Polysplenia associated with semiannular pancreas

Abstract  We report CT images of an asymptomatic, adult case of polysplenia syndrome associated with semiannular pancreas, intestinal malrotation, predouodenal portal vein, and agenesis of the upper abdominal segment of the inferior vena cava with azygous continuation. This rare combination of abdominal abnormalities would be mainly caused by congenital malrotation of lower abdominal organs.

Keywords  Polysplenia · Semiannular pancreas · Malrotation · Agenesis of inferior vena cava · CT

Introduction
Polysplenia syndrome is a congenital disorder and can be accompanied with various abnormalities in thoracic and abdominal organs, including bilateral left-sidedness, cardiac anomalies, and malrotation of abdominal organs. Fifty percent of patients with polysplenia syndrome die by 4 months of age and 75% before 5 years of age, mainly with severe cardiovascular anomalies [1]. Adult patients without any symptoms usually do not have severe cardiovascular anomalies [1, 2]. CT and MR findings in patients with various combinations of anomalies have been reported [2, 3, 4, 5, 6, 7, 8]. However, although the migration anomaly is one of the main etiologies, the reason why the combinations of abnormalities is various has not been clear. In this paper, we report an asymptomatic adult case of polysplenia syndrome with a rare combination of abdominal abnormality.
Case report

A 64-year-old asymptomatic woman who had no previous medical history was found on health screening to have minimally increased serum transaminase. An abdominal CT scan showed multiple spleens along the greater curvature of the stomach in the left side of the upper abdomen (Fig. 1), a short pancreas with agenesis of the dorsal pancreas, a defect of the tail part with semianular shape of the head of pancreas (Fig. 1c–e), intestinal nonrotation (Fig. 1d,e), preduodenal portal vein (Fig. 1d,e), and agenesis of the upper abdominal segment of the inferior vena cava with azygous continuation (Fig. 1). The liver, gall bladder, and stomach were seen in the normal position (Fig. 1). The heart was of normal shape and was located in the left side of the chest on the scout view of the CT scan.

Discussion

In cases of polysplenia syndrome, various combinations of abdominal abnormalities, mainly caused by either bilateral left-sidedness or nonrotation, have been reported [1, 2, 3]. Our patient had no bilateral left-sidedness either in the chest or the abdomen. She did have agenesis of the upper abdominal segment of the inferior vena cava with azygous continuation. In addition, although there was no rotation abnormality of the liver, stomach, gall bladder, or spleen, she had serial rotation abnormalities in the pancreas and intestine. In sum, the organs located in the lower abdomen were malrotated.

The majority of patients with polysplenia syndrome die by the age of 5 years. This high mortality rate is mainly due to severe cardiac anomalies [1]. Some 5–10% of patients with polysplenia syndrome have a normal heart or only minor cardiac defects, reach adulthood without symptoms [2], and are only incidentally discovered. The patient presented here must be included in the latter category because she had no past history, no symptomatic cardiac disease, and showed no abnormality in biochemical and serological tests except for minimally increased transaminase.

To our knowledge, although CT and MRI findings of approximately 36 cases of polysplenia syndrome have been reported [2, 3, 4, 5, 6, 7, 8], only one case with a similar combination of anomalies has been published, by Soler et al. [7]. Therefore, although each abnormality itself is not rare in the case of polysplenia syndrome [1, 2, 3], this combination of abdominal abnormalities is rare [2, 3].

In conclusion, we report a case of polysplenia syndrome with an interesting combination of abnormalities caused by malrotation of lower abdominal organs in the peritoneal space.