Introduction
Amarnath shrine is a holy place of Hindus. It is situated at an altitude of 3888 m (12,756 feet) and is visited by more than 6,00,000 devotees each year. Climbing to these heights (>10,000 feet) by individuals living in plains, requires a moderate physiological adaptation. The stress of this adaptation may not be tolerable by all. In individuals with sickle cell disease, the enlarged spleen can develop splenic infarcts. As the only difference between the sickle cell trait and the sickle cell disease is the proportion of hemoglobin S (HbS) in the blood; one can assume that this complication can also occur in the trait condition. And yes, there are case reports where no cause for splenic infarction other than the trait condition could be found. The hypothesis that sickle cell trait predisposes to splenic infarction becomes stronger with our description of simultaneous splenic infarct in two members of a family. We searched the literature for the evidence of this hypothesis and tried to find the reasons for its rarity.

Case Reports

Case 1
A 55-year-old man from Ganjam (Odisha, India) presented to the casualty with a chief complaint of pain in the left upper abdomen for 4 days. The complaint started during a religious visit to Amarnath cave. The person along with his wife and son reached Baltal using air and then road route. From this place, they started a 16 km long journey on foot. Both the man and his son started to have pain over the left upper abdomen at the end of this journey; however, they managed to reach the cave on their own. Thereafter, the symptoms worsened and they were evacuated to the base. The primary treatment was started after around 6 h of onset of symptoms, and then, they were shifted to the medical college hospital. By this time, he developed fever and had vomited few times. The ultrasonography of the abdomen was performed. The spleen was found have heterogeneous echotexture with multiple irregular hypoechoic areas. A provisional diagnosis of multiple splenic abscesses was made. Due to certain personnel reasons, he was airlifted to the casualty of our hospital. At the time of arrival, his symptoms were 4 days old. He was hemodynamically stable. The abdominal examination revealed severe tenderness in the left upper quadrant. The routine blood investigations including the liver function and kidney function tests were all normal except for the leukocytosis.

Abstract
Sickle cell trait is a highly prevalent condition. It is not a disease. However, it has been associated with few rare complications. Splenic infarction is one among them. The altitude-related hypoxia is the most common predisposing factor for this. The simultaneous occurrence of this complication in more than one member of a family is so rare that possibly, it is the only second such case report. We encountered this in two members of a family, the father and his son. They were on a religious visit to a 12,756 feet high mountain cave, when they developed pain in left upper abdomen. A thorough workup including contrast-enhanced computed tomography abdomen established the diagnosis. Both recovered uneventfully on the conservative management. As it is a rare occurrence, the finding must be interpreted carefully. There is no need to screen the individuals for sickle cell trait before high-altitude travel.

Keywords: Sickle cell trait, splenic abscess, splenic infarction
The counts were raised to 21,520/µL (neutrophils – 76%, lymphocytes – 22%, monocytes – 2%, eosinophil’s and basophils – 0%) against the laboratory normal of up to 10000/µL. Based on these, a provisional diagnosis of multiple splenic abscesses was made. Intravenous antibiotics (ceftriaxone, metronidazole, and amikacin) were started. Being an uncommon finding, a repeat ultrasound at our institute was performed. The findings were again suggestive of splenic abscess. A contrast computed tomography (CT) scan was performed for planning further management. However, its findings changed the diagnosis. The spleen was enlarged and had multiple ill-defined hypodense areas in the parenchyma, suggestive of splenic infarction [Figure 1]. A final diagnosis of splenic infarct was made, and the conservative management was continued. Progressive symptomatic improvement was noticed. He was evaluated for the possible causes of the event by the cardiologist and the medical specialist. Chest X-ray, echocardiography ECG, two-dimensional ECG, coagulation profile, peripheral smear of the blood, rapid diagnostic tests for detecting malaria parasites antigens, and dengue serology were all performed. No abnormality was detected in these tests. Sickling studies also came out to be normal. With a strong suspicion of sickle cell trait as the diagnosis of exclusion, Hb electrophoresis was performed. With 29.8% HbS in the blood, the diagnosis of sickle cell trait as the cause of splenic infarction was reached.

