

A Classic Case Of Polysplenia Syndrome With A Pancreatic Mass And SOLs In Liver

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Abstract

The ultrasound and CT appearance of a classic case of polysplenia syndrome is presented with an overview of associated anomalies. Note was made of a pancreatic mass in the same patient with multiple space occupying lesions (SOLs) in the liver.

INTRODUCTION

The normal viscerovascular asymmetrical arrangement is called 'situs solitus' (situs in Latin means position and solitus means customary). 'Situs inversus' is the mirror image of situs solitus. In between the spectrum of situs solitus and inversus lies 'situs ambiguus'^{1,2}. It comprises of patients in whom right or left-sided structures predominate. Patients with right-sided predominance typically lack a spleen, whereas patients with left-sided predominance typically have a segmented spleen or multiple splenules.

We have recently seen a patient of polysplenia with a classic spectrum of associated anomalies and multiple space occupying lesions (SOLs) in liver.

CASE HISTORY

A 35 year old male patient presented with icterus and abdominal (right hypochondrial) pain for two months. The patient was treated in a primary health centre for the same but his condition progressively worsened and he was then referred to our hospital.

Transabdominal ultrasound revealed a normally positioned liver with multiple SOLs and central intrahepatic biliary radical dilatation. The pancreas was small and the body and tail of pancreas were not visualized. There was a heterogenous mass in the visualized head and uncinata process of the pancreas. The spleen was normal in size and position and showed multiple echogenic septae.

Contrast enhanced CT scan was thereafter performed which revealed an interesting set of findings. Liver was enlarged with multiple, hypodense, rounded, varying-sized lesions. There was central intrahepatic biliary radical dilatation [Fig

1]. The body and tail of pancreas (derivatives of dorsal pancreatic bud) were absent with a heterogenous mass in the existing head and part of uncinata process (derivatives of ventral pancreatic bud) [Fig 2]. Multiple splenules were seen in the left upper quadrant [Fig 3]. A preduodenal portal vein was noted, however no obvious duodenal stenosis was present [Fig 4]. Also, there was intestinal nonrotation with the small bowel lying on the right side of abdomen and the large bowel on left [Fig 5]. The hepatic segment of inferior vena cava was interrupted and its azygous continuation noted in the form of a large retrocrural vessel on right side (the azygous vein) [Fig 1]. The kidneys and adrenals were normal.

The chest radiograph of the patient was normal.

The FNAC of the pancreatic mass could not be performed as the liver function of the patient was deranged with an INR of 3. To our misfortune, the patient then left the hospital against medical advice and was lost to follow up.

Figure 1

Figure 1- CECT of the abdomen shows multiple, varying-sized SOLs in the liver with intrahepatic biliary radical dilatation. Note is also made of interruption of the hepatic segment of IVC in the same patient and its azygous continuation seen in the form of a large retrocrural vessel on right side.

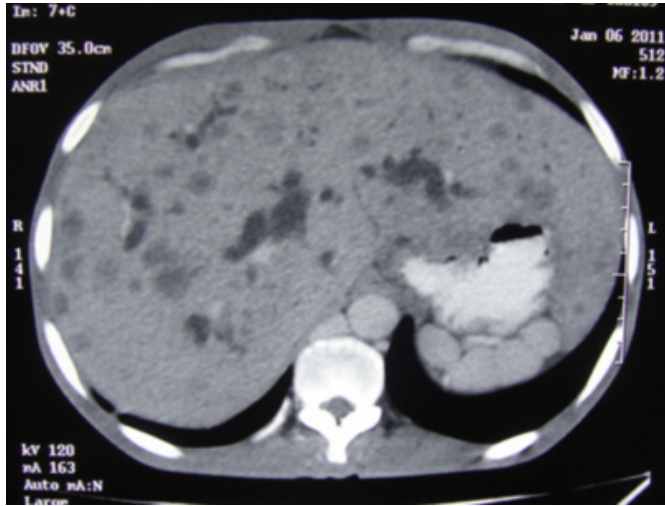


Figure 2

Figure 2- CECT image showing absent body and tail of pancreas and a heterogenous mass in the existing head and part of uncinate process.

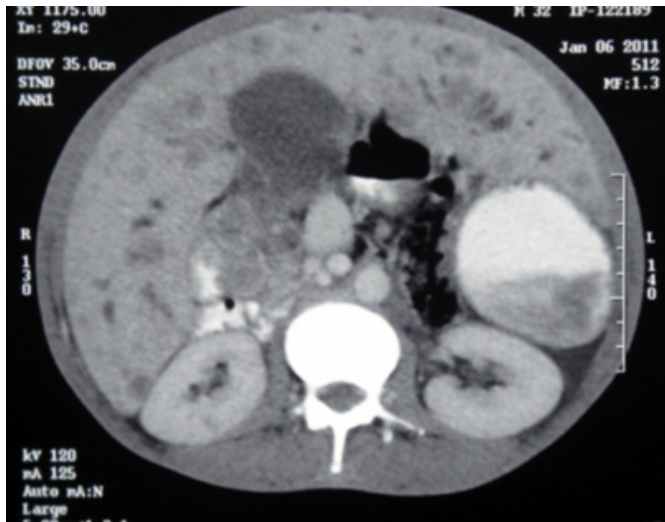


Figure 3

Figure 3- CECT abdomen shows multiple splenules in the left upper quadrant

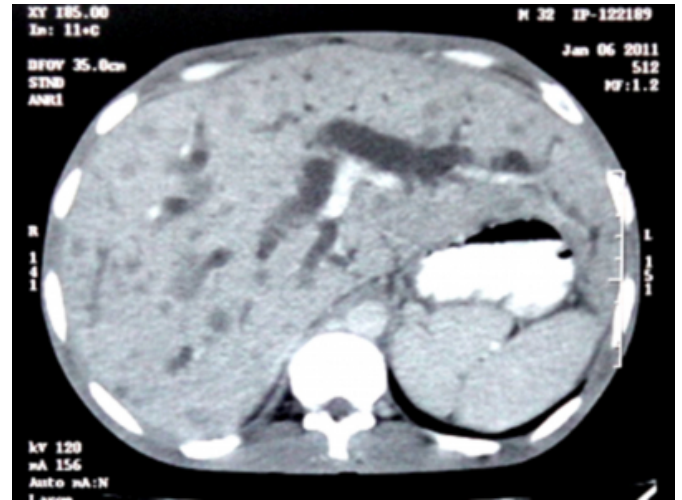


Figure 4

Figure 4- CECT abdomen showing a preduodenal portal vein apart from other features like liver SOLs and multiple splenules.

