Chung Shan Medical Journal 2024; 35:143-146

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Case Report

Rare Synchronous Ampullary Vater Cancer combined with Pancreatic Body Cancer Patient receiving Whipple Operation with Total Pancreatectomy and Splenectomy-Case Report and Literature Review

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Ampullary vater cancer represents about 0.2% of gastrointestinal cancers and approximately 7% of all periampullary cancers. Synchronous ampullary vater cancer combined with pancreatic body or tail cancer is more rare. We presented the first case of synchronous ampullary vater cancer combined with pancreatic body cancer which received Whipple operation combined total pancreatectomy and splenectomy. Pathology report showed ampullary adenocarcinoma with duodenum invasion, pT2N0M0 stage IB and pancreatic body ductal adenocarcinoma, pT2N0M0 stage IB. Then he received the complete course of systemic chemotherapy and tolerated well. Now his clinical condition was stable and the period of disease free was more than 19 months.

Keywords: Ampullary cancer, Pancreatic cancer, Synchronous cancer, Whipple operation, Total pancreatectomy

1. Introduction:

Pancreatic cancer is a rapidly progressing disease and affects patients worldwide. Surgical resection is the primary intervention and the only potentially curative treatment option, but fewer than 20% of patients are surgical candidates.(1) An early report by Ihse et al. of outcomes after total pancreatectomy cited 27% operative mortality and 52% morbidity compared with 3% mortality and 28% morbidity for partial pancreatectomy.(1) Nathan et al. reviewed U.S. Surveillance, Epidemiology, and End Results program data and reported mortality rates after total pancreatectomy ranging from 5.8%–9.3%.(1) Compared with patients who undergo a partial pancreatectomy, those who undergo total pancreatectomy experience a higher incidence of perioperative morbidity and a higher re-operation rate.(1) Ampullary cancer is rare, accounting for about 0.2% of gastrointestinal cancers and 7% of all periampullary cancers.(2) Survival of ampullary cancer depends on stage, grade, and histologic type. Papillary carcinomas arising in adenomas had a more favorable survival than adenocarcinomas not associated with adenomas.(3) Synchronous ampullary cancer combined pancreatic body or tail cancer is more rare. We presented the first case of synchronous ampullary vater cancer combined with pancreatic body cancer which received Whipple operation combined total pancreatectomy and

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splenectomy. Pathology report proved with ampullary adenocarcinoma with duodenum invasion, pT2N0M0 stage IB and pancreatic body ductal adenocarcinoma, pT2N0M0 stage IB. After receiving the complete course of systemic chemotherapy, he had stable clinical condition and disease free about more than 19 months.

2. Case

A 57 years old male patient suffered from jaundice and received computed tomography, (CT) endoscopic retrograde cholangio-pancreatography (ERCP) with biopsy, and magnetic resonance cholangio-pancreatography (MRCP). CT report showed diffuse biliary tract dilatation(Fig.1). ERCP report showed a papilla of Vater tumor with common bile duct obstruction(Fig.2). Biopsy report showed papilla of Vater adenocarcinoma. MRCP report showed focal pancreatic tail duct dilatation which suspicious IPMN (Intraductal papillary mucinous neoplasm) (Fig.3). Then he received Whipple operation on 2021/11/10.

During operation, a hard whitish tumor was found located at pancreatic body area and frozen section reported as adenocarcinoma. Thus addition procedure was performed with Whipple operation combined total pancreatectomy and splenectomy (Fig.4A, 4B, 4C).

After operation, he recovered well postoperative course and was discharged on postoperative 14th date. The final pathology report showed ampullary adenocarcinoma with duodenum invasion, pT2N0M0 stage IB and pancreatic body ductal adenocarcinoma, pT2N0M0 stage IB. Then he received regular follow-up and chemotherapy with Gemcitabine for 17 cycles which was tolerated well. Until now, 2023/06, he

