Pancreas divisum, pancreaticolithiasis and extraordinary ductal variation in a 14-year-old girl

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ÖZET

(Pankreas divizyum, pankreas taşı ve çok nadir kanal varyasyonu olan 14 yaşında kız hasta)

Pankreas divizyum dorsal ve ventral pankreatik kanalların birleşmemesi sonucu ortaya çıkan pankreasın embriyolojik bir anomalisidir. Çalışmada çok nadir görülen pankreatik kanal anomalisi olan çocuk olgu sunulmuştur. Sunulan olguda dorsal pankreatik kanal ana kanal olup, ventral kanal ikincil kanal olarak dorsal kanalla birleşmektedir. Ana papilla sadece koledok ile devam etmektedir. Kız olgu pankreas divizyununun nadir bir komplikasyonu olan pankreas taşları ile de komplike olmuştur. Olgu birden fazla denenen minör papilla sfinkterotomisine cevap vermemiş ve bu nedenle de açık cerrahi tedavi planlanmıştır. Olguya genişletilmiş uzunlamasına pankreatiko-jejunostomi (Frey operasyonu) uygulanmış ve cerrahi sonrası son bir yıldır olgunun hiçbir yakınması olmamıştır.

Anahtar Kelimeler: *Pankreas divizyum, pankreatit, pankreas taşı, cerrahi tedavi*

SUMMARY

(Pancreas divisum, pancreaticolithiasis and extraordinary ductal variation in a 14-year-old girl)

Pancreas divisum is an embryological anomaly of the pancreas that is characterized by a lack of fusion of the dorsal and ventral pancreatic ducts. Following case represents an unusual variant of pancreatic ductal anatomy in a child with pancreas divisum. In the presented case, dorsal pancreatic duct is the main pancretic duct while ventral duct is seen as an accesory duct join the dorsal duct. Major papilla is continued with the only common bile duct. She is also complicated with dorsal duct pancreaticolithiasis which is an uncommon complication of pancreas divisum. She failed multiple attempts at endoscopic minor papilla sphincterotomy and refered for surgical exploration. She underwent extended longitudinal pancreatico-jejunostomy (Frey's Operation) and remains symptom-free after one year.

Keywords: *Pancreas divisum, Pancreatitis, Pancreaticolithiasis, Surgical therapy*

INTRODUCTION

The pancreatic ductal drainage results from fusion of the dorsal and ventral pancreas buds during embryogenesis. The most common anatomical result is fusion of the dorsal duct (duct of Santorini) and the ventral duct (duct of Wirsung), with regression of the distal portion of the dorsal duct leading to drainage of both portions of the pancreas through major papilla. Pancreas divisum results from failure of fusion of the dorsal and ventral ducts of the pancreas during embryologic development leading to variations in pancreatic duct anatomy. Three variants have been described: type 1 or classical divisum in which there is total failure of fusion; type 2 in which dorsal drainage is dominant in the absence of Wirsung's duct; type 3 or incomplete divisum where a small communicating branch is present 1,2. This article reports a case of an 14-year-old girl with reccurent pancreatitis and pancreas divisum associated with dorsal duct pancreaticolithiasis and an unusual ductal anatomic variation.

CASE REPORT

An 14-year-old girl was referred to the Turgut Ozal Medicine Center Pediatric Surgery service with a 5-year history of recurrent bouts of epigastric and back pain associated with nausea and anorexia. She experienced four episodes of pain over the course of 1 year, and these episodes progressively worsened in intensity. Elevation of both serum amylase and lipase levels were documented on each occasion, with resolution of these laboratory abnormalities and resolution of the symptoms. Ultrasonography had shown evidence of pancreatic diffuse enlargement

in the head of the pancreas with ductal dilatation and multiple pancreatic stones during attack **(Figure1)**. Magnetic resonance cholangiopancreatography confirmed the diagnosis of pancreas divisum. Endoscopic retrograde cholangiopancreatography (ERCP) through the major papilla demonstrated the only common bile duct **(Figure 2A)**. ERCP revealed a minor papilla that delineated the dorsal pancreatic duct on cannulation, had massive dilatation, and contained multiple calculi **(Figure 2B)**.

Figure 1:Ultrasonography had shown evidence of pancreatic diffuse enlargement in the head

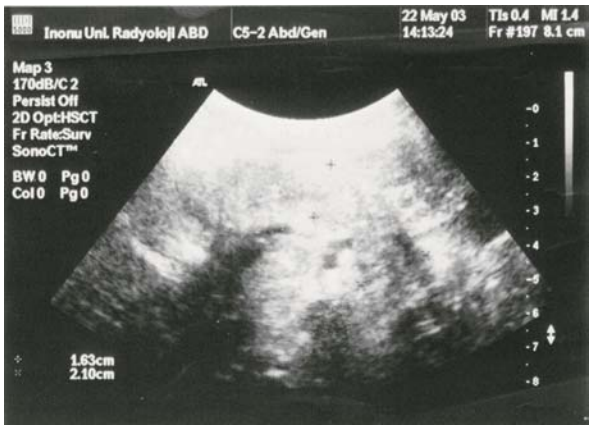


Figure 2A: ERCP through the major papilla demonstrated the only the common bile duct

