Dicle Tıp Dergisi /
Dicle Medical Journal

2013; 40 (4): 681-684
doi: 10.5798/diclemedj.0921.2013.04.0359

CASE REPORT / OLGU SUNUMU

Annular pancreas associated with nutcracker syndrome: A case report

Nutcracker sendromu ile birlikte anüler pancreas: Olgu sunumu

Ayşe Sağıroğlu[1], Afra Yıldırım[2], Niyazi Acer[3], Turgut Tursem Tokmak[2], Emel Duran[4], Mevlüt Başkol[5]

ABSTRACT

The pancreas usually develops from the fusion of the dorsal and ventral pancreatic buds between the first 4-8 weeks of embryonic life. Annular pancreas consists of a ring of pancreatic tissue, which partially or completely surrounds the descending portion of the duodenum. Annular pancreas is an uncommon congenital anomaly. The etiology of this anomaly is still unknown. It has been associated with other congenital anomalies and various clinical symptoms. The nutcracker phenomenon is defined as compression of the left renal vein between the aorta and superior mesenteric artery. We report the case of a 54-year-old patient who presented with a 15-day history of nausea and vomiting associated with weight loss. On magnetic resonance cholangiopancreatography (MRCP) and computer tomography (CT) examination, an annular pancreas and nutcracker syndrome were diagnosed, respectively. In this study, we founded that the radiologic and clinical findings of associated annular pancreas with nutcracker syndrome may be of clinical importance for surgical procedures.

Key words: Annular pancreas, congenital anomaly, nutcracker phenomenon, MRCP, CT

ÖZET

Pancreas genellikle embriyolojik hayatın ilk 4-8 haftaları arasında dorsal ve ventral pankreas tomurcuklarının birleşmesinden gelişir. Anüler pancreas, pancreas dokusunun pars descendens duodeni'yi kısmi ya da tamamen halka şeklinde sarmasıyla oluşur. Anüler pancreas yaygın olmayan bir konjenital anomalidir. Bu anomalinin etyolojisi hala bilinmemektedir. Bu anomali, diğer konjenital anomaliler ve çeşitli klinik semptomlarla birliktelik gösterebilmektedir. Nutcracker fenomeni, v.renalis sinistra'nın aorta abdominalis ile a.mesenterica superior arasında sıkışması olarak tanımlanır. 15 gündür bulantı ve kusma ile birlikte kilo kaybı olan 54 yaşında bir hastanın olgusunu bildirdik. MRKP (Manyetik rezonans kolanjiopankreatografi) ve BT (bilgisayarlı tomografi) incelemelerinde anüler pancreas ve nutcracker sendromu tanısı konuldu. Biz bu çalışmada, radyolojik ve klinik olarak anüler pankreas ile nutcracker sendromunun birlikteliğininin cerrahi girişimler için klinik açıdan önemli olabileceğini gösterdik.

Anahtar kelimeler: Anüler pancreas, konjenital anomali, nutcracker fenomeni, MRKP, BT

INTRODUCTION

The pancreas usually develops from the fusion of the dorsal and ventral pancreatic buds between the first 4-8 weeks of embryonic life [1].

Annular pancreas is a rare congenital anomaly. There are many studies in which the diagnosis was made on autopsy or during surgery. The incidence of annular pancreas is estimated to be 3 in 20,000 [2].

Annular pancreas occurs due to the failure of the ventral bud to rotate, thus it becomes elongated and encircles the early part of the duodenum [1]. The etiology of this anomaly is still unknown [3]. There are 591 cases about annular pancreas reported in the literature. This congenital anomaly affects males more frequently than females [4]. In children, it is usually characterized by severe duodenal obstruction requiring immediate surgical treatment [4].