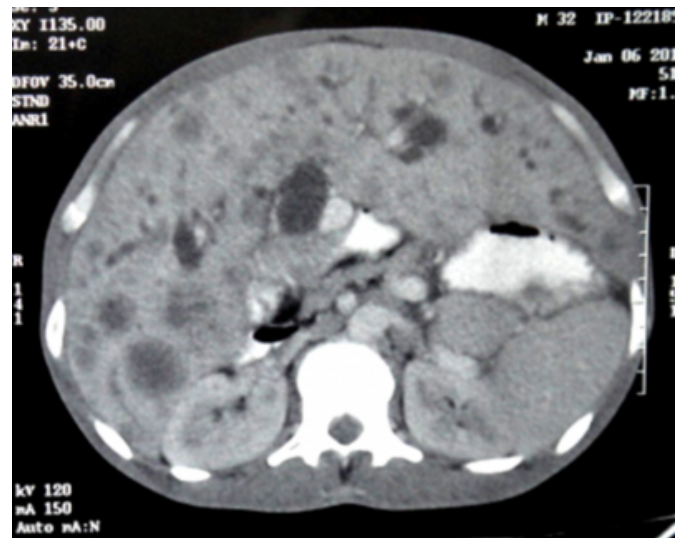
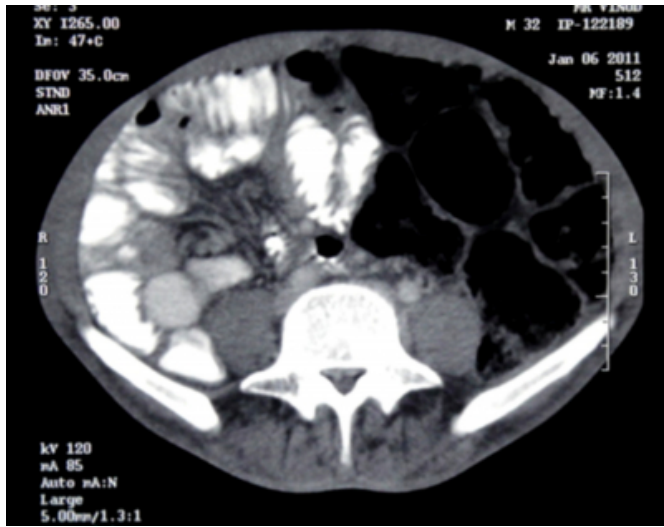


Figure 5

Figure 5- CECT abdomen image showing intestinal nonrotation with the small bowel lying on the right side of abdomen and the large bowel on left.



DISCUSSION

There are estimates that 1.44 infants per 10,000 are affected by situs ambiguous (heterotaxy)³. Polysplenia occurs in an estimated 4 per million liver births in the United States⁴. The control of human somatic asymmetry is still unknown. However, several hypotheses have been proposed. Genetic mutations (in the ‘inv’ gene), reverse embryonic turning and asymmetrical expressions of genes have been identified as the causes of the same. Familial situs ambiguous has been related to both autosomal and X-linked inheritance patterns, although most cases arise sporadically^{5,6}.

Patients with polysplenia may have splenules on both sides of the dorsal mesogastrium or a single spleen which has one or more septae.

Such patients have a variety of intestinal malrotation anomalies ranging from nonrotation to reversed rotation and faulty peritoneal attachments. Duodenal atresia or stenosis and biliary atresia can also be associated⁷.

Few patients may also have a short pancreas as a result of maldevelopment or agenesis of dorsal pancreatic bud¹. One similar case of 53 year old woman with polysplenia has also been reported to have carcinoma in the existing ventral pancreas⁸. A midline adrenal gland and renal anomalies have also been reported.

Interruption of inferior vena cava has been reported in as many as 50-60% of patients with polysplenia. Venous return then occurs via the right or left sided azygous systems. In

patients with intact vena cava, both aorta and vena cava can lie to one side of midline or on opposite sides⁹.

Vitelline anatomy is also skewed in situs ambiguous. The normal retroduodenal course of portal vein is a result of specific resorption of segments of paired vitelline veins. When the larger portion of the left vitelline vein is resorbed, the resultant relationship of the portal vein is preduodenal.

Common cardiac anomalies in patients of polysplenia include atrioventricular septal defects. In patients with asplenia, conotruncal anomalies like truncus arteriosus, conal hypoplasia with pulmonary outflow obstruction or pulmonary atresia, and anomalous pulmonary venous connections are commoner¹⁰.

A small subset of individuals with situs ambiguous also have central nervous system anomalies like holoprosencephaly, neural tube defects and caudal regression syndrome.

Thus, in a case of situs ambiguous the associated anomalies are the major causes of morbidity and mortality.

KEY MESSAGES

The incidence of polysplenia is not well established. However, it is estimated to be as low as 4 per million liver births in the United States. It is essential to know the associated anomalies in order to ascertain the mortality and morbidity.

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