An Incidental Findings of Polysplenia Syndrome in an Adult Patient with Multiple Anomalies

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Introduction

Polysplenia syndrome is a rare congenital subtype of heterotaxy syndrome associated with various visceral and vascular anomalies. Polysplenia syndrome is characterized by presence of two or more spleens and anomalies of other asymmetric organs. It is reported incidence of 1 per 250,000 and more common in females. Here, we report the case of a 53-year-old woman who presented with polysplenia syndrome. The patient had multiple accessory spleens in right sided, dextrocardia, interrupted inferior vena cava with azygos continuation, left-sided liver and congenital short pancreas. Our patient was asymptomatic until now and were diagnosed incidentally.

Case Report

53-year-old female patient was admitted to the our internal medicine clinic with mildly abdominal pain and debility. There was no sigficant evidence in the patient’s medical history. Physical examination showed mild abdominal sensivity and dextrocardia, other findings were normal. Ultrasound examinations were performed simultaneously with physical examination. Ultrasound demonstrate the presence of liver in the upper left quadrant of abdomen and multiple nodular spleen structures in the right upper quadrant of abdomen. Abdominal and chest computed tomography (CT) examination were performed to confirm the diagnosis of polysplenia and detect the accompanied additional findings. Initial CT scan shows partial situs inversus and a right-sided heart which is concomitant to right aortic arch. Liver was the in upper left quadrant of abdomen and multiple nodular spleen structures in the right upper quadrant (Figure 1). There was enlarged azygos vein on the left side of descending aorta and vena cava superior was left side of vertebral column (Figure 2). The corpus and tail of pancreas could not be monitored, there was only head of the pancreas in left quadrant (congenital short pancreas) (Figure 3). Vena cava inferior could not be monitored also. Both renal veins drain into azygos vein. Hepatic vein emptying into right atrium (Figure 3). Accordingly, the right lung consists of two lobes and left lung consisted of three lobes. Patient is diagnosed with polysplenia syndrome.

Figure 1: The coronal and axial enhanced CT show, heart is in the right hemitoxa (dextrocardia), liver is in the left upper quadrant of abdomen (arrow), there are multiple nodular spleen structures in the right upper quadrant (thin arrow). There is enlarged azygos vein on the left side of descending aorta (arrow head) and azygos vein is on the left side of aorta in the abdominal imaging (curved arrow).
More than 40 percent of reported cases have cardiac anomalies and majority of such children can not survive very long in the first five years of life. The most common cardiac anomalies are atrial septal defect (78%), azygous continuation of the inferior vena cava (65%), ventricular septal defect (63%). Death can occur at an early age especially in case of concomitant cardiovascular abnormalities [4].

In conclusion, polysplenia syndrome is an infrequently seen disease, which may be accompanied by multiple systemic abnormalities. It is important to be familiar with this syndrome especially in order not to overlook accompanying abnormalities. Computerized tomography examinations are very helpful in the evaluation of polysplenia syndrome patients.

References