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Pancreas Divisum ： An Infrequent Cause of Acute Pancreatitis in A Case

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Abstract

A 40-year-old female without history of alcohol consumption and medication suffered from persistent epigastralgia for four days and was admitted to our hospital. Elevated serum amylase and lipase levels were noted. She was admitted and treated under the working diagnosis of acute pancreatitis. Abdominal sonography showed normal common bile duct, but enlargement of the whole pancreas and mild main pancreatic duct dilatation without evidence of gallstone. CT demonstrated consistent findings of mild infiltration around the pancreatic tail and body with minimal fluid collection without any space-occupying lesion. Due to persistent symptoms for five days, MRCP was arranged that showed non-opacification of the ventral duct and dilatation of the dorsal duct that drained into the minor papilla. Pancreas divisum (PD) was diagnosed. She was discharged after 6 days of hospitalization with symptomatic and supportive therapy without ERCP or endoscopic intervention. PD is a congenital anomaly predisposing to acute pancreatitis. The diagnosis of PD requires a high level of suspicion, especially in younger patients without evidence of alcohol consumption, hyperlipidemia, or gallstone. Although ERCP can give definite diagnosis, MRCP is the diagnostic tool of choice because of its non-invasiveness and high resolution. The justification of the use of invasive therapeutic measures in PD patients with pancreatitis is still controversial. (J Intern Med Taiwan 2008; 19: 531-535)

Key Words ： Congenital anomaly, Pancreatic duct, ERCP, MRCP, Idiopathic pancreatitis

Introduction

Pancreatic divisum (PD) is a condition that develops if the embryonic fusion of the dorsal and the ventral pancreas buds at the gestational age of 6 weeks is incomplete. The failure of fusion between these two pancreatic ducts results in a short and narrow duct in the head of the pancreas that drains through the major papilla (ventral duct) and a much longer duct that drains most of the pancreatic secretions through the minor

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papilla (dorsal duct). The clinical relevance of PD remains controversial. Some authors consider it a normal variant of the ductal anatomy of the pancreas[1], whereas the others claim that it is a pathological condition that causes a relative stenosis of the minor papilla in the dorsal pancreas and predisposes to the development of pancreatitis. The prevalence of PD has been reported to be approximately 7% to 12.6% in Western populations and is the most common congenital pancreatic anatomical variant[2,3]. On the other hand, it is relatively uncommon in Asia with an estimated incidence of less than 2% in the general population[4].

Case Report

A 40-year-old female was admitted to our hospital due to severe epigastralgia for four days. It was dull and persistent. No history of alcohol consumption and medication was noted. Although gastritis was first diagnosed at another medical center, medical therapy failed to relieve her symptoms. Laboratory work-up at our hospital revealed elevated levels of serum amylase and lipase (812 U/L and 2,566 U/L, respectively), while serum bilirubin, triglyceride, alkaline phosphatase, antinuclear antibody (ANA), Anit-dsDNA and rheumatoid arthritis (RA) factor levels were found to be normal. She was admitted under the diagnosis of acute pancreatitis. Abdominal sonography showed normal diameter of common bile duct, but enlargement of the whole pancreas and mild main pancreatic duct dilatation. No gallstone was noted. CT scan demonstrated consistent findings of mild infiltration around the pancreatic tail and body with minimal fluid collection without any space-occupying lesion. Due to failure in symptomatic relief after supportive treatment for five days, MRCP was arranged that showed dilatation of the dorsal duct that drained into the minor papilla and non-opacification of the ventral duct (Fig. 1). PD was diagnosed. She was discharged on the seventh day with regression of symptoms after supportive medical therapy without ERCP or endoscopic intervention. She was followed with regular abdominal sonography for ten months without any signs or symptoms.

