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SICKLE CELL IN HEART; A RARE CASE REPORT

Medical Science

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ABSTRACT

Being rich in tribal communities, Jharkhand has a significant fraction of population with HbS trait. The disease has a variable presentation and most of the times the patients are asymptomatic for a long period of time. While, some patients gets crippled by repeated vaso-occlusive crisis, other have a mild symptoms. Keeping this in mind, autopsy surgeons rarely think of sickle cell disease as a cause of death in cases of death with no apparent cause, thereby making it an under-reported cause of death even in areas with high prevalence.

A case report of a male with post-mortem findings of sickle cell in the myocardial vessels is been presented here.

KEYWORDS

Sickle cell disease, vaso-occlusive crisis, autopsy, histopathology, heart.

INTRODUCTION:

Sickle cell disease is a common hereditary haemoglobinopathy caused by a point mutation in beta-globin that promotes the polymerization of deoxygenated haemoglobin, leading to red cell distortion, haemolytic anaemia, microvascular obstruction and ischaemic tissue damage.[1]

The sickle cell disease is largely seen in Indian states of southern (Tamilnadu, Andhra) south eastern (Odisha), western (Gujarat) and central (Madhya-Pradesh) region. The eastern and north eastern part of the country reveal only a minor incidence of HbS[2]. SCD is proposed to be a higher prevalence in tribal populations of central, southern and western India[3]. In Jharkhand the average prevalence in tribals is about 3% whereas it is about 13% of tribal population in Chhattisgarh[4].

Sickle cells on autopsy specimen like liver and kidneys are not rare to find but rare cases have been reported of sickle cells in heart. We present such an unusual case.

Specimen from a 28 year old male was received in department of Pathology, RIMS, Ranchi for determining the cause of death. It included heart, a portion of liver, spleen, and both the kidneys.

The deceased had been admitted in the hospital with complaints of chronic abdominal pain and had an unexpected sudden death while receiving treatment. He had never been diagnosed a case of sickle cell disease. The gross autopsy findings were unremarkable.

GROSSLY:- The heart was grossly enlarged measuring 21cm x 11 cm x 10 cm. It showed increased pericardial fat and vascular prominence on the surface. Apart from this the heart was grossly unremarkable.

MICROSCOPICALLY:- The coronary arteries showed sickled red cells and increased perivascular fibrosis.

DISCUSSION:-

Sickle red cells express higher than normal levels of adhesion molecules and are sticky. This, along with reduced deformability of red cells and sluggish blood flow in inflamed tissues result in microvascular occlusions, which are responsible for the most serious clinical features of the disease, mainly vaso-occlusive crisis manifesting frequently as hand-foot syndrome, acute chest syndrome, priapism, stroke, retinopathy etc.

Patients have a variable clinical history majority of them being aymptomatic. While some patients get crippled by vaso-occlusive crisis, others have only mild symptoms.

The clinical profile of sickle cell patients in central India is less severe in compare 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 remains undiagnosed.

Lab disgnosis is done by using an oxygen consuming agent, such as meta-bisulphite, which induces sickling in red cells if HbS is present. Haemoglobin electrophoresis is also used to demonstrate the presence of HbS and exclude other sickle cell syndromes. A pre natal diagnosis can be done by analysis of fetal DNA obtained by amniocentesis or chorionic biopsy.

The most common method of diagnosing sickle cell disease at autopsy is Hb electrophoresis, however Thogmartin et al, in their study, have concluded that histological diagnosis of sickle cell can be done with the sensitivity of 95% and specificity of 100%.

Many a times autopsy specimens of liver and kidneys show sickle cells, which points to diagnosis of SCD in such patients with vaso-occlusive crisis as a foremost cause of death.

But finding of sickle cells in heart with no other organs with such finding is rare. So this finding helped us to offer diagnosis of SCD as a cause of death.

CONCLUSION:

In case of death due to no apparent cause the autopsy surgeon must consider Sickle cell disease as a possible cause of death in areas where the disease is prevalent. Such diagnosis would help the treating physicians to think and work on SCD profile and which may save lives of many patients suffering from the disease.

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