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Duodenopancreatectomy for PDAC Associated with MALS: A Case Report

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Rezumat

Duodenopancreatectomia cefalică pentru adenocarcinomul ductal cefalo-pancreatic asociat cu sindromul de ligament arcuat: prezentare de caz

Introducere: Adenocarcinomul ductal pancreatic reprezintă o patologie malignă agresivă, fiind cea mai des întâlnită afecțiune malignă a acestui organ (peste 90% din cazuri). Duodenopancreatectomia cefalică reprezintă tratamentul curativ al acestei patologii.
Prezentare de caz: Lucrarea de față prezintă cazul unei paciente, în vârstă de 68 de ani, transferată din alt serviciu în vederea investigațiilor şi a tratamentului unei tumori cefalopancreatice. În urma explorărilor clinice şi paraclinice se decelează un adenocarcinom ductal pancreatic localizat la nivel cefalo-uncinat, asociat unui sindrom de ligament arcuat, cu reumplere prin colateralele peripancreatice. În urma unei pregătiri preoperatorii corespunzătoare, se intervine chirurgical şi se practică următoarele: duodeno-pancreatectomie cefalică, hepatico-jejunoanastomoză transmezocolică, pancreato-gastro anastomoză, gastro-entero anastomoză termino-laterală precolică, by-pass aorto-hepatic cu grefon safen şi jejunostomie de alimentare tip Witzel.
Concluzie: Având în vedere lipsa fluxului în artera hepatică proprie în urma ligaturării şi secționării arterei gastroduodenale a fost necesar un procedeu de revascularizație, mai precis realizarea unui by-pass aorto-hepatic cu grefon safen. Evoluția postoperatorie a pacientei a fost favorabilă. Particularitatea

cazului rezidă în ocluzia completă a trunchiului celiac prin sindromul de ligament arcuat şi în faptul că irigarea organelor din etajul supramezocolic era asigurat printr-un flux retrograd al arterei mezenterice superioare în artera gastroduodenală.

Cuvinte cheie: adenocarcinom ductal pancreatic, sindrom de ligament arcuat

Abstract

Introduction: Pancreatic ductal adenocarcinoma (PDAC) is an aggressive form of pancreatic malignancy which occurs in over 90% of such cases. Pancreaticoduodenectomy (PD) is used with a curative purpose for localized PDAC.

Case presentation: A 68-year-old woman presented to our service through a transfer from another service, to be investigated and treated for a head of the pancreas tumor in a tertiary referral hospital. After a complete clinical and paraclinical evaluation, the patient was diagnosed with a PDAC and also with a median arcuate ligament syndrome (MALS). The surgical treatment was considered adequate, therefore, the patient underwent a PD with transmesocolic hepatico-jejunostomy, pancreaticogastric anastomosis, precolic end-to-side gastrojejunostomy, Witzel jejunostomy and with the help of the cardiovascular surgery team from the Heart Institute, Cluj-Napoca, an aorto-hepatic bypass using saphenous vein graft was performed.

Conclusion: Bypass was essential because the blood flow in the proper hepatic artery was not restored after sectioning the median arcuate ligament and clamping the gastroduodenal artery. The patient had a favorable outcome. The particularity of the present case consists of the complete occlusion of the celiac trunk by MALS and the total vascularization of the supra-mesocolic organs due to the superior mesenteric artery through the gastroduodenal artery.

Key words: pancreatic ductal adenocarcinoma, median arcuate ligament syndrome

Introduction

Pancreatic ductal adenocarcinoma (PDAC) is a highly aggressive malignant tumor. PDAC is the most prevalent type of pancreatic cancer, an exocrine malignancy, accounting for over 90% of pancreatic cancer cases (1-3). This type of neoplasm, primarily displays a glandular pattern with duct-like structures and requires a differential diagnosis with carcinomas of the intrapancreatic bile duct, ampulla of Vater or duodenal mucosa (2).