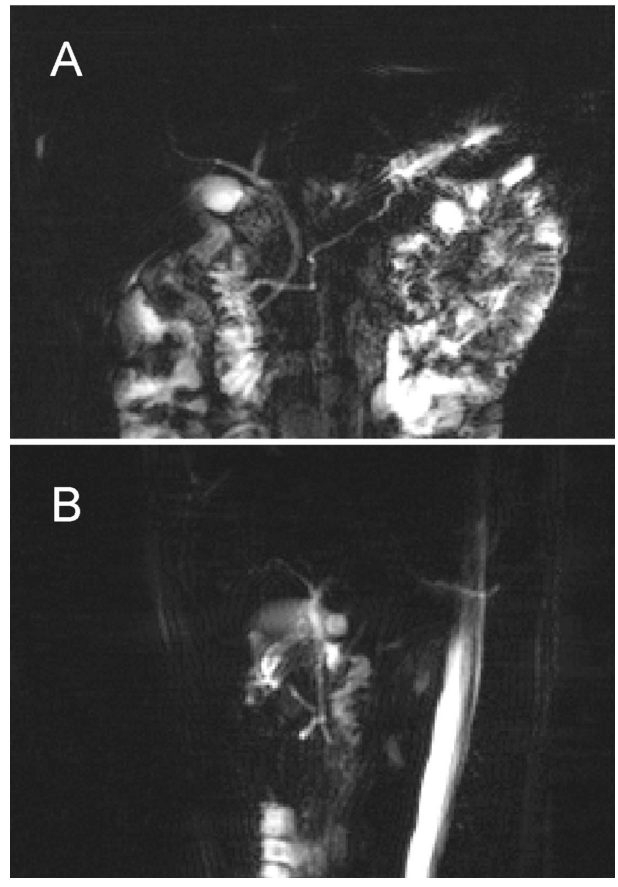

Fig.1.MRCP (PA view, A, and lateral view, B) showed non-opacification of the ventral duct and dilatation of the dorsal duct that communicated with the main pancreatic duct and drained into the minor papilla.

Discussion

PD results from an embryonic failure in fusion between the dorsal and ventral pancreatic buds and is common in the western world with a reported incidence of up to 9.3-10.8% in MRCP series[5, 6] and 5-6% in ERCP series[7]. However, the prevalence of the anomaly in Asia is relatively low with a reported incidence in the general population only between 1-2%[4,8]. Since the minor papilla is usually small but drains most of the pancreatic juice in PD subjects, the increased flow caused by fatty diet in combination with papillary stenosis may increase dorsal duct pressure and lead to the development of obstructive pan-

creatitis[9,10]. As a result, endoscopic or surgical sphincterotomy of the accessory papilla to release the pressure of pancreatic duct in symptomatic patients has been proposed[11,12].

However, other authors argue against the proposal because the prevalence of PD is not increased in idiopathic pancreatitis (IP) and the occurrence of pancreatitis in the ventral duct system in PD patients[2]. Moreover, the estimated incidence of pancreatic symptoms development has been reported in less than 5% of PD patients[2,13]. Accordingly, the indication for the use of endotherapy in IP patients with PD is controversial. While some clinicians proposed a beneficial role of endotherapy in pain relief as well as reducing incidence of recurrent pancreatitis and hospitalization [14], others hold an opposing view due to the ambiguous relationship between PD and IP, questionable accuracy in PD diagnosis, the benign clinical course in patients with IP and PD without endotherapy, and a lack of sufficient case number to justify the use of such invasive procedure[2].

Ultrasonography, CT scan, and conventional MR imaging are commonly used in the work-up of patients with pancreatic symptoms. Although these techniques are useful in evaluating the pancreatic parenchyma, they are of limited value in the detection of pancreatic duct anomaly. In patients with PD, a lobulated appearance of pancreas[12], homogeneous enlargement of the pancreatic head[10,15], or a fat cleft separating the dorsal and ventral pancreatic moieties[10] may be depicted on CT. However, these ancillary signs are of limited diagnostic value. ERCP is currently the definitive modality in the diagnosis of PD[1]. The conduction of ERCP in PD subjects involves the cannulation of the major papilla, followed by the injection of contrast medium into the short, thin, blind-ended ventral duct which typically shows no communication with the dorsal duct. Then the minor papilla is cannulated and the communication between the dorsal duct and the main pancreatic duct is defined[5]. However, an important pitfall of ERCP is the possibility of misinterpretation. For example, a shortened ventral duct may be interpreted as PD (i.e. a "false PD")[16]. The condition is attributable to other pathological causes, including a fibrotic stricture from alcoholic or pseudocyst in chronic pancreatitis, previous pancreatic trauma, partial pancreatectomy, or tumor[17,18]. Another limitation of using ERCP in PD diagnosis is the risk of severe pancreatitis associated with minor papilla cannulation which has been reported to be 2%[19]. MRCP is a new imaging technique that allows noninvasive multiplanar visualization of the biliary tree and pancreatic duct without injection of contrast medium[5]. It may be preferable to assess the anatomy of pancreatic duct because it minimizes iatrogenic complications and pitfalls associated with failed cannulation and injection of the dorsal duct present in ERCP studies. In addition, advances in fast magnetic resonance technology with the use of phased array coil have increased image resolution and enhanced diagnostic accuracy.

