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Unusual association of diseases/symptoms

Hepatobiliary scintigraphy findings of a patient with polianomaly presenting with icterus

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Summary

17-days-old baby girl with continuous crying and persistent icterus attended to our hospital. Hepatobiliary scintigraphy was performed in order to diagnose aetiology of icterus and polianomaly of patient was incidentally found with scintigraphic imaging.

BACKGROUND

Hepatobiliary scintigraphy is an important technique that can identify pathologies of hepatobiliary tract. We wanted to report a baby with icterus and additional multiple malformations.

CASE PRESENTATION

A 17-days-old female infant was brought to paediatrician with complaints of continuous crying and persistent icterus. Physical examination revealed nothing but hepatosplenomegali. Laboratory analysis of patient revealed leucocytosis, thrombocytosis, direct and indirect hyperbilirubinemia and elevation of liver function tests.

INVESTIGATIONS

She was sent to nuclear medicine department for examining the aetiology of icterus. Hepatobiliary scintigraphy of patient revealed no activity passing to the gut supporting the cholestasis, but additionally on dynamic images, lateralisation of heart activity to the right side and greater spleen activity than expected. Also, some intra-abdominal activity consisted with free liquid in abdomen. Further investigation of patient revealed dextrocardia on telecardiography, and hepatomegaly, polysplenia and ascites in abdominal computerised tomography.

DIFFERENTIAL DIAGNOSIS

Hepatobiliary scintigraphy is usually used for differential diagnosis of neonatal hepatitis and biliary atresia. Biliary atresia is a condition which requires surgical intervention and can't differentiate from other conditions without biopsy.[1] Hepatobiliary scintigraphy prevents unnecessary biopsies to patients with neonatal hepatitis. Radiolabelled compounds are used to follow the distribution of bilia in the hepatobiliary tree and gastrointestinal system.[2] Hepatobiliary scintigraphy is indicated in many clinic conditions including any pathologies of hepatobiliary tree like diagnosis of biliary atresia, acute cholesistitis, biliary leak from operation site, biliary obstruction and some familial diseases.[3–7] Diagnostic sensitivity and specificity for diagnosis of biliary atresia is 100% and 75% respectively according to a recent report.[8] In this patient we couldn't recognise the biliary activity of small intestine probably because of acids. Major finding of our scintigraphy was the right disorientation of cardiac activity and gross splenic activity which were confirmed with computed imaging as dextracardia and polispleni (figure 1).

OUTCOME AND FOLLOW-UP

Biliary atresia was not confirmed by biopsy and icterus improved after medical treatment. Hepatobiliary scintigraphy changed patient management and maintained important additional information.

DISCUSSION

We present a polianomaly patient with her scintigraphy images. Scintigraphy is a necessary tool for identification of biliary atresia in infants with icterus. Hepatobiliary scintigraphy is also used for a number of diseases associated with hepatobiliary system. Although our patient had a negative hepatobiliary scintigraphy for neonatal hepatitis her outcome didn't confirm biliary atresia as well.

This anomaly is rarely presented in adulthood but there are some case reports.[9 10] There are also some unusual combinations involving aortic coarctation or patients with operation results.[11 12]

Associated interesting finding of our patient revealed a rare congenital anomaly syndrome of her: polyspleni associated with dextrocardia. This rare condition is previously presented with associated hepatobiliary problems in a case report.[13] This patient which is reported in this case report also had no activity passing to gut on hepatobiliary scintigraphy but his outcome was consisted with neonatal hepatitis.

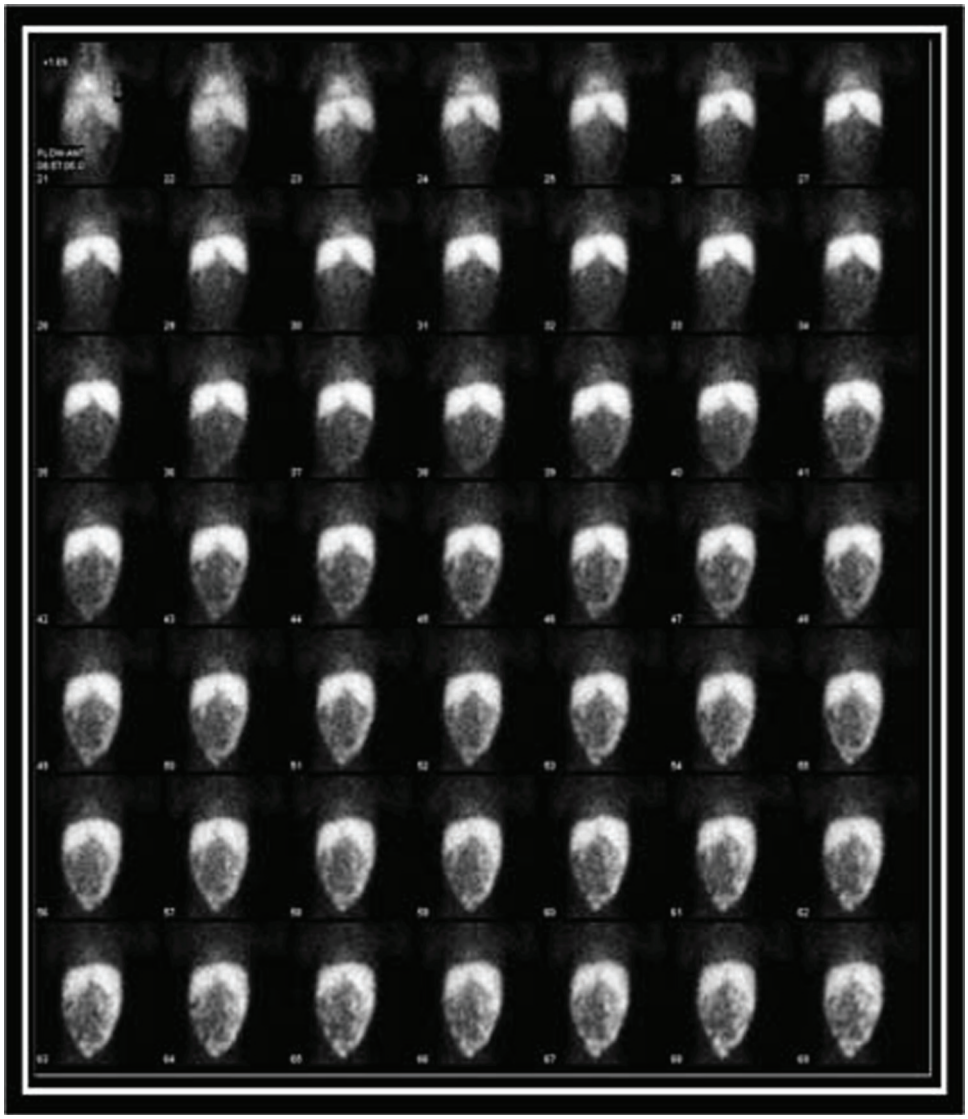

Figure 1 Anterior projection dynamic images of Tc-99m hidroksiiminodiaseticacid (HIDA) scintigraphy.

Learning points

- Hepatobiliary scintigraphy is a required method for persistent icterus
- Hepatobiliary scintigraphy has some misinterpretation causes like acites
- Congenital anomaly associated with polispleni and dextracardia may present with icterus.

Competing interests None.

Patient consent Not obtained.

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