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# Road Traffic Accident in a Case of Sickle Cell Disease: Death Due to the Accident or the Disease?.

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Case Report

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ABSTRACT

Sickle cell Disease a homozygous condition for the abnormal hemoglobin gene HbS, is associated with decreased life expectancy and recurrent episodes of various types of crises like hemolytic crisis, sickle crisis, sequestration crisis, vaso–occlusive crisis and aplastic crisis. Disseminated intravascular coagulation (DIC) can sometimes occur in conditions of tissue trauma and septicemia and rarely without septicemia as well. If the pathogenesis of the disease is not correctly understood and the crises were not attended properly DIC may set in leading to shock,multi-organ failure and even death. We present a case of DIC in a young female patient of Sickle cell Disease who met with a motor accident and developed multi-organ failure resulting in death. It is important to have prior knowledge about the pathogenesis of the disease and attend the crises even before the complication sets in, so that the patient's life may be saved.

## INTRODUCTION

Sickle cell disease is an inherited hemoglobin abnormality where there is production of abnormal hemoglobin, HbS because of a point mutation with replacement of glutamic acid by valine at 6$^{th}$ position of β chain [1]. This may result in polymerization of the hemoglobin and sickling of the RBC with occlusion of vascular channels when exposed to conditions of hypoxemia [2]. Vaso-occlusive crisis may be exaggerated in cases of injury and trauma which may lead to DIC, multi-organ failure and even death [3]. Here we present one such case where an 18 year old female, ~~a~~ known case of sickle cell disease, had met with a motor accident and put on treatment. She developed vaso-occlusive crisis and DIC, went into shock with multi-organ failure and succumbed.

## Case Report

A 18 year old female student, a known patient of Sickle Cell Disease sustained head injury in a road traffic accident and was admitted in a private hospital immediately, from there she was shifted to a Multi-specialty hospital on the same day. At the time of admission the patient was semiconscious and drowsy. There were no seizures and no ENT bleeding. On examination patient had bleeding from the head with contusions in the occipital and parietal regions. BP was 100/70, Pulse 88/min, Respiratory rate 25-30/min. She was attended by a neurosurgeon and all relevant investigations were done. Hb was 7.3gms/dl, TC 23,900 , DC polymorphs show shift to left with shift to left , serum electrolytes were normal. CT scan of the head showed multiple areas of hemorrhagic contusions in bilateral frontal and temporal lobes; Mild cerebral edema; Extra Dural hematoma along occipital convexity with linear undisplaced fracture of Occipital bone; Sub Dural hematoma along Falx and Tentorium. Plan of treatment was medical and the expected outcome was guarded.

The patient general condition was stable for the following three days except for occasional vomiting. On the 4$^{th}$ day the patient developed altered sensorium and seizures. Left side pupil was dilated and fixed. The patient was rushed to the OT and decompression craniotomy was performed with an intention to remove the sub dural hematoma. Intra-operatively neurosurgeons noticed profuse bleeding from the cancellous bone and middle

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meningeal artery .Brain was edematous. They could not secure hemostasis and the wound was closed. The patient developed DIC went into shock and succumbed with multi organ failure as per the history taken from the autopsy record. Autopsy was done. Autopsy revealed heavy, congested and hemorrhagic organs. Bits from lungs, liver, kidneys and spleen were received in our department for study. Sections from Lungs showed edema, hemorrhages and congested pulmonary microvasculature with studded, sickle red cells (Fig. 1). Liver showed congested sinusoids with plenty of sickle cells (Fig. 2) Sections from spleen also showed large areas of congestion with sickle cells (Fig.3) and focal areas showed calcified hemosiderotic nodules(Gamma-Gandy bodies) (Fig.4). Kidneys showed congested glomeruli (Fig.5).

Basing on the clinical, autopsy and histopathological findings the case was reported as death due to head injury with multi -organ failure following vaso-occlusive crisis and DIC.

