Acute Splenic Infarction following Air Travel in a Child with Hb SD Disease

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Key Words
Sickel cell disease · Air travel · Acute splenic infarction

Abstract
Objectives: The relative hypoxia, dehydration and immobility associated with air travel predispose sickle cell disease patients to acute splenic crises; however we are not aware of previous reports involving children with Hb SD. Clinical Presentation: We report on a previously healthy 2-year-old Kuwaiti girl who developed a sudden onset of severe abdominal pain while traveling on a commercial jet aircraft. This was followed by vomiting and fever. On examination in the emergency room she was very irritable with abdominal tenderness maximal over the left hypochondrium. The spleen was palpable 7 cm below the costal margin, and abdominal ultrasonography and CAT scan confirmed a diagnosis of acute splenic infarction. Hematological investigations showed that the patient was a compound heterozygote for Hbs S and D. Intervention: The patient was managed with intravenous fluids and analgesics and made a full recovery. Conclusion: Acute splenic infarction should be considered in the differential diagnosis of a sudden onset of abdominal pain associated with air travel in patients with Hb SD.}

Introduction

The spectrum of sickle cell disease encompasses clinical situations where Hb S coexists with another variant of Hb with which it can copolymerize [1]. Hb D is one such variant and patients with Hb SD disease can have a clinical course comparable to SS disease [2, 3]. Most published reports have, however, been of American patients who usually carry a β0-globin gene cluster haplotype associated...
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with low Hb F and, thus, a severe disease. The coexistence of Hb D with the Saudi Arabia/India (SAI) β^a haplotype has not been reported. We have recently come across a Kuwaiti Hb SD patient who presented with acute splenic infarction following air travel. This is probably the first reported case of this complication in a child with Hb SD disease.

Case Report

B.A. is a 2-year-old Kuwaiti girl who presented in the emergency room of Mubarak Al-Kabeer Hospital, Kuwait, because of a sudden onset of abdominal pain followed by fever and vomiting of 1 day’s duration. The illness started while the family was traveling from Cairo to Kuwait in a commercial jet aircraft. She was previously well and had never been hospitalized. On physical examination, she was very irritable, lethargic and febrile (temperature of 38.5°C). She was pale, but not jaundiced, and with generalized abdominal pain and tenderness. However, the spleen was palpable 7 cm and the liver 2 cm below the costal margin. There was tachycardia and a soft ejection systolic murmur. The chest was clear and the throat was not inflamed. There was no evidence of meningeal irritation. Sepsis workup (excluding LP) was done and she was commenced on intravenous ceftriaxone in addition to intravenous fluids and analgesics. Her Hb was 8.8 g/dl, WBC 22.7 \times 10^9/l and platelets 689 \times 10^9/l. Sickling test was positive. Hb electrophoresis and, later, cation-exchange HPLC showed the following pattern: Hb F 24.2%, Hb S 28.2% and Hb D 45.5%. DNA studies showed that the patient’s β^s mutation was on a chromosome with the Saudi Arabia/India haplotype [4]. All cultures were negative and ceftriaxone was discontinued. Ultrasound examination of the abdomen revealed hyperechoic shadows in the spleen. By the 2nd day in the hospital, the abdominal tenderness was localized to the left hypochondrium and CT showed massive splenic infarction. The symptoms gradually abated and she was discharged after 1 week.

Two weeks after the acute episode, she was doing well. The splenomegaly persisted (5 cm below the costal margin), but it was no longer tender. Liver/spleen 99mTc-labeled colloid scintigraphy showed no splenic visualization. She has now been followed up for 2 years, during which she has had at least three episodes of vaso-occlusive crisis for which she was seen in the emergency room. The spleen is no longer palpable and repeat labeled colloid and denatured RBC scintigraphic studies at 1 year still showed no splenic visualization. She was administered pneumovax and Hib vaccines and is on oral penicillin prophylaxis.

Discussion

There is an increasing interest in studies of the air quality available in commercial jets [5]. These aircrafts travel in a hostile environment of extremely cold, oxygen-poor and dry air at very low pressures. Cabins are pressurized to protect passengers from the effects of hypoxia and the aim is to maintain the pressure at a maximum of 5,000–7,000 feet above sea level. However, when aircrafts fly at an altitude of up to 40,000 feet, cabin pressures reach an equivalent of 8,000 feet of altitude. Research has shown that at these reduced pressures, arterial oxygen saturation drops because the O2 diffusion mechanisms across the alveolar membranes become inefficient. Secondly, although the recommended humidity in an office is 40–60%, that in the plane is commonly less than 25%. This predisposes to dehydration which, in addition to the relative hypoxia and immobility while traveling, constitute real hazards to the sickle cell patient. It is therefore not surprising that air travel can precipitate severe splenic crises in these patients.

Acute splenic sequestration (ASS) or infarction in sickle cell states following air travel was commonly reported in the era of unpressurized aircraft travel. With the introduction of pressurization, reports on ASS have been relatively rare with only a few cases reported in Hb SC or S^β patients [4, 6]. The fact that more SS patients have not been reported with this problem is probably a reflection of the unavailability of viable splenic tissue, since most patients have autosplenectomy relatively early in life. Many Arab SS patients however have elevated Hb F levels even as adults and...
retain their spleen function [7]. Therefore they will appear to be at increased risk for air travel-associated acute splenic crises. There is, however, a need for a prospective study of this problem among our patients.

It is interesting that the patient in the present report was quite well with no previous symptomatology suggestive of sickle cell disease. This is probably due to the high level of Hb F consistent with her age and the fact that her β^0 mutation was on a chromosome with the Saudi Arabia/India haplotype. We are also following another patient with a similar Hb genotype who presented at the age of 4 years with ASS following a presumed viral illness. The ASS became recurrent and splenectomy was carried out after the third episode. Prior to this operation, splenic colloid scintigraphy showed good uptake in this patient. It is therefore important to be on the lookout for acute splenic crises in Arab children with Hb SD disease.

References