Gianotti Crosti syndrome associated with transfusion acquired hepatitis B virus infection in a patient of sickle cell anemia

Sir,

Gianotti Crosti syndrome (GCS) is a self-limiting disease characterized by acrally located monomorphic papular or papulovesicular eruptions and may be associated with various infections including hepatitis B antigenaemia.[1] High frequency of hepatitis B infection has been noted among patients with sickle cell disease and clinicians should be aware that jaundice in these patients may be due to hepatitis B and not only due to hemolytic crisis.[2] This case report reappraises the significance of GCS in evaluation of GCS associated infections in children and especially in patients receiving multiple blood transfusions.

A 9-year-old boy, a known case of sickle cell disease with past history of recurrent jaundice, treated with multiple transfusions, presented with abrupt onset of itchy skin eruptions and yellowish discoloration of skin and sclera of one week duration. Monomorphic, flat-topped erythematous firm papules of varying sizes were seen distributed symmetrically over the face, buttocks and limbs sparing the trunk, popliteal fossae, palms, soles and mucosae. Except for icterus and cervical lymphadenopathy, rest of the general and systemic examination was normal. A clinical diagnosis of GCS was made.

Laboratory investigations showed hemoglobin of 7.8 g/dl, PCV 22.3 and peripheral smear showed target cells, micro spherocytes and sickle cells and total leukocyte count of 6,700/mm^3. Liver function tests showed indirect hyperbilirubinemia (total bilirubin 8.6 mg/ml and direct bilirubin 2.2 mg/ml) and mildly elevated liver enzymes (ALT-757IU, AST- 507IU). Hepatitis B surface antigen (HBsAg) was detected on enzyme-linked immunosorbent assay and rest of viral markers were found to be negative. Screening of parents was negative for hepatitis B. Skin biopsy showed

focal parakeratosis, acanthosis and upper dermal perivascular mononuclear infiltrates consistent with GCS. The patient was managed with antihistamines and skin lesions cleared in two weeks. He had no recurrences in the subsequent six months of follow-up.

Episodes of jaundice in sickle cell anemia may be misleading in many cases, as it may either be part of the chronic hemolysis or due to transfusion related hepatotrophic viral infections. GCS represents a type IV cutaneous hypersensitivity reaction to various viral antigens including hepatitis B virus (HBV), Ebstein-Barr virus, Rota virus, Coxsackie virus (B and A 16), Parvo virus B19, Entero virus, Human herpes virus (HHV)-6, Parainfluenza virus, Cytomegalovirus (CMV), Adeno viruses, Poxvirus and Human immunodeficiency virus (HIV).[3,4] Among these viruses, HBV, CMV and Parvo virus B19 are quite common in patients with sickle cell disease and can be responsible for sickle cell crisis. The prevalence of HBV in sickle cell patients is 15%, as compared to 1% in control patients.[2] Features indicative of a HBV-related GCS include asymptomatic, discrete monomorphic flat-topped papules in the age group between six months to 12 years and perivascular mononuclear infiltrate with minimal spongiosis on histopathology.[3] These features may be different in Hepatitis-B negative individuals (pruritic pink pinhead sized vesicle-like papules.[5]

The asymptomatic individual deep red papules of GCS in HBV infection are 5-10 mm in diameter develop first on the thighs and buttocks, then on the extensor aspects of the arms and finally on the face over a period of three or four days.[4-6] The eruption fades in two to eight weeks with mild desquamation. Recurrence is unlikely, but has been reported.[7] The clinical features of the eruption caused by the other agents cannot be distinguished from HBV-associated GCS.[6] In our patient, it was the cutaneous manifestation that unmasked hepatitis B viremia as he presented with classical manifestations indicative of HBV-related GCS. Hence GCS in a patient with sickle cell disease is of dual importance as it mandates the evaluation for hitherto unrecognized transfusion related infections that may be implicated in GCS.

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