Autoimmune pancreatitis was ruled out in our reported case by the smooth contour of the main pancreatic duct and intrapancreatic portion of the common bile duct as well as lack of sausage-shaped enlargement of pancreas in CT scan. Moreover, she did not show any extrapancreatic presentation of autoimmune disease, such as rheumatoid arthritis and renal disease, that is frequently associated with autoimmune pancreatitis[20]. Biliary microlithiasis-related pancreatitis was also not considered since the inflammatory portion of the pancreas was over the body and tail according to the image findings. The pancreatic secretion of these portions was drained through the dorsal duct into the minor papilla that was not communicated with the bile duct in our case[2].

From January 2004 to January 2008, 4 out of 1548 patients that underwent ERCP were diagnosed as PD at our hospital. All 4 cases revealed a thin and short ventral duct through major papilla cannulation. Minor papilla cannulation was successful only in one case that showed a dilated dorsal duct. Failure in demonstrating

the ventral duct in our reported case may be due to its small caliber that precluded its visibility in MRCP. Our results are consistent with those from previous literature showing that the ventral duct cannot be demonstrated in 71% of PD cases in MRCP[21].

Since the diagnosis of PD is difficult, the incidence of PD-associated pancreatitis is probably under-estimated. Due to the common concept that acute pancreatitis in younger patients without gallstone is associated with either excessive or chronic consumption of alcohol or hyperlipidemia, the need for seeking rare predisposing factors such as PD is often ignored. Also, diagnostic tools such as ERCP, CT scan, and MRCP are not always available in district hospitals. Furthermore, an experienced examiner is required to properly conduct a ERCP and interpret the findings in patients with PD as mentioned above.

In conclusion, PD as a possible cause of acute pancreatitis should always be kept in mind. This is especially important in young adults since most of them are misdiagnosed as alcoholic, hypertriglycemic, or idiopathic. Although the cause-and-effect relationship between PD and pancreatitis is not well established, an accurate diagnosis of PD is important in allowing clinicians to consider more aggressive approach such as sphincterotomy for PD patients with repeated pancreatitis and persistent pain. However, long-term studies still need to be conducted to justify its use. Because of its noninvasiveness and high resolution, MRCP may be the diagnostic tool of choice for patients with PD which is an underestimated cause of acute pancreatitis in young adults and definitely deserves more attention in diagnosis and treatment.

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胰腺分裂症：
急性胰臟炎之罕見原因之一病例報告

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摘 要

一無喝酒及服藥病史之四十歲女性，因持續性上腹疼痛四天入院，血清澱粉酵素及解脂酵素皆有上升情形，臨床診斷急性胰臟炎接受治療；腹部超音波顯示，總膽管大小正常，無膽結石，但胰臟有腫大及主胰管有輕微擴大現象；電腦斷層發現相同情形，且胰臟體部和尾部有浸潤及小量積液情形；因治療五天後，症狀仍未改善，故安排核磁共振膽胰道攝影術檢查（MRCP），顯示胰臟腹側管並未顯影，背側管擴大且連接至副乳頭，診斷爲胰腺分裂症（Pancreas divisum, PD）。病患並末接受經內視鏡逆行性膽胰管攝影術（ERCP）及內視鏡治療術，經症狀和支持治療住院六天後出院；PD爲一先天性異常且可能誘發急性胰臟炎；必須有高度警覺性才能診斷PD，特別針對沒有喝酒、高血脂症或膽結石之年輕病患。雖然ERCP是最明確之診斷工具，但MRCP爲一非侵入性及高解析度之另一選擇工具；至於PD病患合併急性胰臟炎，是否需接受內視鏡治療，至今仍未有定論。