ABSTRACT

Sickle cell trait occurs as a result of inheritance of a normal haemoglobin gene (HbA) from one parent and a mutated sickle haemoglobin (HbS) gene from another. Unlike the sickle cell disease, it is considered a benign condition in itself. However, major vaso-occlusive complications have been reported in association with Sickle cell trait, the acute chest syndrome being the most important one. We report the case details of a 32 years male who presented with features of acute chest syndrome and was later found to have sickle cell trait.

INTRODUCTION

It is estimated that sickle cell trait affects around 100 million people worldwide. It occurs as a result of inheritance of a normal haemoglobin gene (HbA) from one parent and a mutated sickle haemoglobin (HbS) gene from another. Though considered a benign condition in itself, major vaso-occlusive complications have been reported in association with Sickle cell trait.

Acute chest syndrome (ACS) is defined as a newly appearing pulmonary infiltrate on chest X-ray, affecting the lower lobes, or a perfusion defect demonstrable on a lung radioisotope scan in a patient with sickle-cell disease. It is clinically associated with fever and or respiratory symptoms.

In this case report we present details of a 32 years male who presented with features of acute chest syndrome and was identified to have sickle cell trait.

CASE REPORT

A 35 years male, the eldest among his three siblings, was admitted to Chitwan Medical College, a tertiary care centre in central Nepal, with complaints of fever for five days, chest pain and shortness of breath for two days. There was past history of recurrent jaundice from early childhood. He claimed to be a reformed smoker, and occasional drinker, the last alcohol intake being more than a year ago.

On enquiry, there was history of jaundice in his younger sister who had passed away 3 years prior with ailment associated with shortness of breath, chest pain and body pain.

On examination, He was icteric and pale. Blood pressure was 120/60 mm Hg. Pulse Rate was 96 beats per minute. Respiratory rate was 20/min.

The blood picture at presentation was suggestive of microcytic hypochromic anaemia with thrombocytopenia. The laboratory reports at the time of presentation are enumerated below (Table 1-3).

Work-up for Malaria, Dengue, Leptospirosis, Scrub typhus, Hepatitis A virus, Hepatitis E Virus, Blood/ urine cultures all returned negative. Kidney function tests were normal. CT Scan of Chest was indicative of Collapse-consolidation of postero-basal segments of both lungs, bilateral moderate pleural effusion, minimal ascites and diffusely calcified splenic parenchyma.
Echocardiography showed dilated LA, Moderate to severe AR, and mild MR with no regional wall motion abnormality. We sent for Haemoglobin electrophoresis which was suggestive of sickle cell trait- with 42% of HbS and 58% of HbA (Figure 1).

He received total of 5 units of packed RBCs for falling haemoglobin- likely due to haemolysis.

He was managed with IV antibiotics, analgesics among other supportive care. Hydroxyurea was added after haematology consultation once patient’s condition stabilized.

**DISCUSSION**

Vaso-occlusive crises are fairly common in patients with Sickle cell disease. However, in patients with sickle cell trait these occurrences are rare and hence it is considered to be a benign condition. We carried out a comprehensive literature search to look for the available case reports/ series related to vaso-occlusive events in patients with sickle cell trait. We found a few reports on Acute Chest Syndrome in such patients.

The age at presentation varied between 25 to 54 years, our patient’s being 35 years. Almost all of such cases have been reported in patients of Afro-American origin while two were reported in hose with Mediterranean ancestry. We couldn’t find any such case reports from our region during our comprehensive internet search.

Out of these reported cases the percentage of HbS varied from 26 to 45%, a finding which was similar in our patient (42%).

Almost all the reports, except one, reported normal levels of haemoglobin, a finding quite in contrast with that seen in our patient, which might have been seen as a result of sepsis related haemolysis or sickling related destruction. Clinical presentation was heralded by chest pain in almost all patients, including ours, while one case report failed to show association with chest pain while nothing was mentioned of chest pain in one of the reports. Similarly, radiological evidence of multi-lobar involvement was apparent in almost all the reported cases, including in our patient while one report failed to show such association while yet another one mentioned nothing about radiological appearance.

Among the reports which mentioned the precipitating factors of such vaso-occlusive crisis, most were either due to respiratory infections, operations or accident. In our patient it was probably precipitated by a respiratory infection. Outcome-wise, most of the patients, including ours, survived while 3 of the patients reportedly succumbed to the insult.

**CONCLUSION**

To conclude, while sickle cell trait is mostly a benign condition, there have been cases with severe vaso-occlusive events. Most cases usually carry a good prognosis while in a few deaths have occurred. Clinical presentations and precipitating factors may be variable and diagnosis is confirmed with haemoglobin electrophoresis. Early diagnosis in patients with relevant clinical and epidemiological attributes combined with imaging findings could be lifesaving.
REFERENCES:


