An eighty-six-year old male was admitted for fever and jaundice, which prompted an abdominal CT scan and serial radiological investigations. Cholangitis was considered on the basis of the CT, MRI and MRCP findings. Besides, a bizarre-shaped spleen and a small splenule were found. A short pancreas and a preduodenal portal vein were also noted. These anomalies were part of the polysplenia syndrome. Polysplenia syndrome is a rare congenital anomaly frequently associated with cardiopulmonary and abdominal disorders. In adult cases, there are usually only minor associated anomalies. Awareness of these abnormalities helps in recognizing the syndrome. CT is proved to be an excellent imaging modality in diagnosing the abdominal anomalies. MRI and MRCP may give more detailed information.

**Key words:** MRCP; Polysplenia; Preduodenal portal vein; Short pancreas

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impressed on the basis of these imaging findings. T2-weighted MR imaging also disclosed the presence of a short pancreas (Figure 5). A combination of splenic, portal venous and pancreatic anomalies leads to the diagnosis of polysplenia syndrome. Cardiac ultrasound revealed no evident congenital cardiac anomaly, despite of mild diastolic dysfunction.

Under the impression of chronic cholangitis and CBD stricture, CBD excision and hepatojunostomy were performed. A preduodenal portal vein was seen during operation. Finally, poorly differentiated adenocarcinoma was diagnosed on the histopathological examination.

DISCUSSION

Polysplenia syndrome presenting with severe cardiac anomaly is usually diagnosed in early childhood and carries a grave prognosis. In some cases, correct diagnosis may be delayed until adulthood, particularly when there is no associated congenital heart defect. Many kinds of abdominal anomalies are present in this syndrome. Multiple spleens, azygos or hemiazygos continuation of IVC, preduodenal portal vein, short pancreas and visceral heterotaxia are commonly encountered in reported cases.

Figure 1. A bizarre-shaped spleen with a deep fissure (arrow).

Figure 2. a. Preduodenal route of portal vein. Enhanced CT shows a portal vein (arrow) straddling the duodenum (D) and pancreas. Di, duodenal diverticulum from second portion of duodenum. b. Enhanced CT in a more caudal level shows spleno-portal junction (arrow), which is located anteromedially to pancreatic head. Little pancreatic tissue is seen anterior to splenic vein.

Rarely, genitourinary anomalies such as double ureters, renal agenesis or hypoplastic kidney are reported as part of the polysplenia syndrome [1,2]. However, there is no unique pathognomonic anomaly. In our case, only three anatomic anomalies are identified, that is, a polysplenia, a short pancreas and a preduodenal portal vein. These anomalies were consistent with polysplenia syndrome.

Although presence of multiple splenules is the
most consistent criterion for polysplenia, a fairly large spleen segmented by deep fissures has ever been reported [3]. The location of multiple spleens was reported to be either on the left side or on the right side, and almost situated along the greater curvature of the stomach [3]. When CT scan is used to detect this anomaly, adequate small bowel opacification is essential. Otherwise, small splenules could be overlooked or confused with unopacified bowel.

Short pancreas, agenesis or hypogenesis of the dorsal pancreas, is a rare congenital anomaly. It can occur as an isolated anomaly or be associated with the polysplenia syndrome. Congenital short pancreas is related to embryologic failure of the dorsal bud, which develops into body and tail. Anomalies of the dorsal pancreas and spleen are expected to occur together because both develop in the dorsal mesogastrium. Disturbances in blood supply to the pancreatico-splenic region during embryonal life can cause concomitant anomalies. According to the degree of immaturity of the dorsal pancreas development, hypoplasia of the pancreas is classified clinically into three types [4]: A, total agenesis of the dorsal pancreas; B, hypogenensis of the body and tail; C, hypogenensis of the tail. Our patient is a case of type C pancreas hypoplasia.

CT and MRI demonstrate only the head of the pancreas, which may be sometimes prominent. Bowel loops and fatty tissue may fill in the expected normal position of the body and tail. It is important to recognize the congenital short pancreas in order to avoid mistaking the pancreas for a mass lesion. Inability to visualize the pancreatic duct throughout the pancreas is a frequent problem of ERCP (Endoscopic Retrograde Cholangiopancreatography) in the cases of short pancreas. In contrast, MRCP, in our

Figure 3. Coronal enhanced T1-weighted MRI shows wall thickening of common bile duct and wall enhancement (arrow). Abrupt tapering of distal portion is noted.