Pancreatic cancer occurs in the majority of cases in the head of the pancreas (65%), but it can also affect the body (15%), the tail of the pancreas (10%) or it can be multifocal in 2% of the cases. The 5-year survival rate of this condition is lower than 10% (1-3). Despite the fact that the mortality rates of patients with gastric, colorectal and hepatic cancers are decreasing due to therapeutic advances, there has been no sign of such an evolution regarding patients with PDAC. However, surgical interventions and chemotherapy have achieved an improvement in the survival of patients with early-stage PDAC (1,3,4).

In literature it is debatable whether the mortality of pancreatic cancer is a result of late diagnosis or limited response to treatment (1,2).

Pancreaticoduodenectomy (PD) is performed with a curative purpose for localized pancreatic ductal adenocarcinomas even though it is appropriate for only 20% of cases. However, when there is an intrinsic celiac trunk narrowing due to atherosclerosis or an extrinsic compression as a consequence of a median arcuate ligament syndrome (MALS), the ligation of the gastroduodenal artery can result in liver ischemia (5-7).

MALS, Dunbar syndrome or celiac artery compression syndrome (CACS) was first described by Dunbar in 1965 and represents an anatomic and clinical entity characterized by the extrinsic compression on the celiac axis (8). Dunbar syndrome leads in 1% of cases to the following symptoms: postprandial epigastric pain, nausea or vomiting, and weight loss. In about 35% of patients an abdominal bruit can be present. The median arcuate ligament is a muscular arch that connects the diaphragmatic crura thus developing the aortic hiatus (7-9).

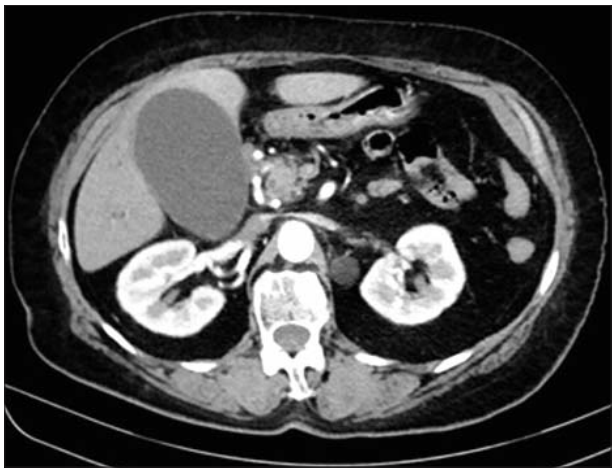


Figure 1. CT scan Coronal section

Case Report

A 68-year-old woman presented to the Prof. Dr. Octavian Fodor Regional Institute of Gastroenterology and Hepatology Cluj-Napoca, Romania through a transfer from another service, to be investigated and treated for a head of the pancreas tumor in a tertiary referral hospital. The patient had the following medical history: high blood pressure, diabetes mellitus type II under treatment with oral antidiabetics, arthrosis and a left knee arthroplasty performed in 2020. On arrival, the patient complained of right upper quadrant and epigastric pain, weight loss and exhibited severe jaundice. The patient had been evaluated using a native computed tomography (CT) in the previous medical service which revealed the following: bilateral intrahepatic biliary duct dilatation, hydropic gallbladder, a dilated common bile duct (CBD), measuring approximately 17 mm, a diffuse nonhomogeneous structure localized in the head of the pancreas near the uncinate process, assessed at 26 mm, two hypodense hepatic lesions in the fifth and sixth segments and a left adrenal adenoma.

Laboratory examinations showed hyperglycemia, hepatocytolisis (GPT=348/U/I, GOT = 312/U/I), cholestasis, expressed by high gamma-glutamyl transferase (GGT = 1027/U/I), alkaline phosphatase (ALP = 3065/U/I) and conjugated and total hyperbilirubinemia (23.8/mg/dL, respectively 28,5/mg/dL), lower albumin level (3.3/g/dL), elevated C reactive protein (CRP=2.31/mg/dL).

