the Digestive System. Lyon: IARC PRESS; 2000. p. 234-6.

7. Solcia E, Capella C, Klöppel G. Tumors of the pancreas. Armed Forces Institute of Pathology, Washington DC. 1997. p. 41-53.

8. Zamboni G, Scarpa A, Bogina G, et al. Mucinous cystic tumors of the pancreas: Clinicopathological features, prognosis, and relationship to other mucinous cystic tumors. Am J Surg Pathol 1999;23:410-22.

9. Suzuki Y, Atomi Y, Sugiyama M, et al. Cystic neoplasm of the pancreas: a Japanese multiinstitutional study of intraductal papillary mucinous tumor and mucinous cystic tumor. Pancreas 2004;28:241-6.

10. Wouters K, Ectors N, Van-Steenbergen W, et al. A pancreatic mucinous cystadenoma in a man with mesenchymal stroma, expressing oestrogen and progesterone receptors. Virchows Arch 1998;432:187-9.

11. Suzuki M, Fujita N, Onodera H, et al. Mucinous cystic neoplasm in a young male patient. J Gastroenterol 2005;40:1070-4.

12. Reddy RP, Smyrk TC, Zapiach M, et al. Pancreatic mucinous cystic neoplasm defined by ovarian stroma; Demographics, clinical features, and prevalence of the cancer. Clin Gastroenterol Hepatol 2004;2:1026-31.

13. Sarr MG, Capenter HA, Prabhakar LP, et al. Clinical and pathologic correlation of 84 mucinous cystic neoplasms of the pancreas; Can one reliably differentiate benign from malignant (or premalignant) neoplasms? Ann Surg 2000;231:205-12.

14. Sohn TA, Yeo CJ, Cameron JL. Intraductal papillary mucinous neoplasms of the pancreas; An interestingly recognized clinicopathologic entity. Ann Surg 2001;234:313-21.

15. Hruban RH, Takaori K, Klimstra DS, et al. An illustrated consensus on the classification of pancreatic intraepithelial neoplasia and intraductal papillary mucinous neoplasms. Am J Surg 2004;28:977-87.

16. Spinelli KS, Fromwiller TE, Daniel RA, et al. Cystic pancreatic neoplasms: observe or operate. Ann of surg 2004;239:651-9.

17. Pitman MB, Lewandrowski K, Shen J, et al. Pancreatic cysts: preoperative diagnosis and clinical management. Cancer Cytopathology 2009;118:1-13.

18. Falconi M, Salvia R, Bassi C, et al. Clinicopathological features and treatment of intraductal papillary mucinous tumors of the pancreas. Br J Surg 2001;88:375-81.

19. Osada S, Imai H, Okumura N, et al. A modified reconstruction method to prevent critical complications after pancreatoduodenectomy. Hepatogastroenterol 2006;53:296-300.

20. Osada S, Sanada Y, Tanaka Y, et al. Clinical evaluation of modified reconstruction method after pancreatoduodenectomy. Hepatogastroenterol 2009;56:619-23.