XIX CONVEGNO TRIENNALE
VERONA
15, 16, 17 e 18 OTTOBRE 2026
Camera di Commercio di Verona
AICE Associazione Italiana Centri Emofilia

3.04 Acquired coagulation disorders

ACQUIRED FACTOR XIII DEFICIENCY: AN UNDERDIAGNOSED CAUSE OF DELAYED BLEEDING AFTER PERICARDIOCENTESIS

M. Luongo[1], J. Di Paolo[1], G. D'Amico[2], P. Massaro[1], M. Di Mascio[1], R. Malizia[1], U. Benedetto[2], S. Verna[1]

[1]Immunohematology Service, Asl 02 Abruzzo, Chieti, Italy; [2] Department of Cardiac Surgery, Asl 02 Abruzzo, Chieti, Italy

Background. Acquired factor XIII (FXIII) deficiency is a rare and underdiagnosed cause of bleeding, characterized by impaired fibrin clot stabilization despite normal results on routine coagulation tests (PT, aPTT, INR). Bleeding is typically delayed and may be difficult to interpret. Pathogenic mechanisms include peri-procedural or post-traumatic consumption, reduced synthesis, and the presence of autoantibodies, particularly in autoimmune diseases such as systemic lupus erythematosus (SLE).

Case Report. We report the case of a patient with SLE, not receiving active treatment, who underwent pericardiocentesis for cardiac tamponade secondary to pericardial effusion. The clinical course was complicated by recurrent hemopericardium despite normal coagulation parameters. Early recurrence of cardiac tamponade required re-intervention, followed by a further episode four days later requiring a third procedure. Given the normal PT and aPTT values and the suspicion of a functional impairment in the final phase of coagulation, FXIII activity was assessed, along with a mixing study and serial monitoring. FXIII activity measured on postoperative day 8 was reduced (24%). The mixing study showed partial correction (55%), suggesting the absence of a clinically significant inhibitor. Serial measurements demonstrated spontaneous recovery of FXIII levels, supporting a peri-procedural consumption mechanism. However, the incomplete correction observed in the mixing study, together with the concomitant administration of high-dose corticosteroids for acute respiratory failure, does not fully exclude a transient immune-mediated component.

Conclusions. FXIII deficiency should be suspected in unexplained, delayed, or recurrent postoperative bleeding with normal routine coagulation tests, particularly in patients with autoimmune diseases. In the absence of clinical suspicion, this condition may remain undiagnosed. In surgical settings, FXIII levels typically decline between postoperative days 4 and 7 and remain reduced longer than other coagulation factors. Although no universally accepted threshold exists, levels below 30–50% are generally considered clinically relevant. Management should be guided by clinical presentation and FXIII activity. In this case, serial monitoring supported a conservative approach, avoiding unnecessary replacement therapy, although earlier recognition might have reduced the need for repeated interventions. The incorporation of FXIII assessment into perioperative protocols may improve management and reduce unnecessary invasive procedures.

Abstracts printing tool powered by pagepress.org