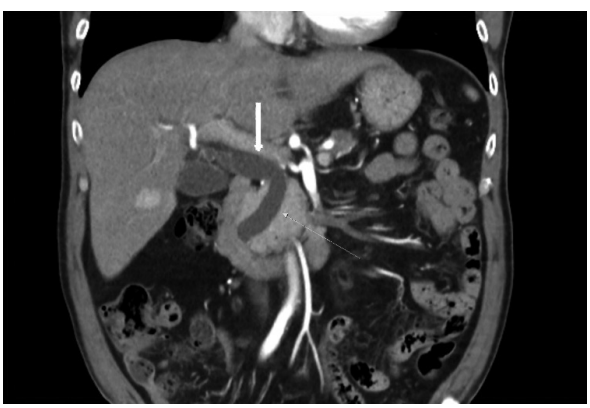


Figure 1. CT report showed diffuse biliary tract dilatation. (blue arrow)

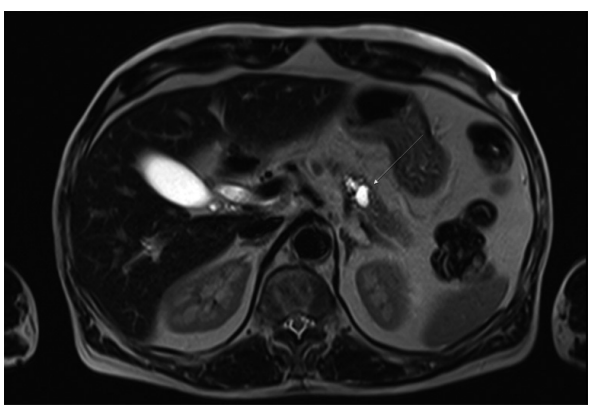


Figure 3. MRI report showed focal pancreatic tail duct dilatation which suspicious IPMN (yellow arrow)

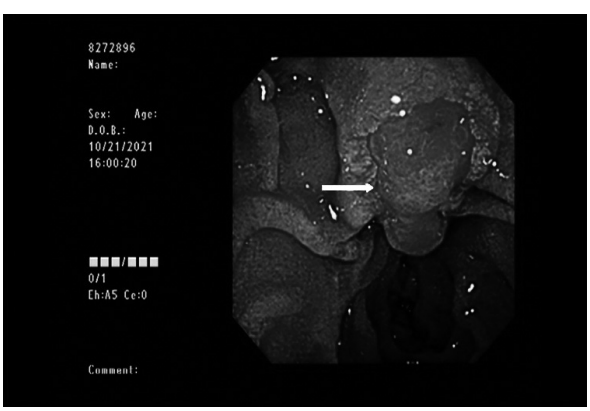


Figure 2. ERCP report showed a papilla of Vater tumor with CBD obstruction. (blue arrow)

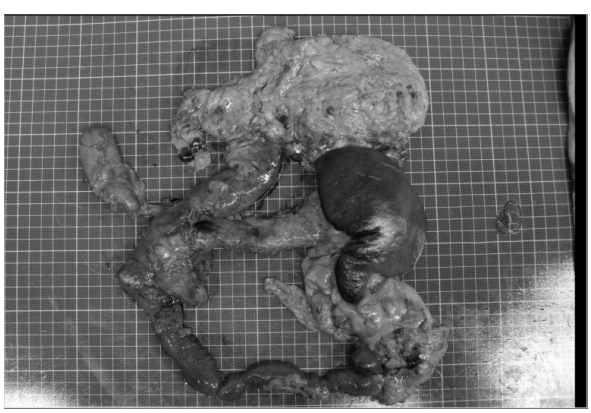


Figure 4A. Whole specimen of Whipple operation combined total pancreatectomy and splenectomy

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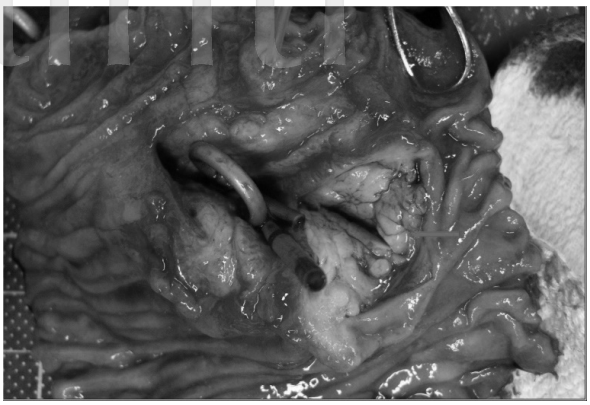


