ORIGINAL RESEARCH PAPER
Volume-8 | Issue-10 | October - 2019 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MDCT FINDINGS OF AN ADULT WITH POLYSPLENIC SYNDROME ASSOCIATED WITH PREDUODENAL PORTAL VEIN AND SHORT TRUNCATED PANCREAS: A CASE REPORT

Radiodiagnosis

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ABSTRACT

Polysplenia syndrome, also known as left isomerism, is a type of heterotaxy syndrome where there are multiple spleens (congenitally) as part of left-sided isomerism. Polysplenia is characterised by multiple splenules without a parent spleen. The most common associated feature is inferior vena cava interruption with azygous or hemiazygous continuation. Rare associations of polysplenia syndrome includes preduodenal portal vein (PDPV) and short truncated pancreas. PDPV is a rare developmental anomaly in which the portal vein runs at the ventral side of the duodenum instead of at an intrapancreatic location. Understanding of this anomaly is of considerable surgical importance, because it may cause unintended portal vein injury during operations involving the gall bladder or duodenum.
We recently and incidentally detected a PDPV and short truncated pancreas associated with polysplenia syndrome in a 22-year-old female patient who complained of dry cough. We report here the Multidetector-row CT findings of a rare case of polysplenia with PDPV, short truncated pancreas and other anomalies.

KEYWORDS

Preduodenal Portal Vein- PDPV; Adult; Congenital Anomalies; MD CT

INTRODUCTION

Polysplenia syndrome is a rare heterogeneous disease. It primarily affects the asymmetric organs, including the heart, lungs and bronchi, liver, intestines, and spleen(1) . Reports indicate that most patients with polysplenia syndrome die before 5 years of age because the disease is often associated with congenital anomalies, such as cardiovascular anomalies(2). 5-10% of the patients lack cardiac involvement, which allows them to reach adulthood(3). Patients with polysplenia syndrome have a normal heart or only minor cardiac defects, are often diagnosed incidentally in patients being treated for other disease(4). However, they may harbor anomalies in abdominal organs or the gastrointestinal tract with a wide range of abnormalities(5). The range of anomalies include multiple spleens of equal volume, visceral heterotaxia, right-sided stomach, a left-sided or large midline liver, malrotation of the intestine, a short pancreas, preduodenal portal vein and inferior vena cava anomalies, bilateral hyparterial bronchi, bilateral bilobed lungs. (5).

We recently saw a case of PDPV and short truncated pancreas combined with polysplenia. There are several reports about PDPV associated with polysplenia syndrome in adult patients, but the detailed Multidetector-row CT (MDCT) findings of these anomalies in the adults are not well identified (2, 5). We report here the MDCT findings of PDPV and short truncated pancreas associated with polysplenia syndrome in an adult patient. Understanding this anomaly is of considerable surgical importance, because the vein lies in the most superficial position ventral to the duodenum and if PDPV is not detected prior to surgery, it can cause severe complications, such as hemorrhage and vascular ligation.

CASE REPORT

A 22-year-old female visited the outpatient centre with complaints of dry cough for which she was advised HRCT chest, which we converted to Contrast CT after seeing plain CT findings . The CT scan showed few calcified sub carinal lymph nodes, bilateral bilobed lungs with bilateral hyparterial bronchi (Fig. 1 A & B). Also an unusual appearance of the portal vein was seen, located ventral to the duodenum. Mesenterico-portal vein and mesenterico-splenic vein anastomosed caudally in the lower abdomen (Fig. 3A, B, C). Portal vein was seen coursing anteriorly to gall bladder and entering into liver at segment IVB, this represents the preduodenal portal vein (PDPV). Few of the intrahepatic branches of portal vein from segments V and IVB were seen reforming the right posterior ascending portal vein at porta and supplying segment VII (Fig. 1D). In addition, 3 spleens are noted in left hypochondriac region (Fig. 1 A). The IVC was interrupted at level of left renal vein with an azygos continuation and absence of the in trahepatic portion of the IVC (Fig. 2A, B, C). Left renal vein shows retro aortic course. Three hepatic veins are draining into right atrium (Fig. 2B). Rest of mediastinal vasculature was within normal limits. Four pulmonary vein seen draining into left atrium. The pancreas body was truncated, with only the pancreatic head, uncinate process and proximal portion of the pancreatic body was seen (Fig. 1C).Pancreatic tail and distal body was absent. There was no evidence of duodenal obstruction, or intestinal malformation.

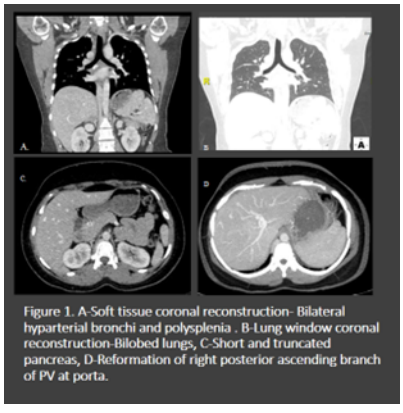

Figure 1. A-Soft tissue coronal reconstruction- Bilateral hyparterial bronchi and polysplenia . B-Lung window coronal reconstruction-Bilobed lungs, C-Short and truncated pancreas, D-Reformation of right posterior ascending branch of PV at porta.

