Acute splenic sequestration crisis in a young woman with homozygous sickle cell anaemia

P.S.A. Sarma

Department of Medicine, Main Hospital and Research Centre, Bhilainager (Madhya Pradesh) 490009, India

Summary: Acute splenic sequestration crisis in a 20 year old female with homozygous sickle cell anaemia (Hb SS) is described. The resemblance of this complication to that of splenic vein ligation is discussed. This is the first case report known to the author of acute splenic sequestration crisis in an adult with homozygous sickle cell anaemia treated successfully.

Introduction

Acute splenic sequestration crisis in sickle cell anaemia was first described by Tomlinson, but has since been reported in children by several authors. It is commonly seen between the ages of 3 months to 6 years and has not been reported in adults, presumably as the spleen begins to fibrose from 8 years of age.

Acute splenic sequestration crisis has, however, been described in adults with sickle cell haemoglobin C disease8-10 and sickle cell thalassaemia.9,10 It is believed that the present case is important in emphasizing that acute splenic sequestration crisis can occur in adults with homozygous sickle cell anaemia, and that the diagnosis can be missed unless the spleen size is regularly monitored in patients with sickle cell crisis.

Case report

A 20 year old woman was admitted as an emergency with a 3-day history of fever, cough, pain in right chest and left upper abdominal quadrant in February 1987. She became drowsy, developed moderate respiratory distress and weakness of the left half of the body one day prior to admission. Homozygous sickle cell anaemia had been diagnosed 6 years previously. When not in crisis her laboratory tests showed the following values: haemoglobin 10 g/dl, haematocrit 30-33%, mean corpuscular volume (MCV) 88 fl, mean corpuscular haemoglobin concentration (MCHC) 30 g/dl, mean corpuscular haemoglobin (MCH) 27 pg, fetal haemoglobin (HbF) 5.5%, reticulocytes 2-7%, white cell count (WBC) 7.1 \times 10^9/l, platelets 180-280 \times 10^9/l, serum unconjugated bilirubin 13.6-22.1 \mu mol/l, and lactate dehydrogenase 240-270 IU/l.

On examination, she was pale, icteric, febrile (38.3°C), clinically dehydrated, tachypnoeic at rest with signs of pneumonia of the right middle lobe, and left hemiparesis. The spleen had enlarged 5 cm below the costal margin and was tender. Initial haemoglobin was 2 g/dl, WBC 36.4 \times 10^9/l (neutrophils 81%), haematocrit 6%, platelets 140 \times 10^9/l, reticulocytes 8%, serum unconjugated bilirubin 28.9 \mu mol/l, and lactate dehydrogenase 740 IU/l. Peripheral blood smear showed 330 normoblasts per 100 WBC. She received fluids, intravenous chloramphenicol and ampicillin, and 2 units of packed red blood cells.

Over the next two days, the neurological deficit disappeared, the spleen was enlarged 8 cm below the left costal margin and tender, and she remained febrile. Her haemoglobin concentration was 4 g/dl, haematocrit 16%, WBC 41.5 \times 10^9/l (neutrophils 38.5 \times 10^9/l), platelet count 150 \times 10^9/l and reticulocyte count 9%. Peripheral blood smear showed 502 normoblasts per 100 WBC. MCV, MCHC, and MCH did not alter significantly when compared with steady state. Sputum and blood cultures grew Staphylococcus aureus sensitive to the antibiotics already started. She received two more units of packed red blood cells over the next 6 days. Simultaneously her spleen extended up to 11 cm below the left costal margin, haemoglobin concentration (8 g/dl) and haematocrit value (22%) remained lower than the steady state and the reticulocyte count did not rise (7%). On day 7, she became afebrile and the normoblast count decreased to 7 per 100 WBC. Chest infection started resolving and pain improved.

At the time of discharge she was afebrile, anicteric, and her haematological and biochemical values were back to her steady state, and the spleen was not palpable. She remained well during 12 months follow-up.

Correspondence: P.S.A. Sarma, M.D., 2A/6/9 Bhilainagar (Madhya Pradesh) 490009, India
Accepted: 25 August 1988

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Discussion

Acute splenic sequestration crisis, a form of anaemic crisis peculiar to sickling disease, is characterized by acute enlargement of the spleen due to trapping of a significant red cell mass, worsening anaemia with or without thrombocytopenia, and frequently signs of hypovolaemia. Patients with low HbF levels are at greater risk of attacks of acute splenic sequestration and mean HbF values at 6 months were significantly lower in patients developing acute splenic sequestration than in those who did not.6

Topley et al.7 have suggested expanding the definition to include all cases with a decrease in haemoglobin of at least 2 g/dl and the use of normoblastemia as evidence of early response when the reticulocyte value fails to rise with worsening anaemia. Surprisingly, its onset in the double heterozygotes is delayed, although an enlarged and distensible spleen persists in half of these patients.11,12 Acute splenic sequestration crisis has been described in adults with sickle cell haemoglobin C disease8,10 or sickle cell thalassaemia.9,10 but this complication has not been reported in adults with homozygous sickle cell anaemia.

The pathogenesis of acute splenic sequestration crisis is not known. The sudden enlargement of the organ and the associated rapid reduction in blood volume and red cell mass suggest that the triggering event may be an acute obstruction to the venous flow to the spleen with consequent sequestration of red blood cells and often platelets as well.10,13 Experimental ligation of the splenic vein produces a similar picture.14 Technetium-99 m/sulphur colloid scanning of the spleen during the crisis has shown splenic enlargement, almost total lack of splenic uptake or decreased uptake with intrasplenic filling defects which resolved following recovery from the crisis.10 Also relevant in this regard are the observations of Itzchak et al.,15 who demonstrated acute splenomegaly, rise in splenic vein pressure (80–600%) and decrease in arterial blood flow (25–67%) following splenic vein ligation in dogs.

Immediate transfusion of red cells, either directly or by partial exchange is the only effective treatment of an acute episode. The response, which is often prompt, is presumably due to the improved rheology of the blood and the consequent 'unclogging' of the venous cut flow from the spleen.9 The patient reported here had an impalpable spleen during 12 months follow-up after the successful treatment of an acute splenic sequestration crisis. She exhibited early normoblastemia when the reticulocyte count failed to rise appreciably with rapid worsening of anaemia during the crisis.

The haematological and clinical findings in this patient during the acute episode resemble those following experimental splenic vein ligation in animals, and can be postulated to be due to acute obstruction to the splenic venous flow with the consequent sequestration of red cells and platelets. Immediate transfusion of red cells (improvement of rheology of the blood) reversed the crisis and brought the spleen size back to that in the steady state.

Acknowledgements

I thank the Director, Medical and Health Services, Bhilai Steel Plant for permitting me to use Hospital records, and Mr J. Tirkey and Mr S.A. Pandey, technicians, for their invaluable help.

References


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*Postgrad Med J* 1989 65: 105-107
doi: 10.1136/pgmj.65.760.105

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