

A Traveler to Holland With a Benign Killer: A Case Report of a Sickle Cell Trait Male with Splenic Infarction

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ABSTRACT

Sickle cell trait is highly prevalent in Saudi Arabia. However, most of the patient are asymptomatic, only few present with symptoms of the complications of the disease. We report a case of 24-year-old male who presented to the hospital with history of dull left upper quadrant, nausea and vomiting while he was on the airplane. Investigation showed a picture of sickle cell trait with hemoglobin S of 40%. Peripheral blood smear was within normal. CT scan of the abdomen confirmed the presence of splenic infarction. This case emphasized on the need to increase the level of threshold to recognize the rare complications of sickle cell trait and how to manage them appropriately.

Keywords: altitude; sickle cell trait, splenic infarction.

INTRODUCTION

We labeled a patient as sickle cell trait when he inherited heterogenetic genes of hemoglobin Hb A. The percentage of Individuals with SCT is high, especially in equatorial Africa and in African-Americans. It reached up to 50% in some enclaves of Africa and up to 10% in African-Americans [1-3]. 20% of the eastern province population of Saudi Arabia are SCT. The sickle cell trait in general lives normally like other people [4]. However, Sickle cell trait is not completely benign state. usually the trait remains a silent condition. But when the SCT person leaves his original place and went to higher altitudes, where high oxygen is needed, he may face many complications one of them is the splenic infarction [5]. We reported here a case of sickle cell trait patient complaint of upper abdominal pain of 3 weeks duration after a trip to Holland. In all patients, the diagnosis of splenic syndrome secondary to SCT gene after exposure to high altitude were made retrospectively.

CASE REPORT

M. M. is a 24 years-old Saudi gentleman who was referred to the hematology clinic at King Fahad Hospital of the University complaining of upper abdominal pain for three weeks duration after a trip to Holland. He was in his usual state of health till one night prior to his trip when he started to do some strenuous exercise in the form of heavy weight lifting and treadmill jogging. He slept for eight hours then headed to the airport thereafter. The flight was a seven hours trip. He had a heavy meal served on the flight then he napped for almost four hours. Eventually, he was woken-up from his sleep because of a sudden onset, severe, generalized abdominal pain, more pronounced on the left upper quadrant. The pain was constant, dull-aching in nature, and radiating to his left shoulder. It was associated with nausea and he vomited almost four times. The

vomitus was food content with no history of hematemesis. During the flight, he noticed some change in his urine color. It became deep yellow-red. He also had a history of dyspnea along the onset of the pain. He tolerated the pain till landing. After landing he sought medical advice at a private hospital's emergency department where he was managed with intra-venous fluid hydration, given some analgesics and was discharged home after eight hours in good condition with the impression of a gallbladder disease. He stayed at Amsterdam for four days. During his stay, he had an on and off left upper quadrant abdominal pain, the pain was partially relieved by analgesics, but it was triggered by deep breathing, coughing, and laughing. He arrived back to Saudi Arabia on the 23rd of October 2013. He immediately visited King Fahad Hospital of the University. In the emergency department, routine laboratory work-up was done and he was given analgesics. His past medical history was significant for an appendectomy three year back, and a septorhinoplasty three months back with no complications. He also had a family history of sickle cell trait mother and four siblings. He was on high protein diet and shakes as a part of a fitness program. He has been a cigarette smoker for the last seven years, two packs per day. Regarding his occupation, he was a heavy field worker. He was discharged home from the emergency department with an appointment follow up at the hematology clinic.

At the clinic, he was complaining of mild left upper quadrant abdominal pain. On examination, he was vitally stable. He looked well, not pale or jaundiced. Systemic examination was unremarkable apart from abdominal examination.

Abdominal examination showed tenderness in epigastric and left hypochondriac area upon

palpation. Traubes' area was dull on percussion. No ascites was noted. Bowel sound was positive.

Investigations were done. Complete blood count showed; white blood cells of 6100 cell/mcL, hemoglobin was 15.5 g/dl, mean corpuscular volume was 87.4 and platelets of 171000/mcL. Reticulocyte count was 1.8%. Differential white cells showed neutrophils of 65.8%, lymphocytes of 26%, eosinophils of 1.4%, basophils of 0.5% and macrophages of 6.3%. A peripheral blood smear was done and was within normal limits.

Renal function test showed blood urea nitrogen of 12 mg/dL, creatinine of 1.0 mg/dL, sodium of 142 mEq/L, potassium of 4.8 mEq/L, chloride of 105 mEq/L, carbon dioxide of 31 mEq/L, and an anion gap of 6.0. Liver function test showed; total bilirubin of 0.9 mg/dL, direct bilirubin of 0.2 mg/dL, total protein of 7.9 g/dL, albumin of 4.5 g/dL, alkaline phosphatase of 95 U/L, aspartate aminotransferase of 28 units/L, alanine aminotransferase of 38 units/L, LDH of 293 U/L, and a GGT of 36 U/L. C-reactive protein was 0.4 mg/dL. serum lipase and serum amylase were within normal.

