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MALIGNANT HYPERTHERMIA IN A PATIENT WITH SICKLE CELL ANEMIA*

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Since as far as we know, there have been no previous reports in the literature describing the malignant hyperthermia syndrome (MHS) associated with sickle cell anemia, we decided to present a patient with sickle cell anemia who had developed malignant hyperthermia after receiving an anesthetic agent[1].

Case Report

A sixteen-year-old male with sickle cell anemia who had been seen at our hospital three or four times a year, had been experiencing painful crises over the last eight years.

Physical examination was unremarkable except for strabismus and the presence of hepatomegaly which was four cm below the right costal margin.

The hemoglobin level was stable at around 8 g/dl. The hemoglobin A_2 and hemoglobin F levels were two percent and nine percent, respectively. The remaining hemoglobin was Hb S. Both parents had Hb S traits.

At the age of sixteen, the patient was admitted to our hospital for surgical correction of strabismus. He was transfused until the Hb S level was fifteen percent. He was given succinylcholine and his temperature rose to 40°C within a few minutes, and surgery was cancelled. Cardiac arrest occurred, and the patient was resuscitated, cooled, and bicarbonate and prednisolone were administered intravenously. He recovered uneventfully within the following four to five hours. The serum creatinine phosphokinase level was found to be 18 units (normal: 8-12 units). A muscle biopsy revealed interstitial edema and a slight increase in fibrous tissue which were said to be characteristic of myopathy.

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Discussion

Malignant hyperthermia syndrome may develop after the use of general anesthetics such as succinylcholine and halothane[2,3]. This condition has been reported to be fatal in 20-40 percent of patients[4]. In some patients treatment with hydantoin derivatives has proven to be effective[3]. Our patient recovered within a few hours, without the administration of any specific form of therapy.

In half of the MHS cases an autosomal dominant form of subclinical myopathy has been observed. The presence of nonspecific musculoskeletal abnormalities such as ptosis or strabismus have been reported in one third of patients with MHS[5]. In addition, an elevated serum creatinine phosphokinase level has been found in some MHS patients with associated myopathy.

The association of MHS with pseudohypertrophic muscular dystrophy, the King syndrome, the Noonan syndrome, and a few malignancies have been reported[5-9]. In some cases virus-like particles have been observed in muscle biopsies, suggesting a viral etiology of the condition[10]. A slight increase in the serum creatinine phosphokinase level and changes in muscle biopsy associated with strabismus suggest the presence of an inherited susceptibility to MHS in our patient. Although there have been no reports in the literature describing a case of sickle cell anemia associated with MHS, it is still not clear whether malignant hyperthermia coincides with sickle cell anemia or if sickle cell anemia patients are susceptible to MHS.

Summary

A sixteen-year-old male with sickle cell anemia and congenital strabismus developed malignant hyperthermia a few minutes after the administration of succinylcholine, used as the general anesthetic for corrective eye surgery. The patient's hemoglobin S level was reduced to fifteen percent before the operation. He recovered uneventfully within a few hours. Increased serum creatinine phosphokinase activity and pathological changes observed in the muscle biopsy along with strabismus suggest that the patient had an inherited susceptibility to malignant hyperthermia.

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