# SPLENIC INFARCTION: an intriguing and important cause of pain abdomen in high altitude

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**Background:** Patients with Sickle cell trait (SCT) are usually asymptomatic. They are usually unaware of their condition unless they have a family history. There are specific situations, where these people suffer from the effects of sickle cell trait. Splenic syndrome at high altitude is one of the specific problems. It is usually seen after a patient with SCT has been inducted to high altitude like in case of mountaineers and military personnel deployed in high altitude warfare. Pain abdomen due to splenic infarction in individuals with SCT is one of the manifestations. These patients, if diagnosed in time, they can be spared from unnecessary surgical interventions. We present herewith our experience of splenic infarction due to SCT in high altitude and their management.

# Keywords: Splenic; infarction; Sickle cell phenomenon; Sickle cell trait; High altitude.

# **INTRODUCTION**

Pain abdomen occurring in troops inducted into high altitude areas (HAA) can be attributed to a number of pathologies. One such intriguing and not much studied phenomenon is splenic infarction (SI). SI is an interesting and important condition usually seen in individuals with sickle cell trait (SCT) and only rarely with sickle cell phenomenon (SCP). The reason for this apparent paradox is that patients with SCP usually become symptomatic in childhood with 'sickling crises' and other complications related to the disease. However in patients with sickle cell trait, the disease may be identified for the first time when they are exposed to the hypoxia of high altitude. This also raises the important issue of whether all troops being inducted into high altitude should be screened for SCT. This study discusses the management of 05 male patients, who presented with acute upper abdominal pain and were diagnosed to have SI. It also deliberates on the management strategies.

### MATERIALS AND METHODS

Over a period of 2 years and 3 months in a hospital located in a high altitude area (13,000 feet), 05 patients of splenic infarction were diagnosed and treated. The most common time of presentation was within 72 hours of induction into the high altitude (HA) area. All patients had been inducted by air. All patients presented with acute left upper abdominal pain with radiation to the tip of the left shoulder.

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### RESULTS

There was accompanying anorexia, nausea and vomiting and subsequently fever depending upon the severity of the condition. The pain was aggravated on deep breathing and coughing. The details of clinical presentations are as depicted in Table-1.

#### Table 1 Clinical Presentations

Case No.	Age (Years)	Induc tion to HA by	Onset of symptoms since induction (Within)	Symptoms
1	27	Air	12 h	Pain left upper abdomen, nausea, vomiting.
2	33	Air	24 h	Left lower chest pain, cough, upper abdomen, nausea, vomiting.
3	24	Air	72 h	Pain left upper abdomen, left lower chest, nausea vomiting.
4	29	Air	12 h	Pain left upper abdomen, left lower chest, nausea vomiting.
5	31	Air	48 h	Pain upper abdomen, nausea.

General examination showed that they were ill and febrile. All patients had tachycardia and tachypnoea. Three of the five patients had pallor. Abdominal examination revealed tenderness and guard in the left hypochondrium. There was tenderness on thumping over the left lower rib cage. Bowel sounds were sluggish in 02 patients. There was no organomegaly or free fluid. Respiratory examination revealed reduced breath sounds in the left lower quadrant in 03 patients. A provisional diagnosis of splenic infarction was entertained. However acute gastritis, gastric ulcer perforation, pleuritic pain, pneumonitis and acute myocardial infarction were kept in mind and the patients were investigated (Table 2). Routine investigations were within normal limits except leucocytosis in 02 patients and subsequently in 01 more patient. There was anemia in 02patients. ECGs were normal. Ultrasound (USG) examination

investigations prome					
Case No	Hemogram	USG-abdomen	X-Ray chest	Sickle Cell Phenomenon	Sickle Cell Trait
1	No Leucocytosis initially, later + Anemia +	No evidence of infarction or collection on 1 <sup>st</sup> USG. Subsequent USGs showed both	Pleural Effusion Lt with raised hemidiaphragm	-ve	+ve
2	Leucocytosis + Toxic granules + Anemia+	Splenic infarction with abscess formation	-do-	-ve	Not done*
3	No Leucocytosis	Small Splenic infarction with minimal collection	-do-	-ve	+ve
4	Leucocytosis + Anemia +	No evidence of infarction or collection on 1 <sup>st</sup> USG. Subsequent USGs showed both	-do-	-ve	+ve
5	No Leucocytosis	Small Splenic infarction with minimal collection	-do-	-ve	Not Done*