Urinalysis showed; specific gravity of < 1.005, color was straw and a trace of blood was found in his urine. Protein, nitrite and leukocytes were negative. Serum haptoglobin was 12.7 mg/dL.

Hemoglobin electrophoresis showed; hemoglobin A1 54.6%, hemoglobin F 3.6%, hemoglobin S 40%, and hemoglobin A2 1.8%. Coombs' test was negative. blood cultures and viral studies were insignificant.

Abdominal ultrasound and abdominal computed tomography were done. Patient was diagnosed as a case of sickle cell trait with acute splenic syndrome secondary to high altitude.

Axial contrast-enhanced abdominal CT scan of the patient showing hypodensity of the posterior affected splenic tissue indicative of splenic infarct.

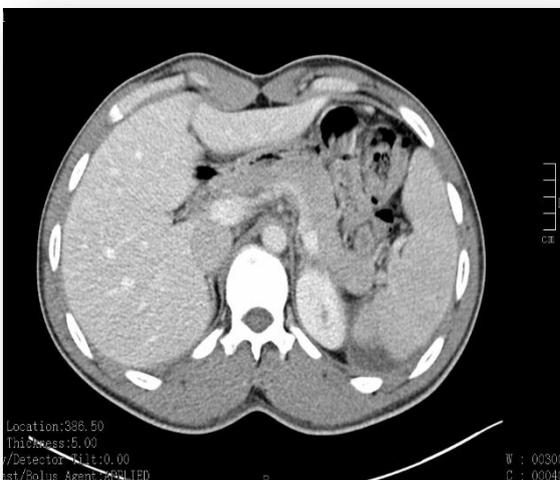


Figure 1:

DISCUSSION

In 1954, Cooley, Peterson, Engle and Jernigan published the first case report of splenic infarction in sickle cell trait after unpressurized aircraft travel [6]. Soon after that in 1955, Smith and Conley described more cases of splenic infarction that was proved in six out of eleven sickle cell trait patients [7].

Splenic infarction in sickle cell trait patient remains a rare complication but there have been more than 50 reported cases since 1954 and most of them were due to high altitude and vigorous exercise like in our patient. The altitudes that have been reported in the previous reported cases range between 1,117 m and 5,000 m (3,663 feet and 16,400 feet, respectively) [8].

hypoxemia is one of the contributing factor to splenic syndrome. Significant dropping in the alveolar partial pressure of oxygen (pO₂) that is happened at high altitude result in increase in the polymerization and subsequently sickling of red cells, leading to increased viscosity, occlusion of microvasculature, and subsequent infarction [9]. Percentage of Hb S > 40% is also potential threshold for initiating sickling process [10]. And this agrees well with the finding in our patient (Hb S of 40%). It is more common in non-black men [11]. In the other hand, autosplenectomy - which happen early in life in patient with Hb SS - may give them some protection in future [12]. This is more suggested by the small number of cases that have been reported.

In most cases, splenic infarcts are mild and self-limited; however, in its most dramatic form, patients present with acute splenic syndrome, characterized by a triad of:

- Severe abdominal pain begins soon after travel to a high-altitude place, initially in the epigastrium or the upper abdomen, but later localizes to the left upper quadrant.

- Splenomegaly

- Left upper quadrant tenderness. It may be associated with fever, anorexia, nausea, and vomiting.

Signs of peritoneal irritation, rebound tenderness, guarding, and rigidity may occur. Left pleural effusion and atelectasis develop in many of these patients.

Increased levels of bilirubin, serum lactate dehydrogenase, reticulocytosis, and anemia may be seen. Chest X-ray study may show a left pleural effusion or possibly pulmonary infiltrates [13-15].

The preferred investigation used in those cases is Contrast-enhanced CT (CECT). The classical finding would be pyramidal, wedge-shaped hypodensity of the affected infarcted splenic tissue. Sometimes, it may show Localized mottling (hyperdense) suggesting hemorrhagic infarction in hyper acute

infarct stage. And in the acute form it may present atypically as Multiple, irregular, heterogeneous lesions of patchy enhancement or total splenic infarction (as in splenic torsion) or infarction of splenunculus^[16]. The majority of sickle cell trait patients can be successfully managed conservatively by Adequate hydration, Supplemental oxygen, Analgesic therapy^[15].

CONCLUSION

Although splenic infarction is a well-recognized rare complication of sickle cell trait. The presence of risk factors such as high altitude and hypoxia should raise a high index of suspicion for diagnosis. Early diagnosis is crucial to manage the patient appropriately and prevent any complications. Physicians and patients should be aware as well as well-educated on this syndrome in addition to the patients.

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