To B19 Again: Transient Aplastic Crisis in Hemoglobin SC Disease

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Learning Objective #1: To identify and treat Parvovirus B19 transient aplastic crisis (TAC) in patients with Hemoglobin SC disease (HbSC)

Case: A 22-year-old African American woman with HbSC presented with one day of severe lower back and bilateral leg pain, not relieved by ibuprofen. Review of systems was otherwise negative. Initial exam revealed mucosal pallor, scleral icterus, absence of hepatosplenomegaly and diffuse tenderness across the lower back with no focal neurologic deficits. The pain improved with the use of hydromorphone and ketorolac. However, by day 3 of admission, she complained of generalized weakness and dyspnea on exertion. She denied cough, fever, runny nose or sore throat. Labs at that time showed hemoglobin (Hgb) 6.1g/dl (a decline from 9.4g/dl on admission), WBC 7.23 x 109/L, Reticulocyte index was 1.6%, Haptoglobin <10 mg/dL, Total bilirubin 1.6 mg/dL and LDH 1049 U/L. No schistocytes were seen on peripheral smear. Parvovirus IgM and IgG antibody titers were elevated. A diagnosis of Parvovirus B19 transient aplastic crisis (TAC) was made, based on decline in Hgb, reticulocytopenia and elevated Parvovirus B19 IgM titers. 2 units of RBCs were given with appropriate increment in Hgb which remained stable over 2 days before she was discharged.

Impact/Discussion: Patients with sickle cell anemia (HbSS) are particularly susceptible to Parvovirus B19 infection due to asplenia and can lead to TAC due to the cessation of red blood cell production. Parvovirus B19 accounts for most TAC particularly among children with HbSS disease. In contrast, patients with HbSC disease often have less frequent crises which may delay presentation of Parvovirus B19 TAC to adulthood as in our patient. TAC should be suspected when there is an acute fall in Hgb that is not accompanied by an appreciate bone marrow response in red cell production. Symptoms usually are due to acute on chronic anemia with varying severity but can be life-threatening, highlighting the need for early recognition. Other Parvovirus B19 related complications such as acute chest syndrome, acute splenic or hepatic sequestration and bone marrow necrosis can occur concomitantly with aplastic crisis. Serum IgM testing is recommended for immunocompetent patients and immunocompromised should be tested by viral DNA. Treatment is usually supportive with antipyretics or ibuprofen (if fever is present) and blood transfusions if symptomatic. Recurrence of infection is rare due to development of immunity.

Conclusion: - HbSC often has a milder clinical course compared to HbSS, which can delay asplenia and its infectious complications.
- TAC should be on the considered in HbSC patients and treated aggressively if symptomatic.

CURRENT PRIMARY CATEGORY (REQUIRED): Hematology/Oncology - V
Secondary Category - V2: (none)
Accuracy - V: I affirm
Policy Verification: SGIM Presenter Registration Policy has been read.