Table 2 Investigations profile

of the abdomen showed no obvious pathology in 02 patients who presented within 12 hours of induction into HAA. However serial follow-up USGs showed perisplenic collections and subsequently areas of infarction (Figure 1). In 02 patients there was only evidence of infarction with no significant collection (Figure 2).



Figure 1 USG showing infarction with perisplenic collection

In 03 patients there was significant peri-splenic collection requiring surgical intervention (Table 3). CT scan was not available at the centre. Chest X-rays revealed lifting up of the pleura with left sided

effusion in all the patients. There was no free gas under the diaphragm. SCP tested in all five patients was negative while SCT which could be performed in only 03 patients was positive an all the three.



Figure 2 USG showing areas of splenic infarction

All patients were initially managed conservatively keeping them nil orally, on intravenous fluids and broad spectrum antibiotics. 02 patients had only small areas of infarction and no significant effusion. They showed progressive significant improvement clinically, biochemically and radiologically without surgical intervention. The pain and fever settled down and patients became asymptomatic. 01 patient with perisplenic collection developed a perisplenic abscess which required drainage. In this patient, the spleen could be spared. The patient with perisplenic abscess started running high grade fever with severe constitutional symptoms. His general condition started worsening, TLC count was rising and there was significant left pleural effusion. An exploratory laparotomy with splenectomy was planned. At surgery, there was an abscess in the left subphrenic space to which the spleen, greater curvature of stomach, splenic flexure of the colon and left abdominal wall were densely adherence. The loculi were carefully broken and about 200 ml of pus drained. The cavity was thoroughly washed and the spleen mobilized with difficulty.

Table 3 Per op findings and treatment profile

Case No	Per op finding	Managem ent	Remarks
1	Large areas of infarction with perisplenic adhesions and a small collection	Splenecto my on Day 10	Uneventful post op recovery
2	Small area of infarction with dense perisplenic adhesions and an abscess loculated around the spleen	Drainage of Perispleni c abscess with preservati on of spleen on Day 20	Grade II Superficial Wound Infection Treated with antibiotics
3	Large areas of infarction with perisplenic adhesions and small collection	Splenecto my on Day 12	No evidence of infarction or collection on 1 <sup>st</sup> pre op USG, but subsequent USGs showed both

On inspection, about 60 percent of the spleen appeared healthy and it was decided to preserve the spleen after much deliberation (Figure 3).



Figure 3 Splenic infarction at laparotomy.

The left subphrenic space was drained. The patient had a hectic post-operative recovery but recovered completely eventually.

In the other 02 patients, splenectomy had to be performed along with the drainage of the collection (Table 3). All patients were subsequently sent to the referral hospital at a lower altitude for convalescence.

# DISCUSSION

Sickle cell disease is the most common of the clinically significant hemoglobinopathies. SCP (the hemoglobin SS homozygous state) is usually identified during childhood. Children present with anemia, stunted growth, increased susceptibility to infection, or painful crisis. On the other hand individuals with SCT (the hemoglobin AS heterozygous state) are usually asymptomatic and usually present when exposed to stress in the form of hypoxia. Because supportive care has improved, the life expectancy of patients with SCP has increased; however, it still remains significantly shortened, by 25 to 30 years. In contrast, life expectancy is not affected by SCT except rare reports of sudden death in some affected individuals while undergoing severe rigorous exertion or sudden ascent to HAA.<sup>1</sup> Hemoglobin AS red blood cells sickle at a much lower oxygen tension than do SS cells. The only clinical abnormality that occurs with any frequency among patients with sickle cell trait is painless hematuria, presumably the result of small infarcts of the renal medulla, where red cells are particularly susceptible to sickling. However hematuria was not seen in any of our patients.

