CASE REPORT

An autopsy case of sudden death in a patient with sickle cell disease

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Abstract An autopsy finding of sudden death due to disseminated intra-vascular sickling of RBCs in a young adult male from Madhya Pradesh while undergoing army recruitment rally, is reported because of its rarity in this part of the country.

Keywords Sudden death · Haemoglobin S · Sickle cell diseases · Rigor mortis

Introduction

The term sudden death (SD) is applied when a person previously in apparent good health falls ill and dies within minutes or at most a few hours [1]. Noninstantaneous SD is further defined as death occurring within minutes of onset of symptoms [2]. Among the cause of SD, many workers reported cardiovascular system as the principal culprit and sudden cardiac death is basically a clinical syndrome that accounts for over half of all the mortality related to ischaemic heart diseases [3, 4]. Death occurs within minutes or hours of onset of an arrhythmia, usually ventricular fibrillation and in most instances, no evidence of any recent coronary thrombosis or acute myocardial infarction can be identified as the trigger event and so there is no obvious morphologic change to explain the fatal arrhythmia. Although SD due to sickle cell diseases (SCD) is not common, it is easily identified on histology, unlike many of the clueless sudden cardiac deaths. But the exact pathogenesis leading to the death of the patient is in fact multifactorial [5]. An autopsy finding of disseminated intra vascular sickling of RBCs without any evidence of organ damage left us to ponder as to what could be the ultimate cause of death other than multiorgan failure and the case is reported for its rarity in this region of our country.

Case report

A dead body of a twenty year old male was brought to the mortuary of Regional Institute of Medical Sciences, Imphal with the recruitment of army personals. No other relevant information regarding his past illness and family history was available. On examination, the body measured 5 feet

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7 inches and weight 70 Kg. Rigor mortis was present all over the body and post mortem lividity was present on the back. The face was mildly congested with cyanosis of finger nails and both lips. There was no external injury. The dead body was presented with the history that while running during the physical training session of the military recruitment training, he collapsed suddenly and dies on the spot. Any relevant history/information regarding past illness and family history could not be elicited.

Internal examination

Head Brain was found normal except congestion.

Chest Right and left lungs weighed 650g and 640g respectively. Both lungs were congested. Heart weighed 360g. No abnormality was detected except congestion in the pericardial vessels. Coronary vessels were patent.

Abdomen Liver, spleen, right and left kidney weighed 1880g, 650g, 155g and 130g respectively.

Histopathology Several sections examined from the brain, liver, heart, spleen, lungs and kidneys revealed striking sickling of RBCs in most of the small blood vessels and capillaries. (Fig. 1) There was marked oedema of interal-

veolar septae of the lungs. The red pulps of the spleen were congested due to the trapping of sickled red cells in the splenic cords. Sickled RBCs were also present in the sinuses. Coronary vessels were patent. There was no evidence of haemorrhage or infarction in any of the organ.

Discussion

SCD is the classical prototype of hereditary haemoglobinopathy. It results from a point mutation in the genetic code such that substitution of amino acid valine for glutamine at the sixth position of beta globin chain produces HbS [6]. Disorders resulting from HbS are of enormous clinical importance because of their prevalence and world wide distribution. SCD is prevalent in high frequency in the indigenous population of certain geographical locations of India. Most affected area of sickle cell haemoglobin in India is the central zone comprising of Madhya Pradesh and Orissa. Both tribal and non tribal of these states are equally affected [7]. The disease is also prevalent in Assam, Rajasthan, Uttar Pradesh, Bihar, West Bengal, Gujarat and Maharashtra. Contrary to the fact that Sickle cell trait is normally asymptom-

Fig. 1 Photomicrograph of cerebral vessel showing sickled RBCs.(H.E stain,400X)

atic as the heterogeneous state of haemoglobin protects the RBCs from sickling, most of the complications associated with sickle cell disease have been described on individuals with sickle cell trait [8]. In most of SD due to sickle cell trait in soldiers and athletes while undergoing strenuous physical condition, the very pertinent information regarding the relative amounts of HbA, HbS and the presence of potential but undetected underlying disease is not available in the literature [6]. However, HbS gene in India and Saudi Arabia has been found to be different from its counterpart gene seen in other parts of the World [9]. Considering the high prevalence of HbS in central India of which the disease was a native, the hepatosplenomegaly and intra-vascular sickling, it is reasonable to presume that he had HbS gene, either heterozygous or homozygous. During the strenuous physical exercise, deoxygenation develop leading to polymerization of HbS and subsequent sickling of RBCs. Dehydration also stimulates more release of von Willebrand factor, decrease in the level of vasodilator substances and increase in the vasoconstrictor substances including endothelin and prostaglandins [8]. Slowing of blood flow and adhesion of RBCs to the vascular endothelium leads to further obstruction and profound deoxygenation and more sickling of RBCs [5]. Sequestration of blood in the enlarged spleen is also a contributing factor of shock and marked tissue hypoxia. Central nervous system hypoxia may produce manifestations of a seizure or stroke and despite such crisis are frequently reversible, they might become fatal [4]. Finally, obstruction to the blood flow in the vital organs leads to multiorgan failure and death. It is imperative therefore

that persons belonging to the high risk population in terms of genotype and particular geographical distribution of HbS, must undergo preliminary screening tests for sickling before undergoing any strenuous physical exercise to avert casualties and death.

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