The patient was further investigated by a native and intravenous contrast substance computed tomography which revealed a head of the pancreas tumor, probably T2N1M0 (*Fig. 1*), a hydropic gallbladder with dilated intrahepatic bile ducts as well as the common bile duct, an adrenal myeloma and the occlusion of the celiac trunk due to a median arcuate ligament syndrome, associated with a voluminous and tortuous gastroduodenal artery and pancreatic collaterals (*Figs. 2, 3* – pointed by the arrow). The blood flow from the gastro-duodenal artery was reverse and the entire supramesocolic vascularization was made secondary to collaterals from the superior mesenteric artery to gastro-duodenal artery.

The patient was admitted to the surgical department with the purpose of continuing the diagnosis process and to receive proper preoperative therapy in case of performing the DP. An echo-endoscopy was performed, confirming the diagnosis through a fine needle aspiration (FNA) showing the presence of a ductal adenocarcinoma. Moreover, an endoscopic retrograde cholangiopancreatography was conducted to carry out a sphincterotomy, to take multiple biopsies and to place a plastic stent in the common bile duct. After a multidisciplinary assessment of the patient, the surgical treatment was considered mandatory, therefore, the patient went under a duodenopacreatectomy with transmesocolic hepatico-

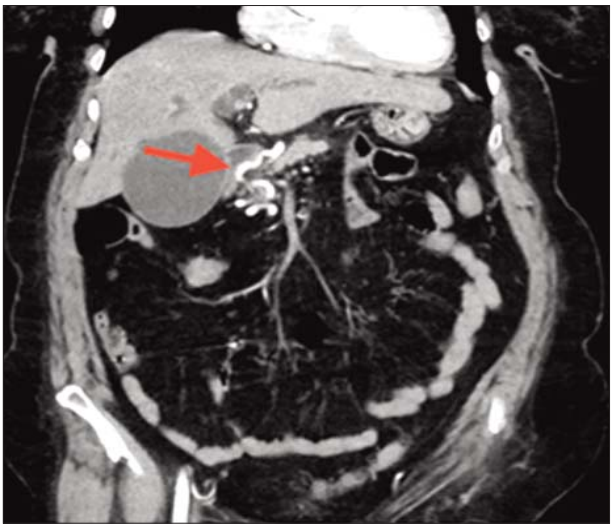


Figure 2. CT scan. The arrow showing the dilatation of the head of the pancreas arteries.

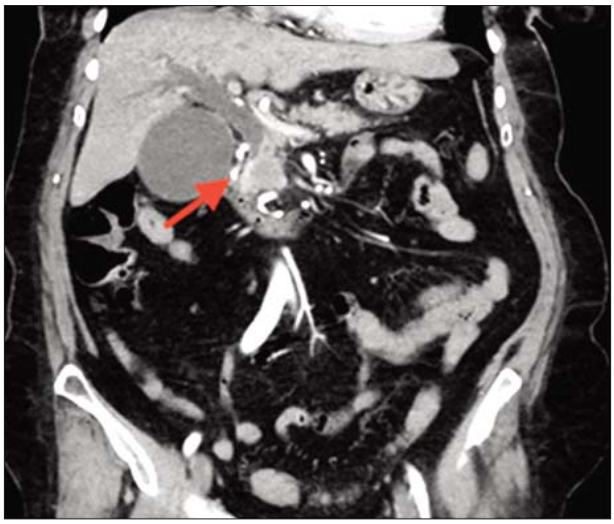


Figure 3. CT scan. The arrow showing the dilatation of the head of the pancreas arteries.

jejunostomy, pancreaticogastric anastomosis, precolic end-to-side gastrojejunostomy, Witzel jejunostomy and with the help of the cardio-vascular surgery team from the Heart Institute, Cluj-Napoca, an aorto-hepatic bypass was performed using saphenous vein graft (*Fig. 4* - the arrow is pointing the bypass). The bypass was necessary after sectioning the median arcuate ligament and also clamping the gastroduodenal artery, due to the lack of revascularization in the proper hepatic artery. The patient had a favorable outcome, being discharged 11 days after the surgical procedure.