The mechanism of splenic infarction in sickle cell disease is attributed to crystallization of the abnormal hemoglobin during periods of hypoxia or acidosis. The rigid erythrocyte leads to rouleaux formation and occlusion of the splenic circulation. In homozygous sickle cell disease, multiple infarcts during childhood commonly result in a scarred, contracted, auto-infarcted spleen by adulthood. Exposure to low oxygen tension, such as unpressurized airplane travel, or vigorous activity, such as skiing in high altitude locations, also can precipitate sickling and splenic infarction in individuals heterozygous for the sickle trait.<sup>2</sup> These patients can also present with deep vein thrombosis, mesenteric or portal vein thrombosis.<sup>3</sup>

Splenic Infarction presents with acute upper abdominal pain or lower chest pain with severe cough and mild to severe constitutional symptoms depending upon the severity and extent of infarction. Patients may develop associated perisplenic abscesses, pleural effusion, empyema, or splenic vein thrombosis. A differential diagnosis of all these conditions including pneumonitis and myocardial infarction should be considered.<sup>4</sup> 01 patient in our series developed an abscess which had to be drained. The diagnosis can be confirmed on the basis of USG/ CT scan. USG may not show the infarct very early on. Serial USGs help to assess the extent and progress of lesion, perisplenic collection, pleural effusion and USG guided aspiration if indicated. It carries the additional advantage of lack of radiation hazard and easy availability. CT scan is more accurate but has the disadvantage of radiation exposure and may not be available at the altitude in discussion. We did not have a CT scan at our disposal. The confirmation of SCP is done with the help of tests for sickling, including the use of 2% metabisulfite solution which is positive in the presence of hemoglobin S. Hemoglobin electrophoresis is requires to be done to confirm SCT which was possible in 03 of our patients at a tertiary care centre where the samples were sent. As discussed, SCP is rarely positive in these patients and hence metabisulfite test is negative as was in our patients also.

Splenic infarction may require splenectomy although they can be managed with conservative treatment as was done in 02 of our patients. Even at surgery, if the spleen is found viable with minimal infarcts and the indication for surgery has been to drain an abscess, splenic preservation should be attempted.<sup>2</sup>

The patients should subsequently be sent to lower altitude when they are stable and when the transfer is feasible and further ascent should be prohibited.<sup>5</sup> This was done in all our patients after their recovery. Genetic counseling should also be carried out subsequently.<sup>3</sup> Whether or not all troops ascending to HAA should undergo screening for SCT remains debatable. There is no study including large number of patients on splenic infarction in high altitude. However literature reveals study on small number of cases. Lane and Githens had published their experience of 06 cases in 1985 with total reviews of 29 cases.<sup>6</sup>

Likewise other authors have published their experience on single case to maximum 04 cases.<sup>5,7-</sup> <sup>13</sup> Though Anwar Seikha managed all the 04 cases with splenectomy, the author had concluded that all the cases could have been managed conservatively without unnecessary splenectomy.<sup>2</sup> In our series of 05 cases only 02 cases required splenectomy. Hence, this is our view that unless otherwise required splenic infarction cases in high altitude due to SCT should be managed conservatively.

### CONCLUSION

Splenic Infarction is an interesting, intriguing and important cause of pain abdomen in high altitude areas. It is seen in troops with SCT rather than SCP. Patients present with sudden severe upper abdominal pain and constitutional symptoms. Investigations may reveal leucocytosis, a raised left hemidiaphragm, area of splenic infarction with or without perisplenic collection. Each case should be managed on individual merit. Patients with small infarcts can be managed conservatively with clinical monitoring and sequential USG, spleen preservation may be attempted and splenectomy may be considered only in exceptional cases. Patients should be evaluated for SCT and should not be reinducted into HAA. The routine screening of troops for SCT remains debatable. Hydration, graded exercise and preventing HAA are important preventive measures once diagnosis is known. For the military personnel involved in high altitude warfare, presenting with left upper-quadrant pain at altitudes above 9,000 feet (>3000 m, SCT should be kept in mind as a probable cause of splenic infarction. Prompt evaluation, management and evacuation to lower altitude may hasten recovery.

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