[1] *Kayseri Education and Research Hospital, Dept. Anatomy Kayseri, Turkey*
[2] *Erciyes University School of Medicine Dept. Radiology, Kayseri, Turkey*
[3] *Erciyes University School of Medicine Dept. Anatomy, Kayseri, Turkey*
[4] *Erciyes University School of Medicine Dept. Internal Medicine, Kayseri, Turkey*
[5] *Erciyes University School of Medicine Dept. Gastroenterology, Kayseri, Turkey*

Yazışma Adresi /Correspondence: *Ayşe Sağıroğlu,*
Kayseri Education and Research Hospital, Anatomy Kayseri, Turkey Email: sagirogluayse@yahoo.com

Geliş Tarihi / Received: 26.03.2013, Kabul Tarihi / Accepted: 27.04.2013

Annular pancreas has been associated with congenital anomalies and a variety of clinical symptoms. The nutcracker phenomenon is defined as compression of the left renal vein between the aorta and superior mesenteric artery [5]. The association between annular pancreas and nutcracker phenomenon has not been reported previously in the English literature. In this study, we present a case of associated annular pancreas with nutcracker phenomenon. The purpose of this case report is to discuss the diagnosis and conservative management of a patient with both annular pancreas and nutcracker syndrome.

CASE REPORT

A 54-year-old man presented with the principal complaint of right upper quadrant pain of fifteen days duration. He was admitted with large bilious emesis and worsening of his epigastric pain. He had been admitted to another hospital with these complaints for 3 days. His renal function was abnormal. He was diagnosed with prerenal acute renal failure. The patient was transferred to Erciyes University Medical Faculty Emergency Department for further examination. His blood parameters were within abnormal limits. Laboratory studies with a reference range revealed mild anemia with hemoglobin of 11.9 g/dl (14.0-18.0 g/dl), leukocyte count of 11.78 ×109/l (4.8-10.8×109/l), blood urea nitrogen of 53 mg/dl(9-23 mg/dl), creatinine of 1.64 mg/dl(0.7-1.3 mg/dl), serum sodium of 130 mmol/l (135-145 mmol/l), serum potassium of 3.9 mmol/l (3.6-4.8 mmol/l), alanine aminotransferase of 56 U/l (8-43 U/l), aspartate aminotransferase of 51U/l (0-34 U/l) alkaline phosphatase of 137 U/l(45-129 U/l) and albumin of 4.3 g/dl (3.5-5.0 g/dl). The patient was evaluated by abdominal ultrasound examinations. Abdominal ultrasonography revealed a gallbladder wall thickness of 3.5 mm, which is consistent with a diagnosis of cholecystitis. Gas was observed in the intrahepatic bile ducts. ERCP (endoscopic retrograde cholangiopancreatography) was planned for the patient. He was admitted to the gastroenterology service. Previous medical history included diabetes mellitus. The patient has smoked one and a half packs a day for 30 years. On physical examination, he appeared cachectic with a weight of 50 kg. There was no associated jaundice. Urinalysis showed hematuria. OGD (Oesophagogastrodudenoscopy) revealed post-bulbar stenosis also food residue was noted, suggestive of gastric outlet obstruction. The pylorus and first part of the duodenum were erythematous and edematous. Gastric peristalsis was decreased. There was evidence of ulceration in the first part of the duodenum. ERCP could not be performed because of gastric outlet obstruction. A contrast-enhanced abdominal CT scan showed normal pancreatic tissue and a pancreatic duct encircling the descending duodenum and that the liver was not cirrhotic. The intrahepatic bile ducts were dilated. The gallbladder was normal. Abdominal CT scan showed compression of the left renal vein between the aorta and superior mesenteric artery (Figure 1). Collateral vascular structures were observed at the level of the renal hilus. MRCP was performed for further evaluation; this showed an abnormal pancreatic duct encircling the duodenum, linked to the main pancreatic duct. There was also a dilation of the intrahepatic bile duct and choledoc duct (Figure 2).