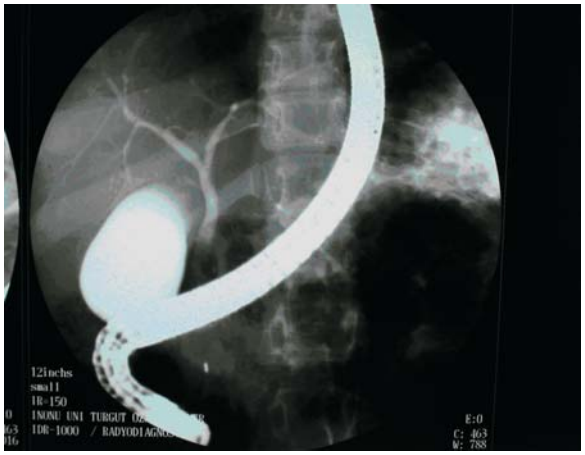
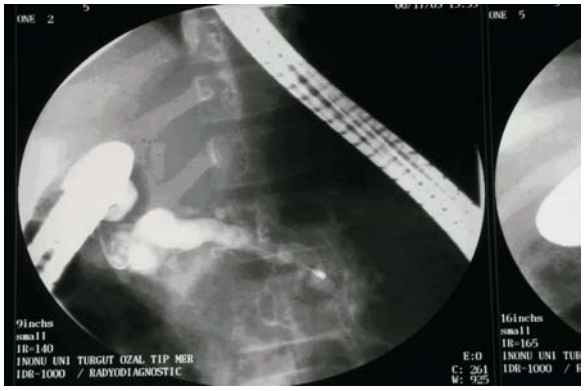
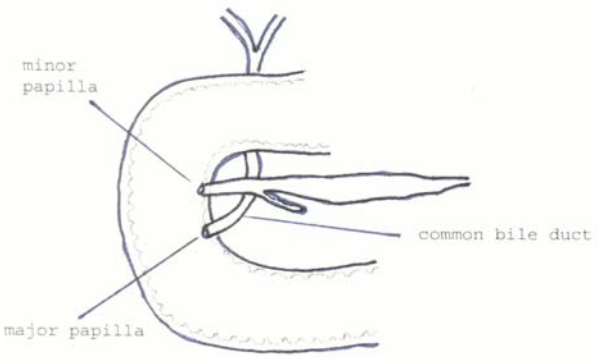


Figure 2B: ERCP revealed a minor papilla that delineated the dorsal pancreatic duct on cannulation, had massive dilatation, and contained multiple calculi



Endoscopic papillotomy with a 7F stent placement in the dorsal pancreatic duct during ERCP to facilitate drainage was performed for two times. Results of minor papillotomies were no favorable for patients with reccurent, well defined bouts of pancreatitis were continued. At surgery, diffuse changes of chronic pancreatitis were found and extended longitudinal pancreticojejunostomy was performed. Pancreatotomy revealed a dilated dorsal duct system with multiple stones, and a tenuous connection between the ventral and dorsal ducts. The pancreatic incision was extended onto the head of the pancreas and into ventral ductal system. Limited excision was made in the head of the pancreas and a Roux-en-Y pancreatico-jejunostomy was created to provide dranaige of the gland (Frey operation).**(Figure 3)**

Figure 3: Unusual anatomic ductal variation was found in our case



Postoperatively the patient recovered uneventfully and was discharged from the hospital postoperative day 10. At the twelve months' of follow-up, the patient has had no evidence of abdominal pain and does not require narcotics.

DİSCUSSİON

Pancreas divisum not rare, being present in 10% of the population 3. A variety of ductal abnormalities fall within the framework of pancreas divisum, including total separation of dorsal and ventral ducts, total absence of the ventral duct, and a tenuous connection between the dorsal and ventral ducts. 1,2. This case represents an unusual anatomic ductal variation of pancreas divisum. As shown in figure 3, dorsal pancretic duct is the main pancreatic duct while ventral duct is seen as an accesory duct join to the dorsal duct. Major papilla continued with only the common bile duct.

Approximately 25 percent of patients with pancreas divisum will develop complications such as recurrent abdominal pain and relapsing acute pancreatitis as a consequence of stenosis of the minor papilla with altered dorsal duct drainage 4,5. Pancreaticolithiasis is rare complication of pancreas divisum and presented cases in the literature are usually adult patients 6,7. Our patient had periodic abdominal pain with a clinical episodes of pancreatitis associated with intense dorsal duct pancreaticolithiasis.

Endoscopic interventions such as minor papillotomy or dilatation and subsequent dorsal duct stent placement provide less invasive alternatives 1. Endoscopic treatment in our patient have also required approaches through minor papillae to address the multiple dorsal duct calculi. Just, endoscopic interventions such as minor papillotomy and subsequent dorsal duct stent placements were failed in our case. We performed extended longitudinal pancreatico-jejunostomy (Frey's Operation) 8. The rationale local resection of the head of the pancreas combined with longitudinal pancreaticojejunostomy was proposed to treath the patient with chronic pancreatitis who has a markedly enlarged fibrotic pancreatic head and uncinate process impacted with calculi and small pseudocyst along the ducts of dorsal ventral and their tributary ducts, as well as a dilated duct in the body and tail of the pancreas. However, any patient, who is a candidate for longitidinal pancreaticojejunostomy may benefit from local resection of the head of the pancreas combined with longitudinal pancreaticojejunostomy due to the advantages associated with decompressing the other duct, the duct to the uncinate process and their tributary ducts. Who responded well to Frey's operation and uneventful at the end of first postoperative year.

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