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SCIENTIFIC LETTER TO THE EDITOR

Osteonecrosis and Leg Ulceration in Indian Sickle Cell Patients

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Sir,

Avascular necrosis of bone and cutaneous leg ulcers are the most common manifestation of patients with sickle cell disease (SCD) [1, 2]. The bone areas most frequently affected are cortical bone of the acetabulum, head of the femur, and head of the humerus. Osteonecrosis of the femoral head is common in patients with sickle cell disease. The hip and shoulder abnormalities are also seen in patients. Multifocal osteonecrosis is defined as disease of 3 or more anatomic sites. Patients diagnosed with osteonecrosis of the knee, shoulder and ankle should be evaluated for other joints for osteonecrosis. In patients with sickle cell disease, clinical complications including osteonecrosis can vary in frequency and severity, presumably due to the effects of genes that modify the pathophysiology initiated by the sickle mutation [3]. Baldwin et al. suggested genetic influence of BMP6 and ANXA2 SNPs with osteonecrosis in SCD [3]. The incidence of leg ulcers in patients with SCD ranges from 25.7% to 75% [4, 5]. Leg ulcers are a common sub-phenotype of sickle cell disease. Their cause is unknown, their prevention is impractical and their management, once present, is often difficult. Leg ulceration and avascular necrosis (AVN) in SCD has become serious problem in a few decades in India, however the incidence of leg ulcer and AVN in sickle cell patients has not been studied extensively in India till date. We had 150 sickle cell patients (60 sickle cell anemia, 75 sickle β-thalassemia and 15 sickle hemoglobin D) recruited from out patient department hematology AIIMS, New Delhi, India. Study period was 3 y. Signed consent form taken from all the patient and study was approved by institutional ethical committee. Diagnosis of patients was done by high performance liquid chromatography (Bio-Rad-Variant™Bio Rad, CA, USA). Complete blood count and red cell indices were measured by automated cell analyzer (SYSMEX K-4500, Kobe Japan). Osteonecrosis screening was done by X-ray and confirmation was done by magnetic resonance imaging (MRI) while leg ulceration was reported on physical examination. MRI is the most sensitive means of diagnosing AVN. MRI facilitates better response to treatment because with the use of MRI, avascular necrosis is diagnosed at an earlier stage and therapeutic measures are more successful the earlier they are begun. Out of 150 cases 14 (8 boys and 6 girls with mean age; 11.5±3.4 y) presented with osteonecrosis where seven were from HbSS, 6 from sickle beta thalassemia and one from HbSD patients. Nine patients presented with femoral head (5 HbSS and 4 HbSβ-thal) while 3 hip (2 HbSS and 1HbSD) and 2 (HbSβ-thal)with shoulder osteonecrosis. Seven patients (4 boys and 3 girls with mean age; 12.3±2.6 y) presented with leg ulceration out of 150 SCD patients (5 HbSS and 2 HbSβ-thal). None of the patients presented both complications *i.e.*, leg ulceration and AVN together. Observation of the study concludes the increased risk of AVN (9.34%) and leg ulceration (4.67%) in Indian sickle cell patients where alpha thalassemia and high HbF is prevalent in patients.

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