Unique Case of Cryofibrinogenemia for Elective Hernia Repair

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Introduction: Cryofibrinogenemia is a disorder first described by Korst and Kratochvil. In patients with this disorder, cryoproteins precipitate under cold conditions in the plasma unlike in cryoglobulinemia, where cryoproteins precipitate in the serum(1). The temperature in which this protein precipitates is 40°C(2). When this occurs, the most common manifestations are cutaneous thrombotic lesions, though more serious lesions can occur at central loci leading to significant CVA. This disorder can be primary in nature or secondary to underlying disorders such as cancers, infections, renal failure, or vasculitis(2). In one series, all patients with biopsy proven cryofibrinogenemia had an occlusive thrombotic diathesis regardless of location of the cutaneous manifestation. Prevention is best accomplished by avoiding cold environments, as a temporal association has been shown between cold exposure and the onset of symptoms and treatment of acute exacerbations has been shown to be most effective with Steroids, Streptokinase and Stanozolol(3).

Case Discussion: The patient is a 40 y/o female with PMH of transient ischemic attack, CVA, superficial venous thromboembolism, vaginal cancer, iron deficiency anemia, atopic dermatitis, and recurrent episodes of intussusception. She was diagnosed with cryofibrinogenemia in 2012. Surgically, she had a prior umbilical hernia repaired with mesh as well as a C-section. She presented to our institution with worsening discomfort in her supraumbilical region and an abdominal CT scan revealed a mass in this area. She opted to undergo elective repair of the defect.

Given her history of cryofibrinogenemia, special anesthetic precautions were taken. Pre-operatively, she was continued on her full dose aspirin and was given a 5000-unit of heparin subcutaneously. Standard ASA monitors were placed; she was pre-oxygenated and induced with Propofol, Fentanyl, Lidocaine and succinylcholine. After easy bag mask ventilation and atraumatic intubation, she was maintained with Desflurane, Fentanyl, and Rocuronium. Given her history, extra precautions to maintain her body temp were performed including both upper and lower body forced air-warming blankets, HME filter and fluid warmers. Throughout the procedure her body temp was carefully monitored by esophageal temp probe and was maintained between 98.1 and 98.40 F. After the procedure, she was extubated, taken to the ambulatory PACU and discharged home later the same day. Post-op, she was anti-coagulated with subcutaneous Enoxaparin 40 mg daily for 10 days.

Conclusion: In a surgical environment prone to hypothermia, we were able to safely provide anesthesia to a 40 y/o F with cryofibrinogenemia without thrombotic complications. This was accomplished through convective warming, fluid warming, and preventing evaporative heat loss. A short course of anticoagulation with enoxaparin further prevented thrombosis in the immediate post-op course.
References:

2) Belizra CC et al. Rheum; 2008 Feb; 47(2): 205-7