Figure 4. MRCP, T2-weighted, 3D TSE, (TR:2300ms, TE:750ms) coronal maximal intensity projection reconstruction shows abrupt tapering of distal extrahepatic bile duct (arrow), more evident in lateral aspect.

Figure 5. Turbo spin echo (TR:2500ms, TE:100ms, turbo factor:23) T2-weighted axial image shows a short pancreas, whose tail terminates at a proximal site (arrow). Bowel loops and fatty tissue fill the expected position of normal pancreatic tail.
case, can give similar information in a non-invasive and safe approach.

Our patient has adult-onset diabetes mellitus. Insulin-dependent diabetes mellitus had been reported in cases of isolated congenital short pancreas [4,5]. However, only one case of short pancreas in polysplenia syndrome was reported to be associated with adult-onset DM [6]. The association between agenesis of the dorsal pancreas and diabetes is yet to be ascertained. Carcinoma of the pancreas was reported in a 53-year-old woman with polysplenia and short pancreas [2]. The association between the adenocarcinoma of the common bile duct and polysplenia syndrome in our case has not been found in the literature.

Preduodenal portal vein is a common venous anomaly in this syndrome. It passes ventral to the duodenum and the head of the pancreas, and appears as a round structure anterior to the pancreatic head on CT and MR. Far anteriorly located portal vein, named as “preduodenal-transhepatic- intraperitoneal” portal vein, was reported in a case of polysplenia syndrome [7]. Preduodenal portal vein might interfere mechanically with pancreatic development, thereby increasing the risk of pancreatic anomalies such as annular pancreas. Potential hazard of preduodenal portal vein in some surgical procedures is obvious. Accidental injury to vein itself was reported in a case of polysplenia syndrome undergoing biliary surgery [8].

CT is excellent in demonstrating these anomalies. Spiral CT, especially with rapid injection of intravenous contrast material and with thin collimation, demonstrates the venous anomaly very clearly. MR is an excellent method to evaluate the venous anomaly because of its multiplanar imaging capability. MRCP shows not only biliary tract obstruction but also relatively short pancreatic duct, which is a common finding of ERCP in congenital short pancreas.

Differentiation of benign from malignant causes of biliary tract dilatation is an important clinical concern. Abrupt termination of a dilated extrahepatic duct is characteristic of a malignant process in the absence of a mass. Gradual tapering of a dilated duct is specific for benign diseases [9]. In another study for wall thickening of bile duct, thickening of greater than 5mm was seen only with cholangiocarcinoma [10]. Other findings, such as degree of intra- or extra-hepatic duct dilatation, presence or absence of a dilated pancreatic duct, and enhancement pattern of duct wall are of no predictive value. A recent study pointed out that the use of nonenhanced T1- and less heavily T2-weighted images with MRCP images significantly improved the diagnostic accuracy of MR examinations of pancreaticobiliary disease. The accuracy, sensitivity and specificity of differentiation of benign from malignant causes of biliary dilatation are 82%, 96% and 71%, respectively [11]. In our case, thickness of abnormal CBD wall reaching about 5.5mm on CT and MRI, as well as relatively abrupt termination of dilated proximal bile duct should raise the suspicion of malignant disease.

In conclusion, polysplenia syndrome is a rare congenital anomaly that may be found incidentally in adults who undergo abdominal CT, conventional MRI or MRCP for other reasons. Awareness of its associated anomalies helps us recognize them as part of the syndrome rather than separate pathological processes.

**REFERENCE**

多脾症伴隨十二指腸前肝門靜脈及短小胰臟：
一個總膽管腺癌病例之偶然發現

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一位八十六歲男性因為發熱及黃疸住院，電腦斷層攝影、磁振造影和磁振膽胰攝影認為是膽管炎。此外，我們發現病人有一個怪異形狀的脾臟和一個小脾，也看到短小胰臟和十二指腸前肝門靜脈，這些異常是多脾症的一部分。

多脾症是一種罕見的先天異常，通常伴隨心肺及腹部的問題，成人的病例通常只有輕微的異常。了解這些異常使我們知道這是症狀的一部分，而非不同的疾病。電腦斷層在診斷此腹部異常方面是很好的工具，磁振造影及磁振膽胰攝影則可提供更多的訊息。

關鍵詞：多脾症；十二指腸前肝門靜脈；短小胰臟；磁振膽胰攝影