Figure 4B. Ampulla vater tumor and dilated CBD (blue arrow)

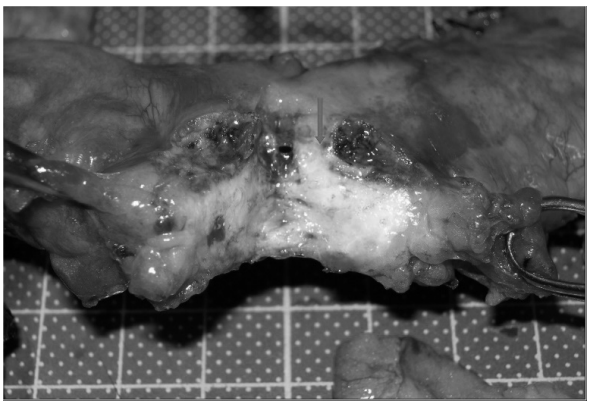


Figure 4C. Pancreatic body tumor (blue arrow)

presents relatively condition with well performance status and no evidence of tumor recurrence.

3. Discussion:

Periampullary tumors can originate from the pancreas, duodenum, distal common bile duct, or the structures of the ampullary complex. Primary ampullary tumors represent <0.5% of all gastrointestinal cancers and approximately 7% of all periampullary cancers.(2, 3)

Pancreatic cancer is a rapidly progressing disease and affects patients worldwide. Surgical resection is the primary intervention and the only potentially curative treatment option, but fewer than 20% of patients are surgical candidates.(1) An early report by Ihse et al. of outcomes after total pancreatectomy cited 27% operative mortality and 52% morbidity compared with 3% mortality and 28% morbidity for partial pancreatectomy.(1) Nathan et al. reviewed U.S. Surveillance, Epidemiology, and End Results program data and reported mortality rates after total pancreatectomy ranging from 5.8%–9.3%. (1) Compared with patients who undergo a partial pancreatectomy, those who undergo total pancreatectomy experience a higher incidence of perioperative morbidity and a higher re-operation rate. (1) Ampullary cancer is rare, accounting for about 0.2% of gastrointestinal cancers and 7% of all periampullary cancers.(2) Survival of ampullary cancer depends on stage, grade, and histologic type. Papillary carcinomas arising in adenomas had a more favorable survival than adenocarcinomas not associated with adenomas.(3)

Curative treatment for periampullary carcinoma is surgical resection. The standard surgical approach is Pylorus Preserving Pancreatoduodectomy (PPPD) or conventional Pancreatoduodectomy.(4, 5) Total pancreatectomy is indicated with extended chronic pancreatitis, neoplasms involving the entire pancreas or locally advanced pancreatic tumors.

In epidemiological studies, the frequency of multiple primary cancer is reported to be in the range of 2–17%.(6-8) One third of synchronous cancer found in related organs (breast-breast, breast-ovary, ovary-endometrium, pancreas-biliary tree).(9, 10) In this case of ampullary adenocarcinoma plus pancreatic body ductal adenocarcinoma, total pancreatectomy is appropriate. Actually, even in simple pancreatic adenocarcinoma or periampullary cancer, survival in patients undergoing total pancreatectomy was not significantly different with in patients undergoing distal pancreatectomy or pancreaticoduodenectomy and may provide comparable surgical and oncological treatment outcomes.(1, 11)

Due to the loss of pancreatic exocrine and endocrine functions, long-term complication of diabetes mellitus, malnutrition and Quality of Life decreasing would happen after total pancreatectomy but overall quality of life was good and showed no significant difference from pancreatoduodenectomy. (12, 13)

In our case, this patient's quality of life and clinical condition showed no obvious worsening. His physiologic reserve recovered well and received regular follow-up and chemotherapy with Gemcitabine for 17 cycles which was tolerated well. We report the first case that synchronous ampullary vater cancer combined pancreatic body cancer patient receiving

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Whipple operation with total pancreatectomy. Curative and completely surgical resection should be first considered for the case.

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