Clinical vignette: An unusual cause of cerebral venous thrombosis

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EPSTEIN-BARR VIRUS CAUSING BOTH THROMBOCYTOPENIA AND CEREBRAL VENOUS SINUS THROMBOSIS

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CASE PRESENTATION
An 18 year old female with no past medical history awoke with a severe generalized headache associated with nausea, vomiting and photophobia. Two weeks prior, she had nasal congestion, mild sore throat, and productive cough with yellow sputum which resolved but fatigue persisted. She was taking oral contraceptives. Physical exam was normal except for significant fatigue. Pertinent labs included a normal WBC, H/H, PT/INR and aPTT. Platelets were 20,000 platelets/mm$^3$. CT Head demonstrated right transverse sinus thrombosis, with empty delta sign. MRI revealed extensive right transverse sinus thrombosis with extension into the right internal jugular vein. Peripheral smear showed marked thrombocytopenia with occasional large and giant forms, some neutrophils with toxic granulation, and occasional reactive-appearing lymphocytes. Her LFTs were mildly elevated with AST 116, ALT 120 Unit/L. Abdominal ultrasound revealed hepatomegaly with mild echogenicity and a normal sized spleen. Viral hepatitis screen was negative. Absolute CD4 count was low (182/mm$^3$); HIV negative. Hypercoagulable workup negative. ANA negative. Anti-heterophile antibody negative. Epstein-Barr antibody to early antigen positive. Epstein-Barr viral capsid antigen IgG positive but IgM negative. Epstein-Barr antibody to nuclear antigen positive. She was diagnosed with Epstein-Barr virus infectious mononucleosis causing immune thrombocytopenia, for which she was treated with dexamethasone and intravenous immunoglobulin (IVIG). The infection plus oral contraceptives likely resulted in the cerebral venous sinus thrombosis, for which she was treated with a heparin drip and then transitioned to warfarin. Oral contraceptives were discontinued. She received Keppra for seizure prophylaxis. Case Illustrates a rare presentation of Epstein-Barr virus associated with immune thrombocytopenia.

Peripheral smear in EBV

MRI head showing filling defect after contrast (empty delta sign)

TEACHING POINTS

1. Epstein-Barr virus can cause immune thrombocytopenia, particularly in children and young adults.
2. The usual screening test for Epstein-Barr virus infectious mononucleosis, the anti-heterophile antibody, can be negative at the time of presentation with hematologic complications, so serology is necessary for the diagnosis.
3. Cerebral venous sinus thrombosis main risk factors are prothrombotic state (including oral contraceptives) and infection. Many cases have multiple risk factors present.

REFERENCES