Discussion

Median Arcuate Ligament Syndrome is one of the comorbidities that must be documented preoperatively when considering a duodeno-pancreatectomy (Whipple procedure). The prevalence of MALS in patients undergoing a PD is of 7.5% of cases. In most of them, MALS has been identified preoperatively, but in some cases, it has been an incidental intraoperative discovery (12-15). If this condition has not been identified preoperatively, it is necessary to raise the suspicion of celiac trunk occlusion during cephalic pancreaticoduodenectomy when the gastroduodenal artery (GDA) has an enlarged caliber and to perform the Bull maneuver (gastroduodenal artery clamping) before sectioning and gastroduo-denal artery ligation (5,7,11,15).



Figure 4. The Aorto-Hepatic bypass (pointed by the arrow)

Computed tomography with intravenous contrast substance in the arterial phase objectifies the existence of a celiac trunk occlusion, either due to an extrinsic (MALS) or an intrinsic cause (atherosclerosis) (10). Patients with MALS have been shown to present with increased peak systolic velocity on the celiac artery during expiration on Doppler ultrasound and narrowing of the celiac artery with more than 50% during expiration on the aortogram (9).

The particularity of the presented case is the complete occlusion of the celiac trunk by MALS and complete vascularization of the supramesocolic organs secondary to branches from the superior mesenteric artery, more precisely a retrograde flux in the GDA. In this case, the revascularization in the proper hepatic artery was not observed after the sectioning of the median arcuate ligament and the Bull maneuver, thus forcing a surgical revascularization technique (16).

To restore blood flow to the branches of the celiac trunk, there are several surgical techniques described in the literature from endovascular stenting to vascular reconstructions (12,14,15). The literature describes the two-step approach of MALS by having a laparoscopic/ robotic sectioning of median arcuate ligament or endovascular stenting of the celiac artery in the first step and then on a second admission performing curative surgery for PDAC. This approach was successful when performed on patients with benign tumors (pancreatic endocrine tumors), but it is not recommended in malignant tumors due to tumor progression during the waiting period and technical difficulty (5). When Whipple procedure is indicated for a benign or premalignant lesion, it is possible to preserve the collaterals and the gastroduodenal artery, as in the case presented by Berselli et al. in which a cephalic pancreaticoduodenectomy with pyloric preservation was performed for an intraductal papillary mucinous neoplasm (IPMN) with moderate degree of dysplasia (borderline) and the superio-posterior pancreaticoduodenal artery and the inferior-posterior pancreaticoduodenal artery were preserved and a successful TT anastomosis between them was made (9). In the surgical treatment of malignant tumors, as in the present case, in order to ensure oncological resection, this cannot be achieved.

A case study documenting a case of pancreatic cancer with concomitant MALS was successfully treated by performing a total pancreaticoduodenectomy and a bypass between the aorta and the proper hepatic artery by interposing an iliac artery allograft with TT anastomosis with the hepatic artery and TL with the aorta (11).

A study conducted in 2007 by Farma and Hoffman in 332 patients for whom cephalic pancreaticoduodenectomy was performed, identified 14 patients with celiac trunk occlusion. In 11 patients, the median arcuate ligament was released with neurolysis and in 3 cases revascularization was required: 1 aortoceliac bypass with saphenous vein, 1 common hepatic artery saphenous vein patch, 1 aorto-hepatic saphenous vein bypass (12).

In our case, the median arcuate ligament was released with the need to perform a revascularization surgical technique, which consisted of an aorto-hepatic bypass with a saphenous graft. In the time-frame between 1978 and 2021 there were only a few cases of MALS associated with a duodenopancreatectomy performed for a resectable pancreatic ductal adenocarcinoma (1-17).

Conflicts of Interests

The authors declare that they have no conflict of interest.

