Case report

ECAST Syndrome(Exercise collapse associated with Sickle Cell Trait): first knock of sickle cell in a Young Bodybuilder

Abstract

ECAST Or exercise collapse associated with sickle cell trait is a rare phenomenon associated with sickle cell trait and is an important presentation of sickle cell disease in sports medicine. Collapse is seen following a vigorous physical activity which is due to excessive heat, dehydration and other factors associated with physical exercise. This rare syndrome is often missed by the treating physicians as a result of lack of knowledge about this rare entity leading to massive underreporting. It is a important to identify ECAST as a cause of collapse in young athletes to prevent mortality and morbidity and in order to provide prompt treatment. We report a case of a 25-year-old young male who was a body builder and reported to the gym after one year long break due to lockdown restrictions of COVID19.After a vigorous exercise session he collapsed in the gym and was brought to the emergency department. After proper history taking and examination he was suspected to be a case of ECAST due to history of a similar episode three years back which was treated as a case of exertional syncope with intravenous fluid therapy and a family history of Sickle cell trait with his mother and father both having sickle cell AS Pattern. Ultimately our patient turned out to be a case of Sickle Cell Trait with evidence of AS pattern on Hb electrophoresis and a small sized spleen visualized on CT Scan of abdomen. The patient was managed successfully with intravenous fluids and blood transfusion and was discharged in a stable condition. He was counselled about moderating his exercise and is doing well on follow up.

Keywords:ECAST,Sickle Cell Trait,Sports Medicine

Introduction

Patients with Sickle Cell trait have a mutation in one of the two genes which code for hemoglobin molecule responsible for transporting oxygen in the red blood cells. This mutation results in change of shape of the Red blood cells from biconcave to. A crescent or sickle shaped cell upon deoxygenation. In sickle cell disease there is mutation in both the genes responsible for hemoglobin molecule formation. This causes sickle cell disease to be a more severe condition than sickle cell trait. Sickle cell trait patients usually have an uneventful course of disease with sickle cell crisis being a rare compilation unlike sickle cell disease. The stable course of sickle cell trait makes the diagnosis of sickle cell trait a difficult

one with far less episodes of crisis and hospitalization and lack of any specific presenting complaints [1].
Upon deoxygenation in a patient with sickle cell mutation there is change of shape of the Red blood cell to a sickled cell which may bock the blood vessels leading to profound damage to the muscles and organs.Sickle cell trait affects more than three hundred million individuals around the globe .Normally individuals with sickle cell trait can participate in athletic events however in rare circumstances exercise can have negative impact on their health.This includes pain,weakness of the muscle and collapse.If collapse occurs in a athlete with AS Pattern following exercise it is referred to as ECAST.It is important to identify and treat ECAST as it causes risk of death to increase by 40 times in athlets with sickle cell trait when compared to other athletes [2].Early diagnosis and treatment of ECAST prevents mortality and reduces the complicatons associated with this rare syndrome.

CASE PRESENTATION

A 25 year old young male was brought to the emergency department following an episode of collapse after an intensive work out session in the gym.The individual had previous history of similar episode three years back for which he was given intravenous fluids and was diagnosed as exertional syncope.There was no history of any bleeding tendencies in the past.There was no history of fever,cough.cold,chestpain,palpitaitons or orthopnea.There was no history of hypertension,diabetes mellitus or bronchial asthma in the past.On family history both his parents were revealed to have sickle cell AS pattern.
On general examination patient was afebrile,pulse was 88 beats per minute,regular in rhythm,normal in volume,all peripheral pulses were well felt,blood pressure was 120/80 mm hg in right arm supine position,orthostatic hypotension was ruled out,Jugular venous pressure was not raised with absent abdominojugular reflux,pallor was present and spo2 was 97 percent on room air.On systemic examination chest was bilaterally clear with equal air entry,Heart sounds were normal with no murmurs,abdomen was soft and non tender with no hepatosplenomegaly and patient was conscious,oriented to time,place and person,deep tendon reflexes were normal,bilateral plantar were flexor and there was no nuerologicaldefecit.Muscle tenderness was present in the upper and lower limbs,themsucles of the fore arm and thigh were soft and tender with a power of 4/5.
Patient was admitted and lab investigations were done and are mentioned in table number 1.Random blood sugar was 152 mg/dl and MRI Brain was done which was normal.
An ultrasonography was performed which showed small spleen which was confirmed on CT Scan of abdomen (Figure 1) and therefore hb electrophoresis was done which revealed AS Pattern.As electrolytes were normal,orthostatic hypotension was ruled out,MRI Brain was normal and heat exhaustion was also ruled out a diagnosis of ECAST was made.
Patient was managed with intravenousfluids,blood transfusion and other supportive therapy.He responded well to treatment and was discharged in stable condition five days after admission .

