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Thick as Thieves: A curious Case of Sjögren's Syndrome

Objective(s):

Method(s): Mrs H is a 38 year old woman originally from Pakistan. She was diagnosed there with Sjogren's/SLE but had clinical manifestations of Sjogren's only (Sicca). She is also known for Hashimoto's thyroiditis, celiac disease and chronic iron deficiency anemia NYD. She had delivered fraternal twins (one boy, one girl) in November of 2017 and unfortunately, her daughter developed complete heart block in utero and needed a pacemaker insertion at birth. This was secondary to SSA/SSB seropositivity in the mother. Her baby was sadly readmitted to hospital late in February in cardiogenic shock with new valvulopathy that is likely related to neonatal lupus, although exact diagnosis remains unclear. She had a previous uncomplicated pregnancy in April of 2014. She had no other significant health problems. Her current medication included hydroxychloroquine only (she had received dexamethasone during pregnancy).

Result(s): She presented to the emergency department on January 27th 2018 complaining of light headedness with falls at home. This was thought to be secondary to a worsened anemia with hemoglobin of 70 and was referred to hematology and back to her treating rheumatologist. It had been noted in the nursing notes that her blood seemed thicker and difficult to draw. In the subsequent week, she noticed decreased visual acuity, more notably in the right eye as compared to the left. She was seen by ophthalmology and was diagnosed with bilateral retinal vein occlusions. Blood work revealed hyperviscous blood, very elevated levels of IgM as well as extremely elevated levels of RF. She was diagnosed with hyperviscosity syndrome likely secondary to hyperIgM/RF. She also happened to have a positive cryoglubulin (type II). She was treated with plasmapheresis and high dose corticosteroids, which lowered her IgM level. She was put on rituximab for maintenance. Unfortunately, her vision loss remains unchanged.

Conclusion(s): This is a rare case of hyperviscosity syndrome causing retinal vein occlusion. This is also a rare case of Sjogren's hyperviscosity syndrome due to high titers of rheumatoid factor. This reflects the already known heterogeneity of clinical presentations in connective tissue diseases and the importance of close follow up and monitoring in patients who appear well but can develop life threatening illness quickly and sometimes without warning.