References

1. Corbo V, Tortora G, Scarpa A. Molecular pathology of pancreatic cancer: From bench-to-bedside translation. Curr Drug Targets. 2012;13(6):744-52.
2. Sarantis P, Koustas E, Papadimitropoulou A, Papavassiliou AG, Karamouzis MV. Pancreatic ductal adenocarcinoma: Treatment hurdles, tumor microenvironment and immunotherapy. World J Gastrointest Oncol. 2020; 12(2):173-181.
3. Ushio J, Kanno A, Ikeda E, Ando K, Nagai H, Miwata T, et al. Pancreatic ductal adenocarcinoma: Epidemiology and risk factors. Diagnostics (Basel). 2021;11(3):562.
4. Adamska A, Domenichini A, Falasca M. Pancreatic ductal adenocarcinoma: Current and evolving therapies. Int J Mol Sci. 2017;18(7):1338.
5. Whistance RN, Shah V, Grist ER, Shearman CP, Pearce NW, Odurny A, et al.

Management of median arcuate ligament syndrome in patients who require pancreaticoduodenectomy. Ann R Coll Surg Engl. 2011;93(4):e11-4.
6. Thompson NW, Eckhauser FE, Talpos G, Cho KJ. Pancreaticoduodenectomy and celiac occlusive disease. Ann Surg. 1981;193(4):399-406.
7. Balachandran G, Bharathy KGS, Sikora SS. Median arcuate ligament syndrome: an important consideration in pancreaticoduodenectomy. Indian J Surg Oncol. 2021;12(2):439-441.
8. Bayati IA, Gajendran M, Davis BR, Diaz JR, McCallum RW. Median Arcuate Ligament Syndrome Clinical Presentation, Pathophysiology, and Management: Description of Four Cases. Gastrointest. Disord. 2021, 3, 44–50.
9. Berselli M, Sperti C, Ballotta E, Beltrame V, Pedrazzoli S. Pancreaticoduodenectomy with unusual artery reconstruction in a patient with celiac axis occlusion: report of a case. Updates Surg. 2010;62(2):117-20.
10. Kazuaki S, Hirofumi K, Tatsuya O, Akihisa N, Shingo S, Kenji W, et al. Pancreaticoduodenectomy with Preservation of Collateral Circulation or Revascularization for Biliary Pancreatic Cancer with Celiac Axis Occlusion: Am J Case Rep. 2018;19:413-420.
11. Celik S, Ringe KI, Boru CE, Constantinica V, Bektas H. A case of pancreatic cancer with concomitant median arcuate ligament syndrome treated successfully using an allograft arterial transposition. J Surg Case Rep. 2015; 2015(12):rjv161.
12. Farma JM, Hoffman JP. Nonneoplastic celiac axis occlusion in patients undergoing pancreaticoduodenectomy. Am J Surg. 2007;193(3):341-4; discussion 344.
13. Kurosaki I, Hatakeyama K, Nihei KE, Oyamatsu M. Celiac axis stenosis in pancreaticoduodenectomy. J Hepatobiliary Pancreat Surg. 2004;11(2): 119-24.
14. Nakano H, Yamamura T, Yamaguchi S, Otsubo T. Celiac axisocclusion of a patient undergoing pancreaticoduodenectomy after distal gastrectomy. Hepatogastroenterology. 2007;54(74):595-8.
15. Halazun KJ, Kotru A, Menon KV, Patel J, Prasad KR. Stenting of coeliac axis stenosis facilitates pancreatectomy. Eur J Surg Oncol. 2006;32(7):811-2.
16. Bull DA, Hunter GC, Crabtree TG, Bernhard VM, Putnam CW. Hepatic ischemia, caused by celiac axis compression, complicating pancreaticoduodenectomy. Ann Surg. 1993;217(3):244-7.
17. Nara S, Sakamoto Y, Shimada K, Sano T, Kosuge T, Takahashi Y, et al. Arterial reconstruction during pancreatoduodenectomy in patients with celiac axis stenosis--utility of Doppler ultrasonography. World J Surg. 2005; 29(7):885-9.