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Cardiac arrest in evan's syndrome (ES): Did coronary vasculitis cause the catastrophy?

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Introduction: ES is a rare syndrome characterized by Coombs-positive warm autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) and/or autoimmune neutropenia. We report a rare case of sudden cardiac arrest and STEMI in a young woman with ES.

Clinical vignette: 32-year-old Guyanese woman, known case of ES on prednisone, cyclosporine, eltrombopag, and on bactrim and valacyclovir prophylaxis, status post splenectomy presented with cardiac arrest, following epigastric discomfort. ROSC was achieved after 45 minutes. Hypothermia protocol was initiated. EKGs showed STEMI in anterior leads. Hemoglobin was 9.4g/dl, WBC and platelets (149,000/microlitre) were normal, direct coombs positive, and troponin at presentation was 1.69 and peaked at 94. CT chest suggested ARDS/bilateral pneumonia. Patient developed thrombocytopenia and required platelet transfusions. Patient became ventilator dependent and remained in coma, secondary to anoxic encephalopathy. EKG at different time points revealed NSR, accelerated junctional tachycardia, bidirectional ventricular tachycardia, all associated with STEMI. On day eight, patient was found to be pulseless, cardiac resuscitation was unsuccessful and patient was pronounced dead.

Discussion: ES was first described in 1951. ES, characterized by Coombs-positive warm autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) and/or autoimmune neutropenia is considered idiopathic. It may be associated with SLE, lymphoproliferative disorders, or primary immunodeficiencies. ES is associated with immune dysregulation though specific defects have not been identified. Steroids are the first line of treatment followed by immunosuppressive medications like cyclophosphamide, azathioprine, cyclosporine, mycophenolate mofetil, and rituximab. Splenectomy is the last resort in cases that are refractory to medical management. Response to rituximab is an indirect indication of B cell dysregulation. Cardiac arrest following STEMI in our patient may be attributed to possible coronary thrombosis or coronary vasculitis. Coronary vasculitis, occurring either independently or associated with autoimmune diseases, is characterized by inflammation of the blood vessel wall and fibrinoid necrosis, occlusion, stenosis, or aneurysmal dilatations. Cardiovascular diseases are one of the major leading causes of death in autoimmune diseases. These patients develop accelerated atherosclerosis of the coronary arteries secondary to the chronic inflammatory process and endothelial cell dysfunction. We emphasize the need for guidelines to screen and manage coronary vasculitis in patients with autoimmune diseases to prevent possible acute coronary events and catastrophic sudden cardiac deaths.

Biography

Purva Sunil Chhibar, MD graduated from medical school in India at the age of 24 years. She served as a Visiting Physician and Research Assistant at the University of Southern California, Los Angeles, for three years before she started her residency training in Internal Medicine at Brookdale University Hospital Medical Center, NY. She passed her USMLE exams in the 99th percentile. At her residency program she took the initiative to organize the First Annual Research Day. She won the first prize at the Oral Presentation Competition at the 8th Annual Symposium for Gender Differences in Cardiovascular Diseases. She will be chief resident starting July, 1, 2015 at Brookdale Hospital.

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