Rare disease

Splenic infarct with polysplenia syndrome and situs inversus

Tejas Suthar,¹ Hiral Banker,¹ Mukesh Shah,¹ Gurudatt Thakkar²¹Radiology Department, VSGH, Ahmedabad, Gujarat, India;²Radiology Department, SCL Hospital, Ahmedabad, India

Correspondence to Dr Hiral Banker, heerbanker@gmail.com

Summary

A rare case of spontaneous splenic infarction with polysplenia and situs inversus is presented. The diagnosis was made by ultrasound and confirmed by CT which showed multiple aberrant right-sided spleens with situs inversus and one of the spleen showing low attenuation areas representing infarct. Polysplenia syndrome is a rare entity associated with heterotaxy syndromes, congenital cyanotic heart diseases, biliary atresia, intestinal malrotation, immotile cilia syndrome, annular or short pancreas, preduodenal portal vein and vena caval anomalies.

BACKGROUND

If we encounter a case with acute abdomen and multiple spleens, do consider a polysplenia syndrome which is associated with manier abnormalities.

And rather to treat the acute cause aggressively, the patient should be investigated thoroughly.

CASE PRESENTATION

A 12-year-old boy was admitted with right lumbar pain, fever and vomiting since last 5 days. The patient did not have history of trauma or any similar complaints in the past. On examination, an ill-defined lump, hard in consistency was palpable with significant tenderness in right lumbar region. Chest x-ray and abdominal ultrasonography (USG) were done along with other routine investigations.

INVESTIGATIONS

USG: it revealed diagnosis of situs inversus with multiple spleens in right hypochondrium and lumbar region with one of the spleen in lumbar region showed ill-defined hypoechoic area with no vascularity on colour Doppler imaging raised possibility of infarct (figure 1).

All the routine blood investigations (blood counts, renal function test, liver function test, G-6 PD) were within normal limits.

Echocardiography: it showed dextrocardia with no other anomalies.

Contrast enhanced CT: it confirmed the findings of total situs inversus with polysplenia and non-enhancing hypo dense areas in one of the spleen in right lumbar region suggesting infarct. There was no evidence of diverticula or atresia in the duodenum and proximal jejunum. Intrahepatic biliary radicals IHBR and pancreas were normal (figures 2 and 3).

OUTCOME AND FOLLOW-UP

The patient was operated thereafter and spleen with infarct was removed. Histopathology showed dilated and congested splenic sinusoids and changes of splenic infarction.

DISCUSSION

The normal position of the organs is known as situs solitus. Situs inversus is a rare congenital anomaly with an approximate incidence of 1 in 10 000 live births.^{1–4} It is a condition in which the usual positions of the organs are reversed from left to right as a mirror image of the normal anatomy. Situs ambiguous covers everything in between these two anatomic configurations on a continuum of asymmetric thoracic and abdominal organ formation. The term heterotaxy has been used to encompass these syndromes, which involve abnormal symmetry and malposition of the thoracic and abdominal organs and vasculature, including complex congenital heart and extra cardiac abnormalities.

Helwig first described heterotaxia (polysplenia syndrome) in 1929. Polysplenia syndrome (PPS) is a type of situs ambiguous characterised by left isomerism,^{5 6} confirmed by group of visceral anomalies of unknown aetiology in which multiple aberrant splenic nodules and wide range of organic malformations exist.

Figure 1 USG image showing spleens with normal parenchyma and infarction.

Figure 2 MDCT venous phase image showing situs inversus with multiple spleens in right hypochondrium.

Figure 3 MDCT venous phase image showing situs inversus and multiple spleens in right hypochondrium and lumbar region. Note one of the spleen in right lumbar region shows non-enhancing hypodense areas, that is, infarct.

The abnormalities that integrate PSS are wide⁷; the most constant being cardiac anomalies—atrial septal defect, endocardial cushion defect, bilateral left atrium, partial anomalous venous return (to the ipsilateral atrium), ventricular septal defects, double outlet right ventricle, pulmonary stenosis and interruption of hepatic segment of inferior vena cava with continuation of azygos and hemiazygos is seen in few cases. The others are biliary atresia, intestinal malrotation, immotile cilia syndrome, annular or short pancreas, preduodenal portal vein and vena caval anomalies.

Most of the patients do not reach adulthood because of serious cardiac anomalies. 5%–10% of patients that lack cardiac involvement reach adulthood.

Patients present with cyanosis, congestive heart failure or persistent jaundice in the newborn period. Those who survive present with abdominal complaints or diagnosed because of incidental finding of situs abnormalities.

In patients with heterotaxy syndrome, the spleen is almost always affected, although the reason is not clearly understood. Three types of splenic anomalies have been described: (A) the spleen may be absent; (B) the spleen may be composed of a cluster of smaller splenules, a large spleen may be accompanied by several smaller splenules, or it may be multilobed; and (c) the spleen may be of normal size but located in the right upper quadrant of the abdomen. In a study of 109 autopsies of visceral heterotaxy with congenital heart disease, 58 patients (53%) had asplenia, 46 (42%) had polysplenia, and 5 (5%) had a single right-sided spleen. The polysplenia and asplenia syndromes, each have their own associated characteristic anomalies.^{2–4}

The combination of ultrasound and CT imaging can diagnose the splenic infarct. However, the patient with polysplenia and situs inversus may need additional investigations to evaluate any other associated anomalies which are important to know preoperatively. MRI with MR cholangiopancreatography may be useful for biliary and pancreatic anomalies. An upper gastrointestinal series can help diagnose duodenal diverticulum, bowel atresia and malrotation. An ECG should be performed to rule out cardiac conduction disturbances and other cardiac rhythm disorders. Echocardiography is the main diagnostic tool for diagnosing complex cardiovascular anomalies.

Learning points

- ▶ Any patient with heterotaxy/polysplenia syndrome with acute abdomen should be aggressively investigated for structural and biochemical abnormalities that could potentially contribute to the condition.
- ▶ The finding of a surgically correctable cause of acute abdomen can lead to curative surgery.
- ▶ Other abnormalities associated with the polysplenia should be corrected early in life which are difficult to manage later because it can lead to significant morbidity and mortality.

Competing interests None.

Patient consent Obtained.

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