Multiple Vertebral Necrosis in a Sickle Cell Trait: A Rare Manifestations

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Introduction

Sickle cell trait also called sicklemia is a condition in which a person has one abnormal allele of the hemoglobin beta gene and is heterozygous, but does not display the severe symptoms of sickle cell disease that occur in a person who has two copies of that allele. Sick cell trait is a hemoglobin genotype AS is generally regarded as a benign condition [1]. However, individuals with sickle cell trait are the carriers of abnormal hemoglobin and rarely may have complications. Of all the complications in sickle cell disease avascular necrosis is the commonest presentation involving the epiphysis of long bones, such as the femoral and humeral heads and the femoral condyles. Osteonecrosis of multiple vertebrae are very uncommon in sickle cell disease. And out of that osteonecrosis of vertebrae in sickle cell trait is very rare. We present a rarest case of young female of sickle cell trait having multiple vertebrae necrosis.

A Case

A young female of 35 years old presented to the medicine out patient department with the complaint of low back ache and pain while walking since last 6 months. 6 months before she delivered her second child by normal vaginal

delivery, since then the pain has started. The pain was mainly localized to the lower back with no radiation; no tingling or any sensory complaints. The pain increased while walking which has restricted her movements. She had some relief of pain only in laying down position. There was no history of fever, weight loss, loss of appetite, white discharge pervagina, trauma to back or weakness of lower limbs. On examination she was pale, unicteric, spleen 2 cm palpable, soft and non tender, paraspinal muscles spasm was present and no neurodeficit. The bilateral hip movements were painful. She was a known case of sickle cell trait with no past history of any blood transfusion or any admissions to the hospital for any crisis. Her blood reports revealed Hb-6.5 gm %, TLC-6,700/cu mm, Platelets-231,000/cu mm, MCV-77. Peripheral smear was suggestive of microcytic hypo chromic anemia. Urine gross and microscopic examination was normal. Hb electrophoresis showed 'AS' pattern. Her liver function including the alkaline phosphatase and kidney function tests were within normal limits. X ray hip joint was s/o avascular necrosis of bilateral femoral heads as shown in Fig. 1. Magnetic resonance imaging of lumbo sacral spine with contrast revealed diffuse abnormal marrow signal intensity affecting the entire axial skeleton. Lower dorsal and lumbar bone marrow necrosis with ineffective spondylodiscitis at L2–L5 level as shown in Fig. 2a–c. Fine needle aspiration cytology (FNAC) of the necrotized bone from the L4 vertebral body was taken and the report showed crenate and non crenate red blood cells, few macrophages, clusters of leucocytes and cell debris along with plasma at some places. Cytomorphology was suggestive of cystic hemorrhagic necrosis of bone probably due to sickle cell disease. No granuloma, no cessation, no malignant cells seen. In view of multiple necroses of vertebrae we retrospectively investigated this patient to rule out other causes. Blood

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culture was normal and showed no growth of any organism. Serum uric acid levels were normal. Her antinuclear antibodies (ANA) and double stranded DNA (dsDNA) test was negative for collagen vascular disease especially systemic lupus erythematosus. Her blood was negative for HIV, HBsAg and Anti HCV. After ruling the other causes of bone marrow necrosis we were forced to consider the sickle cell trait to be culprit for her severe back ache because of lumbar spine necrosis.

As per the treatment, the orthopedic surgeon provided her the supportive measures like the lumbar brace and has planned the surgery.

Discussion

Sickle cell trait considered being non disease because of its benign complications. Some of the literature derived from case reports and observational studies describes the morbidity of sickle cell trait [2]. Severe tissue hypoxia, dehydration, acidosis, increased viscosity and hypothermia increases the red cell sickling and polymerization leading to some fatal conditions like exercise related deaths, renal medullary cancer, splenic infarctions, gross hematuria, hyposthenuria, venous thromboembolism, complicated hyphema and fetal loss [3, 4]. Necrosis of the femoral head is considered to be unproven or unlikely association with sickle cell trait [5]. Osteonecrosis of femoral head is common but vertebral involvement is very rare in sickle cell disease. Osteonecrosis in sickle cell trait is unimaginable. To our knowledge after our extensive search we could find only two case reports mentioning osteonecrosis of femoral head in patient of sickle cell trait [6], where the diagnosis was delayed because the association of osteonecrosis and sickle cell trait was not recognized. Bone marrow necrosis of spine is not mentioned in the literature. Pregnancy would have precipitated or complicated the symptoms as this patient of ours developed the symptoms after her pregnancy. Severe anemia leading to hypoxia could also have contributed for her manifestation. The risks of complications in sickle cell trait during pregnancy are due to the metabolic demands, hypercoagulable state, and vascular stasis associated with pregnancy. They are at increased risk for fetal loss, low birth weight, and preeclampsia compared with women without sickle cell trait

[7, 8]. Placental abnormalities might play a role, due to frequent acute ascending amniotic infection and meconium histiocytosis as shown by Taylor et al. A prospective study carried by Larabee and Monga [8] showed that in patients with sickle cell trait the rate of preeclampsia was significantly increased (24.7 vs 10.3 %, $P = .0001$). There also was a statistically significant decrease in gestational age at delivery, lower birth weight, and an increase in the rate of postpartum endometritis.

Conclusion

In the coming years, with the emerging complications associated with sickle cell trait, it would no more be a benign condition. Especially in the pregnancy one has to take care of the patient in the same fashion as that of a sickle cell disease. We have to prevent these patients from developing dehydration, acidosis, hypothermia or severe anemia. We imply that one has to seriously notify the complaints of sickle cell trait patient and do not let them pass off easily considering it to be benign condition. The treating doctors and the patients should be well educated about all these possibilities.

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