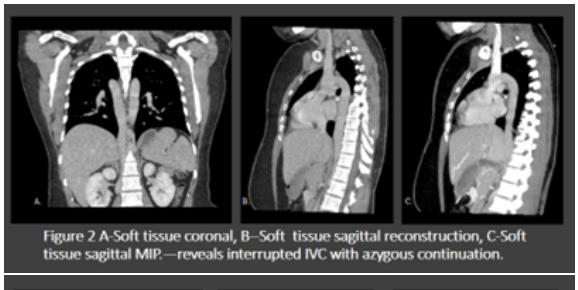

Figure 2 A-Soft tissue coronal, B--Soft tissue sagittal reconstruction, C-Soft tissue sagittal MIP.—reveals interrupted IVC with azygous continuation.

Figure 3 A-Soft tissue axial, B--Soft tissue coronal reconstruction, C-Soft tissue coronal MIP.—reveals PDPV

DISCUSSION

Polysplenia is a rare heterotaxy disorder with a reported incidence of 1 per 250 000 live births. Various studies have attempted to classify the broad spectrum of anomalies into asplenia and polysplenia(). Classical polysplenia results in hyparterial bronchi (both main bronchi are below the pulmonary arteries) and bilobed lungs. Asplenia results in trilobed lungs and eparterial bronchi (both main bronchi are located superior to the main pulmonary arteries).

In embryonic life, the venous blood from the primitive gut consists of two vitelline veins of the yolk sac. These two vitelline veins are connected by three interconnecting veins (cranial, middle, caudal). With further development, caudal and cephalad interconnecting veins, along with the caudal segment of the right and cephalic segment of the left vitelline veins, regressed. Only the middle interconnecting vein remained at the dorsal aspect of the duodenum, and formed an S-shaped PV from cephalad segment of the right and caudal segment of the left vitelline veins (). Anomalous regression of the cranial and middle interconnecting veins, which connected the two vitelline veins that run on either side of the duodenum in embryo result into preduodenal portal vein. The residual caudal interconnecting vein runs ventral to the duodenum and results in an anomalous position of the portal vein().

In the pediatric population, PDPV can be presented as duodenal obstruction, intestinal malformation or congenital heart disease (,). Patients without cardiac anomalies may reach adulthood, and account for 10-15% of cases of polysplenia(). In most adult cases, the diagnosis of the anomalies is made preoperatively or incidentally. In our case, the preduodenal portal vein and polysplenia syndrome was diagnosed incidentally while examining a complaint of persistent cough. It has been reported that PDPV increases risk to the patient during surgery, especially when involving the biliary tract. Tearing of the vein could result in serious hemorrhage().

Short pancreas, agenesis or hypogenesis of the 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 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 develops in the dorsal mesogastrium. Blood supply disturbances 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 : A, total agenesis of the dorsal pancreas; B, hypogenesis of the body and tail; C, hypogenesis of the tail. Our patient is a case of type B pancreas hypoplasia—().

Clinical significance of dorsal pancreatic agenesis is the development of pancreatitis owing to poor drainage from the remnant ventral duct. CT demonstration of a short pancreas is not synonymous with agenesis and is a pitfall well avoided. Fat replacement in the distal pancreas can mimic agenesis. Also partial *vs* complete dorsal duct agenesis can only be differentiated by pancreatic duct studies using either MRCP or endoscopic retrograde cholangiopancreatography.

Cross sectional imaging such as CT or MRI of the abdomen may be the first modality used to detect these anomalies in adults, preoperatively or incidentally (,). In comparison, MDCT with three-dimensional reconstructions including MIP and volume rendering are much more useful in demonstrating the various venous and organic anomalies.

In our case, PDPV and short truncated pancreas with polysplenic syndrome was diagnosed incidentally CT. Conventional axial and reconstruction images more clearly demonstrated other manifestations of polysplenic syndrome such as a truncated pancreas , interrupted IVC with azygos continuation of the IVC , bilobed lungs with bilateral hyparterial bronchi.

Our literature review disclosed variable rate of azygos continuation of IVC accompanied with PDPV ranging from 8% and 15%(). Also some radiologic reports revealed that abnormalities of the IVC-agygos system were more frequently associated with PDPV than previously thought. The advent of MDCT with reconstruction and MRI seems to improve the detection rate of the association between PDPV and IVC anomaly(,).

CONCLUSION.

Based on this case, we concluded that MDCT with reconstruction images is a useful and noninvasive diagnostic method for depicting PDPV. The diagnosis of polysplenia in adults is usually made during investigation for unrelated causes. Increased awareness of such anatomical anomalies would prevent serious complications during abdominal surgery.

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