The case was managed conservatively. The symptoms subsided completely in 10 days and counts also returned to the normal. The patient was discharged after 2 weeks from the hospital.

Case 2

The son who was 27 years old and was with his father during the journey also developed the symptoms almost simultaneously. He was also evacuated and admitted to the medical college where first ultrasound was conducted. Patchy areas of heterogeneous echogenicity were noted in the spleen. A provisional diagnosis of splenic abscess was made and then he was shifted to our hospital.

The blood reports were all normal except for the leukocytosis. The total count was 19,200/µL (neutrophils – 71%, lymphocytes – 24%, monocytes – 2%, eosinophil’s – 3%, and basophils – 0%). The repeat ultrasound gave the same conclusion. Finally, contrast-enhanced CT confirmed the splenic infarct [Figure 2]. The workup for the causes yielded no abnormality except for the sickle cell trait. The proportion of HbS was 32% in the blood.

The case was managed conservatively. The only significant difference in the clinical course of young man was the speed of recovery. He was symptom-free within 5 days of hospital stay.

Follow-up

They were followed till 3-month postdischarge. They were symptom-free and were doing fine. Thereafter, they stopped coming to the follow-up clinic.

Discussion

Sickle cell trait is common in many parts of the world. It has a prevalence of around 10% in the states of Odisha and Chhattisgarh in India.\cite{1} It is not a disease, but recently few complications have been observed more frequently with this condition. Splenic infarction is one of them.\cite{2} The association of splenic infarction and sickle cell trait was first described by Sullivan in 1950.\cite{3} This association is rare. Only 43 reported incidences are there in literature from 1985 to 2005.\cite{4} The causes are not known. A possible hypothesis can be that it requires a combination of following two factors.

A hypoxemic stress should be there

It classically happens during sudden movement to higher altitudes as in case of flights or during mountain climbing. If oxygen saturation in the blood, reaches to <85%, the RBCs of individuals with sickle cell trait may sickle.\cite{5} This leads to vascular occlusion in tributaries of splenic artery...
and subsequent infarct. The stress is so important that this problem is not described in population acclimatized for living at higher altitude. This is also rare when movement to altitude <4000 feet is made. Most of the reported incidences have occurred above 10,000 feet.

The sudden change of altitude along with physical activity during mountain travel increases the risk of hypoxemia, and hence the chances of splenic infarction.\textsuperscript{[6]}

**Probably a higher proportion of hemoglobin S predisposes to the complication**

Classically, the population with sickle cell trait has been divided into three subgroups with an average of 41%, 35%, and 28% HbS in the blood.\textsuperscript{[4]} Those populations having higher percentage of HbS has been associated with increased susceptibility to infarction. This is probably the reason behind the racial differences with respect to the complications.

Overall, splenic infarction is an uncommon condition. Sickle cell trait is one of the less possible causes of this condition. The other common causes for it are hematological malignancies, bacterial endocarditis, intracardiac thrombus, and other medical illnesses.\textsuperscript{[7]} Hence, before reaching the diagnosis as “splenic infarct due to sickle cell trait” it is mandatory to exclude the other medical causes.

Routine screening for sickle cell trait before high mountain climbing is not recommended at present. It can lead to a huge number of travelers being diagnosed as at risk, when actually they are only probably predisposed to a very rare and almost nonlife-threatening condition. Apart from waste of resources, it can lead to undue discrimination among the travelers and fear for some dreaded genetic disorder in the individual.\textsuperscript{[8]} At present, only members of the family with documented evidence of spleen infarct during high-altitude travel may be advised screening and warned against the same if found positive.

This is only the second case report where the sickle cell trait positive father and son, have had the splenic infarct, almost simultaneously. The only other case was reported in 1985 by Goldberg \textit{et al.}\textsuperscript{[9]}

Despite having such a huge population with sickle cell trait and definitely many of them were visiting high altitude areas at some point in their life, why this complication is so rare is difficult to explain. Documentation of only two familial cases over 30 years for a genetic condition which has a prevalence as high as 10% in the population may points to some gap in understanding the disease process. There may be some other factor, apart from sickle cell trait, that we do not know at present. Further reports and studies in the future may solve the issue.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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