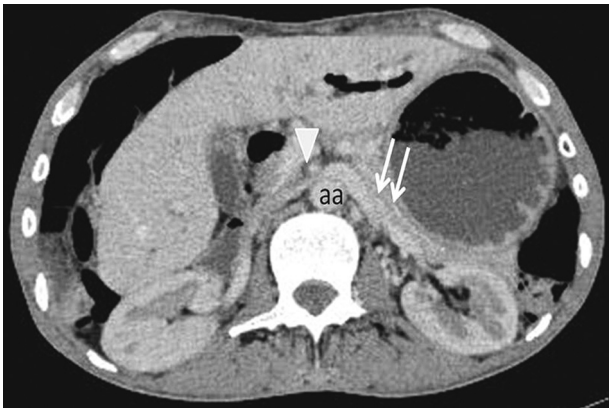

Figure 1 Axial contrast-enhancement CT demonstrates compression of the left renal vein (arrows) between the abdominal aorta (aa) and superior mesenteric artery (arrowhead)

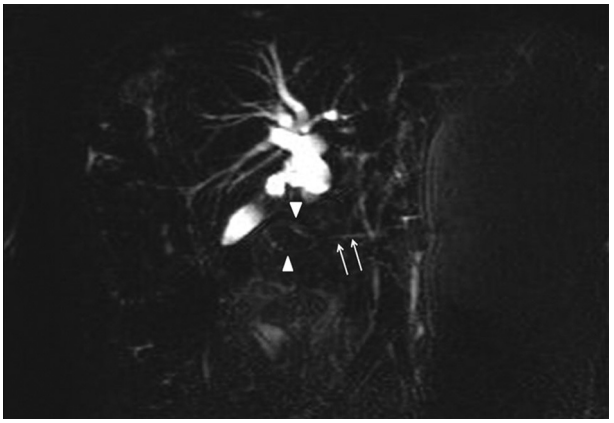

Figure 2 MRCP shows annular pancreatic duct (arrowheads) and Wirsung duct (arrows). There is a dilation of the intrahepatic bile duct and cholodec duct

The main pancreatic duct was of normal size. Due to the annular pancreas, there was an obstruction at the level of the first part of the duodenum. Finally, it was decided to proceed with gastrojejunostomy+antrectomy surgery in the patient. However, the patient did not agree to the operation. He was discharged on his own volition.

DISCUSSION

Annular pancreas, a rare congenital anomaly is found 1 in 20,000 newborns and the male population is more affected [1,6]. It consists of a ring of pancreatic tissue which partially or completely surrounds the descending portion of the duodenum [2,7]. There are two major theories about the embryologic development of annular pancreas. These theories were proposed by Lecco and Baldwin [8-10]. Lecco stated the adherence of the ventral bud to the duodenal wall results in a ring of tissue surrounding the duodenum during dorsal rotation. Additionally, Baldwin suggested that this condition arose from abnormal migration of the ventral pancreatic bud around the duodenum [1]. The regions of the duodenum which are usually affected are D2 (level of the second part of the duodenum) in 74% of cases and D1 (level of the first part of the duodenum) and D3 (level of the third part of the duodenum) in 21% [11]. In our study, there was an obstruction at the level of the first part of the duodenum. Diagnosis is made on the basis of CT and MR imaging findings that indicate pancreatic tissue and an annular duct surrounding the descending part of duodenum [3]. ERCP and CT are the methods used in the diagnosis of annular pancreas [12]. In addition, annular pancreas can be diagnosed non-invasively with MRCP [13,14].

