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Case Report

The Post-Mortem Diagnosis of Vaso occlusive Crisis in Sickle Cell Disease in a Previously Undiagnosed Sickle Cell Disease Of An Asymptomatic Adult – A Case Report

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Abstract: Sickle haemoglobin is highly prevalent in central region of India. It has variable clinical presentation and most of the Indian patients remain asymptomatic for longer period of time. Less numbers of deaths due to sickle cell crisis are reported because of ignorance of Autopsy surgeon in considering this disease as a cause of death. While doing autopsy in case of death with no apparent cause, medical officer must keep in mind that the possibility of death due to vaso-occlusive crisis in sickle cell disease. Presenting here with a case report that highlight the importance of considering sickle cell disease as a cause of death especially in highly prevalent areas. Similarly at the time of autopsy, the role of proper histopathological sampling, haematology slides, Hb electrophoresis and toxicological analysis is considered with proper analysis of results.
Keywords: Sickle cell Disease, Vaso occlusive crisis, Autopsy.

INTRODUCTION

Sickle cell disease (SCD) comprises a group of genetic blood disorders that affect the haemoglobin molecular structure and in some cases the association with haemoglobin synthesis. In sickle cell anaemia, the Glutamic acid is replaced by Valine at the 6th position on the beta chain results in the synthesis of the abnormal haemoglobin called haemoglobin S (HbS) [1]. Several studies highlight that acute chest syndromes, cerebrovascular events, splenic dysfunction or sequestration and aplastic crises are the main causes of sudden death. Although any presentation of sickling can lead to death if one of these more serious complications when arises [2]. Recently, sickling crisis followed by bone marrow necrosis and fat embolism was shown to be associated with high index of mortality in a patient with double heterozygosis to HbS and HBc [3].

Sickle haemoglobin is highly prevalent among the tribal of central, southern and western India [4, 5] with variable frequency ranging from 10-23%. Increased prevalence is also reported in the non tribal communities of these areas. Death in clinically asymptomatic patients with sickle cell disease or sickle cell trait is not uncommon. But, unfortunately less numbers of deaths were reported due to sickle cell anaemia because of ignorance of Autopsy surgeon in considering this disease as a cause of death despite of its high prevalence.

CASE HISTORY

A 25 year old unmarried male was brought for medico-legal autopsy at MKCGMCH, Berhampur. The deceased suddenly expired while undergoing treatment for diarrhoea. As the severity of dehydration was minimal, relative of deceased demands for Post-mortem examination & suspected that this may be a case of poisoning. There was no history of prior hospitalization or treatment. Also family history of the deceased was not significant.

At autopsy on external examination an average body built, dark complexion, 168cm length, both eyes are open & shrunken, conjunctiva dry & congested, clear cornea, mouth partially open, tongue inside, Teeth 16/16 in no., food particles comes out from the mouth, nail bed bluish, post-mortem lividity on back, rigor mortis present all over the body, Micropore adherent to both cubital fossa. No other significant external findings present.

On dissection Lungs show focal oedema, on cut section spongy & no focal lesion. Heart shows concentric type of Left Ventricular Hypertrophy (thickness of 2cm), Right Ventricle thickness of 0.4cm, and Atherosclerotic changes in LAD artery. Stomach intact & contains partially digested food material without emitting any characteristics smell. In Brain, kidney, spleen & liver no gross pathology seen.

Visceral study shows no significant poison could detect in the preserved viscera & splenic smear shows no malarial parasites. Histopathology report of all organs Brain, kidney, heart & lungs show vascular congestion with sickle RBC.