**Figure 1: Photomicrograph of lung parenchyma showing pulmonary vasculature studded with sickle cells (H&E 400X )**

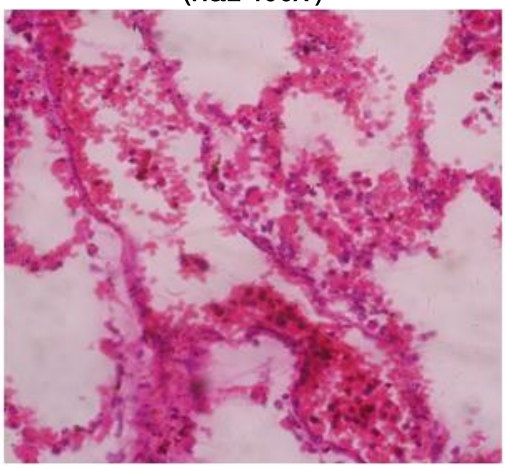

**Figure 2: Photomicrograph of liver parenchyma showing congested blood vessels studded with sickle cells (H&E 400X )**

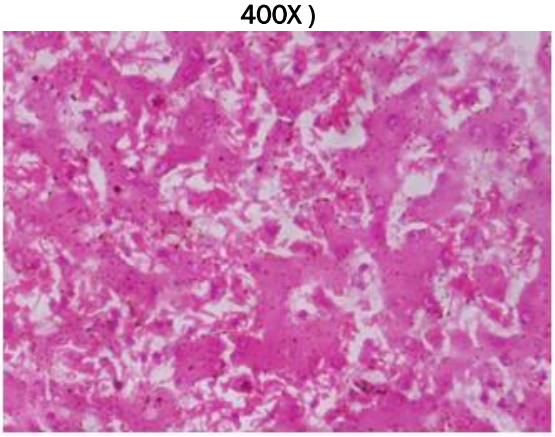

**Figure 3: Photomicrograph of spleen showing congested blood vessels studded with sickle cells (H&E 400X)**

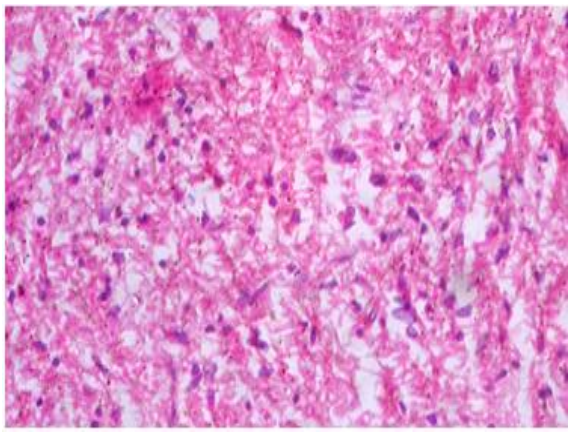

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Figure 4: Photomicrograph of spleen showing Gamma –Gandy bodies (H&E 100X )

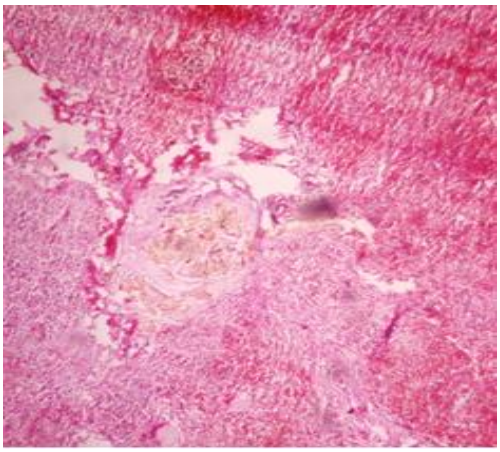
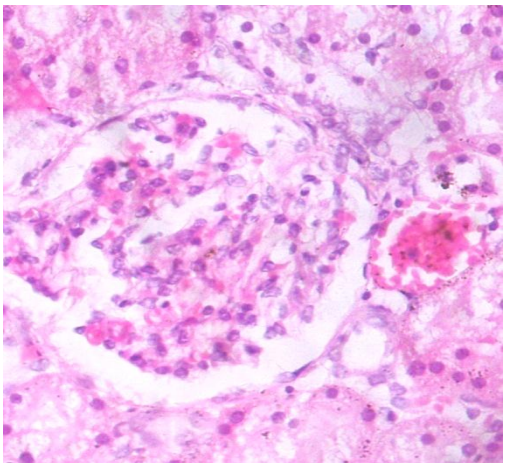


Figure 5: Photomicrograph of kidney showing congested glomeruli and blood vessels ( H&E 400X )



## DISCUSSION

Sickle cell disease is responsible for a wide spectrum of disorders that vary with respect to polymerization of HbS, degree of anemia, frequency of crisis, extent of organ injury, and duration of survival [4]. Risk factors for the development of sickle cell vaso- occlusive crisis include HbS polymerization, sickle cell deformability, sickle blood viscosity, hypoxemia, thrombus formation [4,5], fraction of dense sickle cells [6], sickle cell –endothelial cell adherence and endothelial cell activation [7]. Sickled RBC demonstrates abnormal adhesion to vascular endothelium, mostly the young RBC rather than the deformed RBC [8]. Auto–oxidation with excessive production of free radicals is associated with increased adherence [9]. Tissue necrosis factor promote the expression of adhesion receptors by the vascular endothelial cells. In case of inflammation the white cells comes into contact with the vascular endothelium and trigger the phenomenon of vaso-occlusion [10].

DIC Can occur as a result of crisis in conditions of sepsis and sometimes even in the absence of sepsis [11]. The mechanism of vaso-occlusive crisis may vary in different circumstances and in different anatomic sites as microvasculature architecture vary from organ to organ. Sometimes it becomes a challenge to the pathologist to decide whether the sickling and vaso-occlusion has occurred ante-mortem or post-mortem. On the basis of the post–mortem examination, death in sickle cell disease can be divided into three groups [12].

- Those with sufficient sickle cell related pathology to cause death
- Those in which sickle cell disease related pathology in association with unrelated pathology caused death
- Patients in whom sickle cell disease related pathology is insignificant and therefore unlikely to have caused the death.

In the present case acute blood loss and tissue trauma led to the disturbance of blood flow and hypoxemia with polymerization of hemoglobin. Increased viscosity of the blood, dense cells and sickled RBC obstructed the blood flow with stagnation of blood. Inflammation due to injury, release of free radicals and tissue necrosis factor, were

responsible for the activation of the endothelial cells to produce vaso-occlusive crisis which in turn has activated coagulation cascade to produce DIC with resultant shock, multi-organ failure and death.

To conclude it is important to have prior knowledge about the pathogenesis of the sickle cell disease and attend the crises properly even before the complication sets in, so that the patient's life may be saved.

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