CASE REPORT

An adolescent with hereditary spherocytosis who presented with splenic infarction

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SUMMARY

A 16-year-old male patient with known hereditary spherocytosis presented with a 4-day history of chest pain and lethargy. On admission, he had a low-grade fever and was grossly anaemic; examination revealed splenomegaly. An ultrasound scan confirmed splenomegaly with areas of splenic infarction. Subsequent tests suggested possible Epstein-Barr virus infection. The patient recovered well and had a functional spleen on discharge. This case report presents an unusual complication of isolated hereditary spherocytosis.

BACKGROUND

Hereditary spherocytosis (HS) is the commonest type of haemolytic anaemia resulting from a red cell membrane defect. Affected patients are prone to aplastic crises that can be triggered by infections, but very rarely suffer splenic infarctions.¹ In the few published cases of patients with HS and splenic infarctions, HS always coexists with another condition such as thalassaemia or sickle cell trait²⁻⁶ and rarely with infection from the Epstein-Barr virus (EBV).^{7,8} In this case, the only precipitant that can be found is EBV infection.

CASE PRESENTATION

A 16-year-old Caucasian male patient with HS diagnosed in infancy presented to our emergency department with chest pain. He reported a 4-day history of left-sided chest pain radiating to his shoulder tip with associated lethargy. He had already started a course of clarithromycin prescribed by the general practitioner for a presumed lower respiratory tract infection.

There was no other significant medical history and no previous hospital admissions with complications of HS. The patient was fully immunised, except for the pneumococcal vaccination which was not part of the childhood vaccination schedule during the patient's infancy. There was a strong family history of HS; his grandfather, mother and sister were all affected by HS.

On presentation, he had a low-grade fever and was mildly tachycardic, but oxygen saturations were 97% in room air. On examination there were no signs of respiratory distress and he was pale, but not visibly icteric. He had a tender palpable spleen measuring 8 cm from the costal edge.

INVESTIGATIONS

Initial blood tests revealed significant anaemia (haemoglobin 56 g/L) and a serum bilirubin of

52 µmol/L. He had a positive antiglobulin test with cold agglutinin disease. The chest X-ray did not show any lobar consolidation, but the abdominal ultrasound scan showed a spleen measuring up to 20 cm with a small established infarct near the inferior pole and a more acute infarct in the mid-pole region.

TREATMENT

The patient received packed red blood cells, analgesia, folic acid and oral azithromycin to treat a suspected mycoplasma infection. He was discharged after 3 days.

OUTCOME AND FOLLOW-UP

The patient presented 2 days following discharge with recurrence of his initial symptoms. On this admission, he had a temperature of 38.3°C, and was tachycardic and tender in the left upper quadrant of his abdomen. Intravenous broad spectrum antibiotics were started.

A repeat ultrasound of the abdomen showed at least three areas of wedge-shaped infarction; the largest of these measured 34×44 mm with some perisplenic fluid, particularly at the superior pole, indicating evolving infarction and necrosis (figure 1).

After a discussion among paediatricians, haematologists and radiologists, it was assumed that the patient still had a functioning spleen despite infarcted areas. His paediatrician decided to delay administration of the pneumococcal vaccination. He was discharged after 4 days of close observation.

Later we confirmed the following, a normal haemoglobin electro-phoresis, antinuclear antibody (ANA) negativity, nondetectable parvovirus IgM, mycoplasma pneumoniae levels <240, equivocal EBV viral capsid antigen (VCA) IgM, non-detectable EBV Epstein-Barr virus nuclear antigen (EBNA) IgG and detected EBV VCA/EA (early antigen) IgG. In

Figure 1 Subcostal longitudinal ultrasound view of spleen showing the largest area of splenic infarction. Measuring 34.7×44.5 mm in the superior pole.

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view of the positive Coombs test, these results make the diagnosis of EBV very likely.

DISCUSSION

A literature search, using terms 'spherocytosis' and 'infarction' yielded nine papers which detailed the reports of 10 cases of splenic infarction/sequestration in association with HS.^{1–9}

There are two other documented cases of isolated splenic infarction in patients with HS following infection from the EBV.^{7 8} One reported case is that of a 13-year-old boy with coexistent protein C deficiency.⁷ The second case is that of an 18-year-old Japanese male patient.⁸

The mechanism of splenic infarction in these cases is postulated to be as a result of an inadequate blood supply to a rapidly enlarging spleen.⁸ During EBV infection there is proliferation and expansion of EBV-infected cells. This causes a release of cytokines which along with the deposition of EBV-induced haemolysed erythrocytes cause splenic enlargement.⁸

Learning points

- ▶ Patients with known or a strong family history of hereditary spherocytosis who present with left-sided abdominal pain/lower chest pain should be investigated to rule out a splenic infarction.
- ▶ If splenic infarction is confirmed on ultrasound scan, a second pathology, including Epstein-Barr virus should be actively investigated.
- ▶ The management of the spleen will depend on the degree of infarction, but consideration should be given to prophylactic antibiotics and pneumococcal vaccine.

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Competing interests None declared.

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