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Short Communication

Polyspenia Syndrome: A Rare Congenital Disease

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Abstract

Polysplenia syndrome is a rare congenital disease characterized by the presence of two or more spleens and the affection of other asymmetric organs. The splenic structures are separated into masses of roughly the same size, ranging in number from 2 to 6 and in diameter from 1 to 6 cm. Adult polysplenia is frequently discovered during the investigation for other reasons.

Communication

Polysplenia can be isolated or associated with other congenital malformations that are part of the polysplenia syndrome. Polysplenia syndrome is a rare congenital disease characterized by the presence of two or more spleens (Figure 1) and affection of other asymmetric organs It is more commonly identified in childhood than in maturity. It's an uncommon heterotaxy condition that affects one out of every 250 000 live births. Several studies have attempted to categorize the wide range of abnormalities into asplenia and polysplenia. Polysplenia has an underlying etiology that is unknown. it has been linked to a variety of causes, including embryonic, genetic, and teratogenic components [1,2].

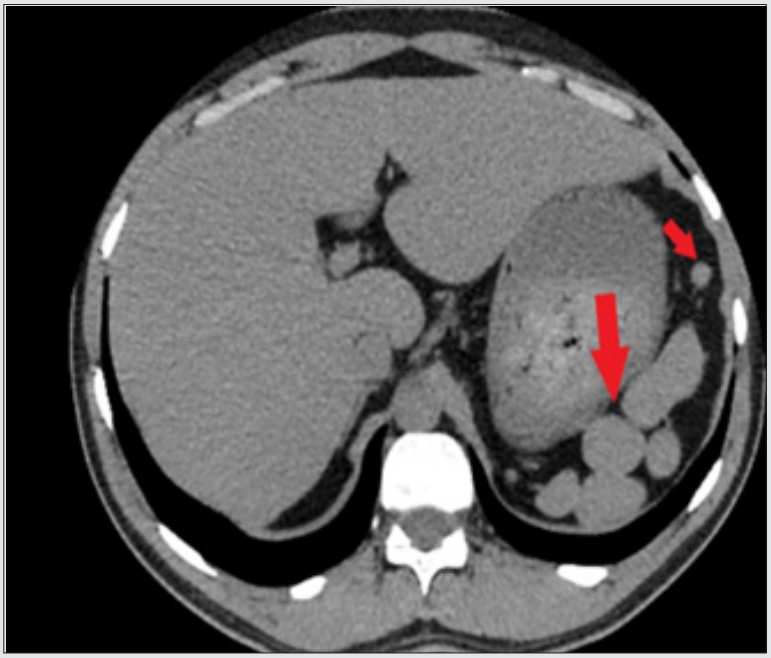

Figure 1: Abdominal CT scan in an axial section without injection of contrast medium showing in the left hypochondrium multiple nodular formations, well limited, homogeneous with clear contours (red arrows) concerning a polysplenia syndrome.

It is characterized by an abnormal arrangement of thoracic and abdominal organs. The variable involvement of these different organs results in a clinical polymorphism. Adults may experience atypical biliary and pancreatic duct discharge, cholecystitis, and intestinal blockage as a result of symptomatic polysplenia [1].

More than 40% of reported instances had cardiac abnormalities, and the majority of these children do not survive past the age of five; atrial septal defect (78 %), and azygous continuation of the inferior vena cava (65%), and ventricular septal defect are the most frequent cardiac defects (63 %) [2]. There is no specific pathognomic aberration that characterizes polysplenia syndrome, although it has a wide spectrum of abnormalities. Multiple spleens of comparable volume, visceral heterotaxia, right-sided stomach, left-sided or huge midline liver, malrotation of the intestine, a short pancreas, and inferior vena cava anomalies are only a few of the abnormalities [1]. The splenic structures are separated into masses of roughly the same size, ranging in number from 2 to 6 and in diameter from 1 to 6 cm. The splenic structures are separated into masses of roughly the same size, ranging in number from 2 to 6 and in diameter from 1 to 6 cm. The total quantity is equal to the mass of a typical spleen (Figure 1). The spleens can be found in either the left or right upper quadrant [2]. Polysplenia is often discovered by coincidence during an abdominal ultrasound or CT scan for another reason [1]. Adult polysplenia is frequently discovered during the investigation for other reasons. To summarize, polysplenia syndrome is a rare condition that can be accompanied by a variety of systemic disorders.

References

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