Sickle Cell Disease Presenting with Chronic Tophaceous Gout

LK Meher1, VP Tushar2, PK Hui3, SN Nayak4

A 32 year old male patient who was a known case of Sickle cell disease (SCD) presented with pain and swelling of small joints of both hands and feet for last 5 years. On general examination there was moderate pallor, mild icterus, splenomegaly (1 cm), swelling of proximal interphalangeal joints of both hands, swelling and tenderness over metatarsophalangeal joints of both feet with irregular hard nodular swellings (tophi) over dorsum of both feet (Figures 1, 2). Routine investigations revealed Hb-5.8 gm%, MCV-85 fl, MCH-28 pg, MCHC-33 g/dl, reticulocyte count-4%, DC-N(70) E(3) L(25) M(2) B(0), ESR-140 mm in first hr, TC-9800 cells/mm³, TPC-1.72 lakhs/mm³. Rheumatoid factor was negative, BUN-42 mg/dl and serum creatinine - 1.4 mg/dl with creatinine clearance of 53.57 ml/min, serum uric acid-6 mg/dl, 24 hour urinary excretion of uric acid-350 mg and liver function tests showed total bilirubin - 3.5 mg/dl, indirect-2.8 mg/dl, AST-25 IU/L, ALT-28 IU/L, ALP-110 IU/L. Hemoglobin electrophoresis showed HbS-88%, HbF-9%, HbA2-3%. X-ray of both hands and both feet showed lytic and sclerotic lesions with destruction of 1st metatarsophalangeal joint and soft tissue deposition in both feet (Figures 3, 4). FNAC from the nodular swelling over interphalangeal joint of great toe demonstrated putty like material (Figure 5) which in cytosmear showed clumps of needle shaped crystals (Figure 6). It was consistent with a diagnosis of tophaceous gout. Patient was treated with colchicines, folic acid and the acute attack subsided. After the acute attack subsided he was put on febuxostat and colchicine.

Thus, it was a clear case of sickle cell disease with tophaceous gout. Though the incidence of hyperuricemia (defined as a serum urate concentration above 0.39 mmol/l or 6.5 mg/100 mL) is high in patients with SCD,1 development of gouty arthritis and presence of tophaceous deposits is rarely documented.2-4 There are many hypothesis trying to explain this peculiar complication, but none has been proven conclusively delineating the need for further studies in this topic. It is hypothesised that this association is rare due to circulatory impairment resulting from congestion and thrombosis of small vessels, in synovia.4 This prevents white blood cells from responding to chemotactic stimulus of uric acid crystals. Activity of the polymorphs is also reduced by anaerobic condition present in SCD. Another hypothesis is that ageing and degenerative changes in joint play a major role for urate crystallisation in joint fluid, but patient of SCD don’t survive for this to occur.2 Presence of hyperuricemia in a SCD patient warrants a detailed evaluation of the renal function as it might be one of the earliest marker of deteriorating renal function.

References

1Professor, 2PG Student, 3Associate Professor, 4Assistant Professor, Post Graduate Department of Medicine, MKCG Medical and Hospital, Berhampur, Orissa

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