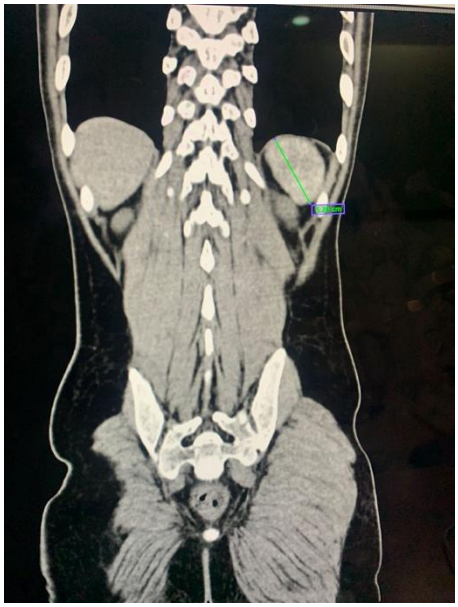

Figure 1:CT Scan showing small sized spleen suggestive of sickle cell anemia

Lab Parameter	Measured Value
CBC	Hb-6.6 gm/dl MCV-73fl Platelet count-180000/dl WBC Count-8900/dl
LFT	Total Protein-7.2gm/dl, Albumin3.6gm/dl, Globulin3.6gm/dl, aspartate aminotransferase 31 units/l , alanine aminotransferase 27 units/l, AlkanlinePhophatase102IU/l, Total Bilirubin :1.9mg/dl Unconjugated 1.2mg/dl Conjugated 0.7 mg/dl
KFT	Creatinine:1.2 mg/dl, Urea 32mg/dl, Sodium133 mmol/l, Potassium -4.3mmol/l
D-Dimer	0.45

Table 1:Showing lab investigations of the case

Discussion

Patients with sickle cell trait have one gene mutation coding for the two chains of hemoglobin leading to formation of sickle cell red blood cells in conditions of deoxygenation. CAST associated with sickle cell disease has a complex mechanism. It is unexpected to encounter collapse or death in an individual with sickle cell trait who otherwise have stable course of disease.Extremely low muscle oxygen ,raised body temperature and dehydration all contribute to sickling of red blood cells when an individual with sickle cell trait exercises [3].Also,when there is physical exertion the level of epinephrine in blood rises rapidly which makes the sickled red blood cells to become sticky and block the blood vessels. It is not clearly understood why only some individuals with sickle cell disease develop ECAST.The risk factors likely to contribute in predisposing to ECAST are intense physical activity over a long period of time, inadequate rest between intense physical activity,return to athletic activity after prolonged period of rest, high altitude and excessive exposure to heat.Other contributing factors such as increased humidity,asthma(exercise induced),fatigue due to illness and stimulants used as dietary supplements may also play crucial role in predisposing an individual to ECAST [4].Presenting symptoms of ECAST include weakness of muscle, muscular tenderness, dropping to the ground ,tachypnoea and fever(usually less than 103 degree Fahrenheit).

There are several ways to differentiate ECAST from other causes of collapse. Athletes with muscle cramps usually have tight, hard muscles which is different than soft and weak muscles of ECAST. Athletes with exertional heat stroke will have a body temperature above 104 degree Fahrenheit whereas in ECAST the temperature will be normal or slightly elevated with a conscious and oriented patient. The most definitive symptom of ECAST in increasing pain and weakness in muscles [5]. ECAST can lead to serious medical conditions such as explosive rhabdomyolysis and therefore should be diagnosed and treated promptly. Treatment of ECAST includes immediate intravenous fluid transfusion and monitoring of electrolytes as well as heart function.

Prevention of ECAST includes screening for Sickle cell trait in the athletes,provision of adequate recovery time during work out session,increasingexcerxise intensity gradually,carrying out mild physical activity routinely throughout the year,giving time for acclimatisation to altitude or temperature,proper hydration before during and post exercise.If an athlete is known case of sickle cell trait he/she should be councelled about the risk factors and symptoms of ECAST and to report to medical staff immediately in case of appearance of any symptoms.

Before an athlete returns to physical activities post an episode of collapse,he/she should be tested for sickle cell trait.Other diseases such as diabetes,electrolyte imbalance and muscle damage should also be ruled out.Training or physical activity should only be resumed slowly and under a doctor's supervision.

Our patient had history of sickle cell trait,he collapsed after intensive work out which was carried out after a long period of abstinence from physical activity due to lockdown restrictions,on examination muscle tenderness and weakness was also present.Random blood sugar was normal,orthostatic hypotension was ruled out,body temperature was normal ruling out heat exhaustion leaving behind only ECAST as likely diagnosis in our case.Our case was managed successfully with intravenous fluids and blood transfusion.A small spleen pointed towards Sickle cell trait which further confirmed the diagnosis of ECAST with hb electrophoresis showing AS Pattern.Therefore timely diagnosis and management of ECAST can yield successful results with prevention of adverse outcome.

Conclusion

ECAST is a rare phenomenon encountered in sickle cell trait individuals carrying out physical activities without proper precautions. The treating clinicans should therefore be

made aware of this rare but important consequence of sickle cell trait in order to enable prompt diagnosis and prevention of complications.

References:

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