In one study, 55 recent cases of annular pancreas in adults were reviewed [15]. Diagnosis was made with ERCP in 47%, MSCT (multislice computed tomography) in 18%, with MRCP in 16%, in 13% of the patients, diagnosis was made at the time of surgery [2]. According to a study made by Sandrasegaran et al [10], although ERCP is a well-accepted reference standard for pancreatic ductal disease there are cases in which obstruction of the annular duct close to its origin resulted in false-negative ERCP findings. ERCP was not an applicable diagnostic alternative due to duodenal obstruction by the pancreatic ring [2]. Annular pancreas has been associated with other congenital anomalies. These anomalies include the following: Down's syndrome, tracheoesophangeal fistula, intestinal atresia, pancreas divisum, intestinal malrotation, cardiac anomalies, malignancy and chronic pancreatitis [1,2]. Nutcracker syndrome leads to elevated renal venous pressure. As a result, continuous compression of the left renal vein and varicose dilatation of collateral vessels may affect the gonadal vein [5,16]. Complications include consequent hematuria, proteinuria and left flank pain. Only hematuria was observed in our patient. After excluding other causes of hematuria annular pancreas was diagnosed [16]. Some imaging techniques such as USG, MR angiography, and CT angiography, as well as measuring pressure gradients between the inferior vena cava and left renal vein during venography, are helpful in confirming this diagnosis [16]. Nutcracker syndrome with cases of Henoch Schonlein purpura, IgA nephropathy, membranous nephropathy, idiopathic hypercalciuria and concurrent urolithiasis have been described [17].

In this study, we founded that the radiologic and clinical findings of associated annular pancreas with nutcracker syndrome may be of clinical importance for surgical procedures. Clinicians should note the possibility of annular pancreas in patients with acute pancreatitis.

REFERENCES

1. Whittingham-Jones PM, Clayton G, Thompson HH. Annular pancreas - a rare cause of gastric obstruction in an 82-year-old patient. Ann R Coll Surg Engl 2005;87:W13-5.
2. Brönnimann E, Potthast S, Vlajnic T, et al. Annular pancreas associated with duodenal carcinoma. World J Gastroenterol 2010;16:3206-3210.
3. Singh P, Sandhu P, Saggar K, Ahluwalia A. Annular pancreas. Indian J Gastroenterol 2010;29:250.
4. Jarry J, Wagner T, Rault A, et al. Annular Pancreas: A Rare Cause of Acute Pancreatitis. JOP 2011;12:155-157.
5. Ulusan S, Koc Z. Left Inferior Vena Cava Associated with Nutcracker Phenomenon. Fırat Tıp Dergisi 2007;12:151-153.
6. Paraskevas G, Papaziogas B, Lazaridis C, et al. Annular pancreas in adults: embryological development, morphology and clinical significance. Surg Radiol Anat 2001;23:437-442.
7. Cunha JEM, Lima MS, Jukemura J, et al. Unusual Clinical Presentation of Annular Pancreas in the Adult. Pancreatology 2005;5:81-85.
8. Baldwin WA. A specimen of annular pancreas. Anat Rec 1910;4:299-304.

9. Lecco TM. Zur Morphologie des Pankreas annulare. Sitzungb Akad Wissensch 1910;119:391-406.
10. Sandrasegaran K, Patel A, Fogel EL, et al. Annular Pancreas in Adults. AJR 2009;193:455.
11. Hollander L, Marie A. Pankreas Annulare. In: von Allgower M, Harder F, Hollender F, Peiper J. (eds) Chirugische Gastroenterologie. Berlin: Springer 1981;1058-1060.
12. Reinhart RD, Brown JJ, Foglia RP, Aliperti G. MR imaging of annular pancreas. Abdom Imaging 1994;19:301-303.
13. Desai MB, Mitchell DG, Munoz SJ. Asymptomatic annular pancreas: detection by magnetic resonance imaging. Magn Reson Imaging 1994;12:683-685.
14. Hidaka T, Hirohashi S, Uchida H, et al. Annular pancreas diagnosed by single-shot MR cholangiopancreatography. Magn Reson Imaging 1998;16:441-444.
15. Zyromski NJ, Sandoval JA, Pitt HA, et al. Annular pancreas: dramatic differences between children and adults. J Am Coll Surg 2008;206:1019-1027.
16. Barsoum MK, Shepherd RFJ, Welch TJ. Patient with both Wilkie syndrome and nutcracker syndrome. Vascular Medicine 2008;13:247-250.
17. Kurklinsky AK, Rooke TW. Nutcracker Phenomenon and Nutcracker Syndrome. Mayo Clin Proc 2010;85:552-559.