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Splenic Infarction in a High Altitude Traveller with Undiagnosed Sickle Cell Trait

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Abstract
Splenic infarction at altitude in individuals with sick-le cell trait is a well-documented phenomenon. Initial presentation of sickle cell trait can occur at altitude as carriers of the sickle gene are usually unaffected by their sickle cell status unless the body is challenged by certain conditions such as high altitude. This report details the case of a 23 year old white male who suffered a splenic infarction at altitude prior to being diagnosed with sickle cell trait. His presentation was classic of splenic infarction and he was treated by emergency splenectomy. Evidence suggests that conservative management of splenic infarction in sickle cell trait should be first line and that splenectomy is often unnecessary. This report serves to remind clinicians working in high altitude areas or performing pre-departure medical assessments, of the dangers of travelling to altitude with sickle cell trait. It further suggests that ethnically vulnerable clients planning trips to high altitude should be offered pre-departure screening for sickle cell trait to ensure that any ensuing complications are managed appropriately, avoiding unnecessary diagnostic delays and unneces-sary splenectomy.

Case Report
In March 2007, I was the doctor on a high altitude expedition in Peru when a young man with undiagnosed sickle cell trait (SCT) suffered a splenic infarction. Although SCT-related splenic infarction at altitude is a well-documented phenomenon,[1-3] it is a relatively rare event and, therefore, this is a worthwhile case to share amongst relevant health professionals. Shortly after arriving in Cusco, Peru, a physically fit, 23 year old white male, with an unremarkable medical history, developed acute abdominal pain. He had flown from the UK in a commercial, pressurized aircraft. Cusco is situated at an altitude of 3300m. This was the patient's first trip to such a high altitude, previously he had not ventured above 1500m.
The initial symptom was a mild-moderate, continuous epigastric pain. Abdominal examination revealed a soft abdomen, tender in the epigastric region with no guarding and normal bowel sounds. Half an hour later the patient began vomiting and over the course of the next couple of hours the upper abdominal pain progressively worsened. The patient had not overtly exerted himself, having only under-taken gentle walking during the time between the flight landing and the onset of the pain. The expedition group had journeyed about an hour from Cusco by bus, so taxi transfer of the patient back to a hospi-tal in Cusco was urgently arranged.

On arrival at the hospital the patient's observations were: pulse 78bpm, BP140/80, temperature 36.5, respiratory rate 22. He was managed with oxygen, opioid analgesia and intravenous fluids. A surgical referral was made and abdominal examination at that time revealed generalised tenderness, worst in the upper abdomen with marked pain on percussion over the spleen. As time progressed the patient became mildly clinically jaundiced. The laboratory blood test results are detailed in tables 1 and 2. An ultrasound scan revealed splenic enlargement and a diagnosis of probable splenic infarction was subsequently made. A decision for surgical management was made immediately after the ultrasound scan and a total splenec-omy was completed within 11 hours of symptom onset. The patient made a good

recovery and four days post-operatively was transferred to a hospital in Lima to benefit from the lower altitude.

On return to the UK the patient was referred to a haematologist and haemoglobin electrophoresis revealed SCT with 37.9% of HbS (35-45% of the Hb is HbS in SCT). This patient was born in the UK and has some Greek ancestry (his maternal grandfather was Greek), although he was unaware of any family history of SCT or disease. Screening for other haemoglobinopathies and thrombophilias was undertaken and all results were negative. (Table 3) Histological examination of the spleen revealed extensive areas of ischaemic infarction in an enlarged spleen (19x14x9cm), with sickled erythrocytes within the clots and some of the blood vessels.

Discussion

This case was typical of the presentation of SCT-related splenic infarction: initial epigastric pain which soon localises to the left hypochondrium.[1,2,3] Vomiting is common and breathing may become shallow and difficult, often with end-inspiratory pain.[1,2,3] A left pleural effusion may also develop.[1,2]

Conservative management with analgesia, fluids, oxygen and descent has been found to be effective in the management of splenic infarction in SCT.[1,2,3] Blood transfusion may also be necessary. Use of the hyper-baric chamber may also aid management by normalising oxygen tensions. According to the literature, splenectomy is often unnecessary in SCT-related splenic infarction and can be avoided with good conservative management.[1,3] However, surgical intervention (partial or complete splenectomy) may be needed if conservative management fails or complications arise.[3,4] Haemorrhage, overwhelming sepsis and abscess or pseudocyst formation are indications for surgical intervention.[4] In this particular case if the clinicians involved had been aware of the patient's sickle cell status and of the appropriate management of SCT-related splenic infarction, greater emphasis could have been placed on the conservative management and splenectomy may have been avoided. I would therefore suggest screening individuals for SCT prior to high altitude travel. Should patients with known SCT develop symptoms consistent with splenic infarction, the diagnosis can be expedited and prompt, appropriate management initiated.

