

Letters to the Editor

Massive splenic infarction leading to sickle cell disease

Key words: Massive splenic infarction. Splenic abscess. Sickle cell disease.

Dear Editor,

We report the case of a female patient diagnosed with sickle cell disease following a massive splenic infarct.

Case report

A 26-year-old woman from Ecuador reported episodes of abdominal pain accompanied by anaemia, always in high-altitude places. At an altitude of 2,000 metres she began to feel general discomfort, nausea and pain in the left hypochondrium and was diagnosed with autoimmune haemolytic anaemia. Three months later, she was admitted to the emergency department of our hospital with a temperature of 38.6 °C, dysuria and pollakiuria. Her abdomen was soft on exploration and she had a painful splenomegaly of some 5 cm. The following tests were done:

- Computed axial tomography of the abdomen with oral and i.v. contrast (Fig. 1): Splenomegaly with massive splenic infarction. There were no vascular alterations to justify it.
- Blood test: Leukocytes 16,900 (82 % PMN), haemoglobin 11.5 g/dl, MCV 84, RDW 18, platelets 524,000.
- Urine: Positive nitrites, leukocytes 5-10/ field, erythrocytes 0-5/ field.

- Peripheral blood smears: Marked anisopoikilocytosis, abundant dianocytes and sickled red blood cells.
- Haemoglobin electrophoresis: Hb A2: 4 %, Hb F: 0 %, Hb S: 41 %, Hb C: 36 %.

She was consequently diagnosed with haemoglobinopathy S and C, with haemolytic crises and massive splenic infarction in the context of an acute pyelonephritis. Once the episode of acute pyelonephritis was resolved she was discharged for Haematology follow-up.

Two weeks later the patient was readmitted for a pain in the left hypochondrium and a temperature of 38 °C. Abdominal ultrasound revealed massive splenic infarction with probable superinfection. Emergency surgery was decided upon for superinfected massive splenic infarction, and a large-sized spleen was found adhering to the left liver lobe, stomach, pancreatic body and tail and abdominal wall, with old dark haematic content. A



Fig. 1. Enlarged liver, with an 18cm maximum craniocaudal diameter and a very prominent left liver lobe. Spleen: Note that the spleen is enlarged (14 cm x 9.6 cm) with no enhancement except in the form of a peripheral ring corresponding to the capsule and in some small areas of parenchyma near the hilum, findings which suggest massive splenic infarction.

regulated splenectomy was performed. During the postoperative period she developed a collection at the surgical site and a low-debit pancreatic fistula, which were resolved with radiological drainage and octeotride. The histopathological study of the surgical specimen revealed a massively necrotic splenic parenchyma with large areas of abscessification.

Discussion

Sickle cell disease is an autosomal recessive hereditary disease characterised by the presence of Haemoglobin S (HbS) (1), which when deoxygenated is less soluble and polymerises, giving rise to sickled red blood cells or drepanocytes, which become rigid and join with other red blood cells to cause vascular occlusion (2,3).

The hypoxia caused when climbing to altitudes above 2,500 m may trigger vaso-occlusive processes and give rise to infarction, the most common of which is splenic (2,4). Other precipitating factors are the postoperative period, postpartum, salmonella sepsis and strenuous exercise (5,6).

Clinical features appear suddenly and progressively, beginning with abdominal pain in the left hypochondrium, general discomfort, fever and nausea, symptoms which may cause confusion with acute mountain sickness (2).

Ultrasonography and computed axial tomography diagnose splenic infarction, and electrophoresis of the red cell haemolysis enables the haemoglobinopathy to be typified.

Initial treatment for splenic infarction must be conservative, aimed at correcting the precipitating factors and achieving an adequate oxygenation and hydration. Surgical treatment will only be necessary in the presence of complications (haemorrhage, abscess or pseudocyst) and in massive infarction when symptoms persist (7-9).

Initial management of cases of splenic abscess is with antibiotics and percutaneous radiological drainage; a splenectomy is indicated if this fails or it is multilocular (10).

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