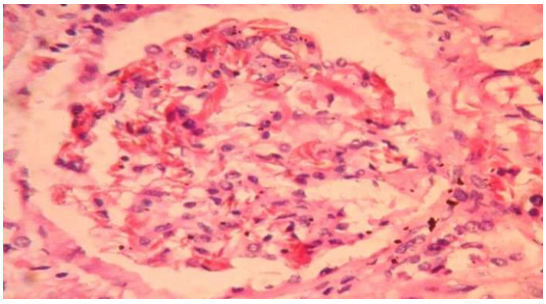

Fig-1: Photomicrograph of the Kidney showing sickle red blood cells & glomerulus

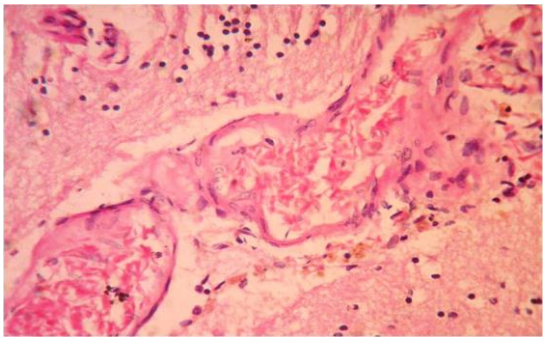

Fig-2: Photomicrograph of the Brain showing sickle red blood cells

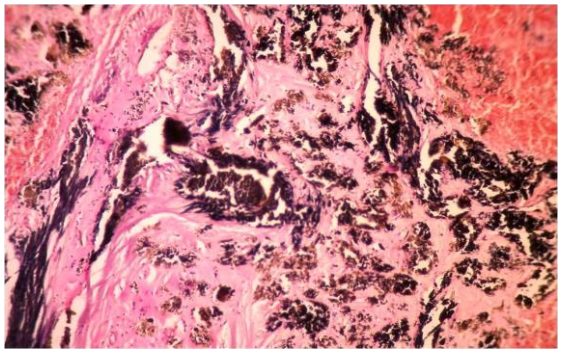

Fig-3: Photomicrograph of the Lung showing sickle red blood cells & carbon particle deposition

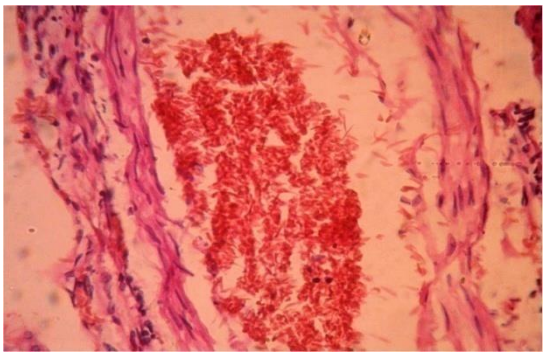

Fig-4: Photomicrograph of the Heart showing sickle red blood cells filling a medium-sized myocardial vessel

Opinion

After considering all facts & findings, Cause of death of the deceased was due to the complication arising out of sickle cell crisis which is a natural disease process further precipitate due to dehydration.

DISCUSSION

We hereby presented a case of unexpected sudden death of an adult male. The patient had a very short clinical history of illness of 24 hours. He had no clinical evidence of acute haemolysis, abdominal pain, breathlessness or anxiety to suggest sickle crisis. The recognized putative factors for sickle cell crisis include infection, fever, excessive exercise, anxiety and hypoxia, [6] which were absent in our patient. The autopsy findings of our patient did not reveal any major organ infarcts. The microscopic findings seen in this case revealed congested blood vessels packed with sickle red blood cells (RBCs) in multiple organs. Haemoglobin electrophoresis is the most common method used to diagnose SCD at autopsy. However, Thogmartin *et al.;* [7] in their study, have concluded that histological diagnosis of sickle cell can be done with the sensitivity of 95% and the specificity of 100%.

In India, SCD is seen mainly in the tribal belts of Central zones comprised of Maharashtra, Madhya Pradesh and Odisha [8]. Our patients were residents of Odisha An only meagre history were available for this patient. The patient was virtually asymptomatic and was apparently well never diagnosed as having SCD. SCD is remarkable for its clinical heterogencity. The clinical profile of sickle cell patients in India is less severe compared to that of African countries and is characterized by delayed presentation, pauci symptomatic cases, less frequency of vaso occlusive crisis and low mortality. Therefore, most of the patients remain undiagnosed [9].

Hypoxia due to exertion induces a chain of events in a person with sickle cell anaemia that causes sickling of RBC leading to vascular occlusion, potentiating hypoxia and culminating in sudden death. Similarly infection, fever, anxiety, abrupt changes in the body temperature or hypertonic dyes are precipitating

factor for sickle cell crisis [10] but, in many cases no cause is obvious. The factor precipitating sickle cell crisis was dehydration may be the triggering cause. Hence, in sickle cell disease related deaths circumstances of death as well as gross and histopathological findings must be considered [7].

CONCLUSION

We conclude that limited clinical history, inadequate evaluation of pathological findings at autopsy and a lack of laboratory resources (electrophoresis) are major shortcomings in the diagnosis of SCD in medico legal cases and cases of sudden death. Therefore, autopsy surgeon should be aware of this possibility in such type of cases. However, precise histopathological examination of tissue in the absence of other ancillary techniques serves as an important tool in diagnosing SCD at autopsy.9Hence during autopsy, circumstances of death, gross pathological findings in the organs, proper histopathology, Hb electrophoresis, molecular studies and toxicological analysis is important.

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