Travel and expedition companies organising trips to high altitude should provide pre-departure guidance to their clients, to ensure they are aware of the health risks associated with high altitude travel. Ideally, clients should receive a pre-departure health questionnaire that should include questions relating to ethnicity and personal/family history of sickle cell disease/trait. Doctors involved with pre-departure health assessments need to be aware of this risk associated with SCT and advise their patients accordingly. Individuals deemed at risk of SCT should be offered a screening test prior to high altitude travel. SCT carriers can then make informed decisions as to whether to take the risk of travelling to high altitude.
A careful family history and determination of et-nicity, along with knowledge of the global distribution and prevalence of SCT will aid risk assessment for SCT carriage. SCT has a wide geographic distribution: although it is most common in Sub-Saharan Africa, it is also found in people of Mediterranean, Caribbean, Middle-Eastern, Asian and Indian origin. Country specific estimates of SCT prevalence are available at: www.chime.ucl.ac.uk/work-areas/cab - last accessed December 2010.[5] Nowadays, some countries have newborn screening programmes that detect SCT, how-ever, many adults travelling to high altitude will not have undergone screening.

Quantification of the risk of splenic infarction at altitude in SCT carriers is difficult as the incidence of the phenomenon is unknown. However, patients should be advised that altitudes above 5000ft (1524m) are deemed hazardous,[3] an altitude that includes many ski-resorts around the world. Exertion increases the risk of splenic infarction in SCT.[2] There is also an association between sudden death in SCT and exertional heat illness and rhabdomyolysis.[2,3] It is therefore prudent to warn SCT carriers travelling to altitude against dehydration and over-exertion. As people with SCT are also at risk of hyposthenuria[3] (inability to concentrate the urine) they can easily become volume depleted and should be warned that urine colour is not a reliable marker of hydration status.

Conclusions

With increasing numbers of people participating in high altitude travel, it is important that travellers, tour operators and doctors are aware of this complication of SCT. Increased awareness of this phenomenon and pre-departure screening of at-risk individuals may reduce the occurrence of SCT-related splenic infarction, promote appropriate management and reduce unnecessary splenectomies.

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Table 1: Haematological laboratory results at presentation

Haemobglobin	Haematocrit	Leucocytes	Clotting Time	Bleeding Time
12g/dL* (14-18g/dL)	36% (42-54%)	8.100/mm^3 (5000-10,000/mm^3)	7min 30sec (5-10 min)	2min 30 sec (l-3min)

Reference ranges in brackets. Abnormal results marked *

Table 2: Biochemical laboratory results at presentation

Urea	Creatinine	Glucose	Alkaline phosphatase	Bilirubin	Aspartate aminotransferase	Alanine aminotransferase
75mg/dL* (25-45mg/dL)	1.5mg/dL* (0.8-1.4mg/dL)	146mg/dL (70-I lOmg/dL)	160U/L (68 - 240U/L)	4.3mg/dL* (<lmg/dL) Conjugated 0.7mg/dL Unconjugated 3.6mg/dL	I2U/L (8-40U/L)	14U/L (I0-50U/L)

Reference ranges in brackets. Abnormal results marked

Table 3: Haematological investigations (3 months post- splenectomy)

Haemoglobin (Hb)	14.7g/dL(13-17g/dL)
Haematocrit	43.5%{40-50%)
Mean corpuscular volume	93fL (84-98fL)
Mean corpuscular Hb (MCH)	3l.3pg(27.5-32pg)
Mean corpuscular Hb concentration	33.7g/dL <31-35g/dL)
Platelet Count	570* 10^{y}/L(150-400 10^{Y}/L) (post-sp1e nectomy pheno me non)
Total white cell count	9.4 lO'VL (3.5-10 IO'VL)
Thrombophilia screen (ie. factor VIlIc level, plasma antithrombin III level, protein C function estimate, plasma activated protein C resistance, factor V Leiden genotype, protein S function estimate)	All normal
Lupus anticoagulant screening	Normal
Prothrombin gene screen	Normal
International normalised ratio Activated partial thromboplastin time ratio	L I (0.8-1.2) 0.9 (0.8-1.2)
Fibrinogen level	3.7 (1.6-4.4g/L)
Blood film	Thrombocytosis and a few Howell-Jolly bodies * (post-splenectomy phenomena)
Sickle solubility test (sickle cell screening test)	Positive *
High pressure liquid chromotography	Two peaks at HbA and HbS * HbS level 37.9% *(35-45% in SCT) HbF level <(%(<!.5%)

Reference ranges in brackets. Abnormal Results marked *