34 YEAR OLD WOMAN WITH THROMBOCYTOPENIA AND WEAKNESS

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Patient is a 34 year old Liberian Female with past medical history significant for Systemic Lupus Erythematosus (SLE) that was recently admitted to the hospital for weakness and was diagnosed with encephalomyelitis due to SLE. At that time the patient was also noted to have low platelets felt secondary to Immune Thrombocytopenic Purpura (ITP). She has history of thrombocytopenia with her four pregnancies. During this past admission the patient was treated with hydroxychloroquine and high dose steroids for the CNS involvement of SLE but no platelet response was noted with administration of steroids. She was discharged to a rehabilitation facility, but returned to the hospital with a new episode of worsening weakness.

On admission the patient was again noted to be thrombocytopenic and anemic. She denied epistaxis, easy bruising, rash, fatigue, fevers, chills, shortness of breath, or symptoms related to upper respiratory infection. Her only complaint on presentation was weakness in the upper and lower extremities that worsened over the last week.

Medications she was taking included prednisone, hydroxychloroquine, alendronate, calcium carbonate and pantoprazole. She denied any drug allergies. Past medical history was significant for SLE complicated with encephalomyelitis and questionable history of ITP with exacerbations during pregnancy. Past psychiatric history included possible conversion disorder. No previous surgeries were noted. She denied any alcohol, tobacco, non-prescription medications, herbal substances or illegal substance use. Patient had previously worked with her husband in their restaurant, but was currently on disability. She is married with 4 children.

Physical exam revealed she was afebrile with a blood pressure 120/66 mmHg, heart rate 79 beats per minute, respirations 12 breaths per minute, and 98% oxygen saturation on room air. She was alert awake and oriented times three, pupils were equal, round and reactive to light and extraocular muscle movements were intact. Sclera was anicteric and no petechiae were noted on buccal mucosa. No lymphadenopathy in cervical, axillary or inguinal regions was appreciated. Lungs were clear and heart was regular without murmurs to auscultation. Abdominal exam was benign and there was no edema in the lower extremities. On neurological exam, cranial nerves and cerebellar exam were within normal limits. The patient had decreased strength in the upper and lower extremities bilaterally, graded as 4/5 and 1/5 respectively. Deep tendon reflexes were symmetrical and graded as 2/4 and plantar reflex was down-going. On examination of the skin no rash, petechiae or purpura were noted.

Her labs and studies were remarkable for hemoglobin of 10.8 g/dL, platelets 57000/microL, MCV 70fl, RDW 21.7%, reticulocyte 2.2%, LDH 188 IU/L, haptoglobin 115 mg/dL, total bilirubin 0.5 mg/dL, creatinine 0.5 mg/dL, INR 1.21, PT 15.8 sec, PTT 27 sec. Fibrinogen 480 mg/dL, D-Dimer 6.49 mcg/ml. Peripheral smear showed decreased platelets without clumping and scarce schistocytes.

The patient’s blood smear was not consistent with TTP. ITP was felt to be the most likely cause of her thrombocytopenia and she underwent treatment initially with high dose steroids and subsequently with IVIG, both without significant response. Her bone marrow biopsy showed hyperplastic megakaryocytosis consistent with peripheral destruction. She also underwent treatment with rituximab for encephalomyelitis without platelet response. A CT Angiogram of chest was ordered during her hospitalization for an episode of shortness of breath. There was no evidence of pulmonary embolism but a large heterogeneous splenic mass was visualized (Figure 1). This mass upon further evaluation with MRI was hypervascular and the differential diagnosis included angiosarcoma and hemangiomia.

The patient underwent splenectomy and pathology revealed a large splenic hemangioma with necrosis. Platelet counts steadily rose from the day of splenectomy and remained stable during the remainder of her hospitalization.

Discussion

This patient had thrombocytopenia from splenic sequestration due to a large hemangioma. Hemangiomas are the common primary tumor of the spleen with a prevalence of 0.03-0.14%. This is a congenital lesion arising from sinusoidal epithelium resulting from proliferation of vascular channels lined by a single layer of endothelium, most often resulting in a cavernous lesion.

Hemangiomas are variable in size ranging from a few millimeters to several centimeters. The majority of these tumors are less than 4 cm, but there are reports of lesions up to 17 cm in diameter. Hemangiomas may be single or multiple as in Klippel-Trenauney-Weber Syndrome.

The majority of hemangiomas are asymptomatic and incidentally discovered. However, larger lesions may enlarge the spleen leading to fullness and left upper quadrant discomfort, spontaneous splenic rupture or Kasabach-Merritt phenomenon (thrombocytopenia and/or coagulopathy, now called disseminated intravascular coagulation or DIC, that results from platelet trapping within a vascular tumor). Thrombocytopenia results from shortened platelet survival caused by sequestration of platelets in the vascular malformation. Episodes of acute DIC have been reported in pregnant women with congenital hemangioma and in one woman during two successive pregnancies. The hormonal
altered and increase in blood volume in pregnancy may affect pre-existing lesions, triggering episodes of acute DIC.

This patient presented with an uncommon cause of thrombocytopenia in the general population. Our approach to the patient with decreased platelets should first be focused on ruling out potential fatal causes such as TTP with the peripheral smear and pseudothrombocytopenia or clumping. In the differential diagnosis of low platelet counts one should also include medications, alcohol abuse, viral infection, hypoplasia, disseminated intravascular coagulation, myelodysplastic syndrome. ITP should be considered as a diagnosis of exclusion. Common medications associated with thrombocytopenia include heparin, glycoprotein IIb/IIIa inhibitors, diuretics, trimethoprim-sulfamethoxazole and quinine.

References