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Catastrophic antiphospholipid syndrome and heparin induced thrombocytopenia: Related diseases or chance association?

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Introduction: Catastrophic Antiphospholipid syndrome (CAPS) and heparin-induced thrombocytopenia (HIT) are immune-mediated thrombotic conditions caused by antibodies targeted to a protein-antigen complex. CAPS and HIT share similarities inclinical presentation and can have dangerous thrombotic outcomes if untreated. We describe two cases of CAPS coexisting with HIT, each with near fatal outcomes.

Case Report: A 37-year-old Caucasian male presented with non healing ulcers, complicated by cellulitis of both lower extremities for one and a half months. He developed respiratory distress during hospitalization and was diagnosed with pulmonary embolism. The patient was emergently intubated and heparin was started. Further workup showed positive anti-cardiolipinIgG at 50 GPL U/ml(RR<10 GPL U/ml) and anti cardiolipinIgM at 28 MPL U/ml(RR<10 MPL U/ml). He also developed acute hepatic injury and acute kidney injury (AKI).His multi organ failure was felt to be secondary to thrombotic angiopathy from CAPS.Treatment with intravenous immunoglobulin therapy and corticosteroids did not improve his multi organ failure and was subsequently treated with three cycles of plasmapheresis. Five days after starting heparin therapy, he developed profound thrombocytopenia (20×109/L). Heparin associated platelet antibodiesdone by ELISA were negative. However, due to strong possibility of heparin induced thrombocytopenia, heparin was stopped and was started on argatroban. His platelet count improved to baseline in 10 days after substituting heparin. He also received renal support with continuous venovenoushemodiafiltration (CVVHDF) for acute kidney failure. He recovered completely and was discharged home on anticoagulation with warfarin.A 42 yr old female presented with shortness of breath for 2 weeks. She failed outpatient oral antibiotics for left lower lobe pneumonia. CT angiogramof chest showed pulmonary embolus in the right main pulmonary artery. She was started on a heparin infusion. Overnight, she developed encephalopathy and had cold mottling of both lower extremities with abdominal pain while on heparin. Abdominal exam revealed diffuse tenderness with hypoactive bowel sounds. Right dorsalispedis pulse was not detectable by Doppler. Right femoral embolectomy was emergently performed. CT angiogram of abdomen showed thrombus in mid superior mesenteric artery. Multiple infarctions were seen, including the right lobe of the liver, spleen and both kidneys. Small bowel resection with ileocolic anastomosis was done. Platelet count dropped from 307×109/L to 115×109/L over 7 days while on heparin drip. HIT antibody by ELISA was positive on the third test. Heparin was stopped and Arixtra was started. Thrombocytopenia resolved. Due to the presence of multi-organ infarctions, CAPS was clinically suspected. Anti-Cardiolipin and beta-2 glycoproteins antibodies were negative.Lupus anticoagulant was positive. ANA titres were 1:160, speckled pattern. Double-stranded DNA antibodies were normal. Due to high suspicion of CAPS, patient was treated with plasmapharesis for five cycles. Patient symptomatically improved in terms of mental status and physical well-being and discharged home.

Discussion: CAPS and HIT share many clinical features in terms of causing thromboembolic complications with similar pathogenesis of platelet activation, damage to vascular endothelium and widespread coagulopathy.Because thrombocytopenia is one of the possible clinical features of APS it can be difficult for clinicians to distinguish it from HIT in the midst of an acute thrombotic state. The above two patients wereinteresting because CAPS and HIT were diagnosed one after another on the same admission. Therefore, it is possible that CAPS and HIT may be interrelated immune-mediated disease states.

Biography

Nay Thi Tun is currently in Residency Training of Internal Medicine Program in Easton Hospital, PA. She has obtained M.B.B.S in Yangon, Myanmar. She has done her residency training in Central Women's Hospital in Yangon, Myanmar for Obstetrics and Gynecology. At present she is a committee member of "Blood Blank" in Easton Hospital and "Instiutional Review Board" in Easton Hospital as well as she is an acting as a student coordinator. She has participated in many medical meetings and publishes number of research works.

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