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Case Report
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A Successful Surgical Intervention in Sepsis Associated Acquired Lupus Anticoagulant with Prolonged Coagulation Screening: Against All Odds

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Abstract

Deranged coagulation profile is commonly seen in critically ill patients due to severe sepsis, Disseminated Intravascular Coagulopathy (DIVC), and drug-induced. Lupus Anticoagulant (LA) is another possible differential diagnosis to be considered, not uncommonly reported in critically ill patients. However, LA with a prolonged Prothrombin Time (PT) is a rare condition.

Keywords: Lupus anticoagulant; Prolonged prothrombin time; Coagulation profile; Mixing test; Sepsis

Introduction

Coagulation profile is commonly sent as a part of initial investigations for bleeding tendency, pre-operative screening, as well as for patients with severe sepsis. The routine coagulation profile consists of Activated Partial Thromboplastin Time (APTT), Prothrombin Time (PT), and International Normalized Ratio (INR).

Deranged coagulation profile is commonly seen in critically ill patients due to severe sepsis, Disseminated Intravascular Coagulopathy (DIVC), and drug-induced. It complicates the management of critically ill patients requiring surgical intervention. Transfusion of Fresh Frozen Plasma (FFP) and other blood products such as cryoprecipitate and platelet concentrate is indicated prior operation to prevent severe bleeding intra- and post-operatively.

Uncorrected coagulation profile despite transfusion of blood products should raise the suspicion of alternative diagnoses such as Lupus Anticoagulant (LA), which is not uncommonly reported in critically ill patients. It is usually characterized by a persistent prolonged APTT despite correction. However, strong LA can present with a prolonged APTT, PT and INR.

Case Presentation

A 54-year-old man was admitted for sepsis secondary to infected right Below-Knee-Amputation stump (BKA). He had a background history of ischemic heart disease and diabetes mellitus with End Stage Renal Disease (ESRD) on regular hemodialysis and history of right BKA for diabetic foot ulcer.

As he was in severe sepsis, he required urgent Above-Knee-Amputation (AKA). However, his preoperative coagulation profile was deranged with prolonged Activated Partial Thromboplastin Time (APTT) 131.9 sec, PT 35.3 sec, and International Normalized Ratio (INR) 3.70. Full blood counts showed hemoglobin 9.2 g/dL, white cell count 21×10^9/L, and platelet count 502×10^9/L. He was not on heparin or any anticoagulant.

Initial impression was coagulopathy secondary to sepsis. Despite transfusion with Fresh Frozen Plasma (FFP), his coagulation profile worsened with the highest APTT>180 sec and INR 10 without any bleeding event. APTT and PT were not corrected by immediate and 2 h incubation mixing tests, with factor VIII level of 600% and hyperfibrinogenemia. These findings were suggestive of strong LA.

Intravenous methylprednisolone was administered, and plasmapheresis was performed twice

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Table 1: Serial coagulation profile monitoring.

Day of Admission	Coagulation Profile			Remarks
	APTT (second)	PT (second)	INR	
Day 1	52.9	16.1	1.31	
Day 3	131.9	35.3	3.7	
Day 4	112.6	43.5	4.87	Transfused 2 units FFP
Day 5	135.6	46.3	5.29	Transfused 4 units FFP
Day 6	133.5	47.8	5.52	Transfused 2 units FFP
Day 7	155.7	50.4	5.92	
Day 8	169	48.9	5.69	
Day 10	137.3	49.1	5.72	
Day 13	176.8	51.5	6.1	
Day 14	>180	62.1	7.8	Transfused 2 units FFP
Day 15	>180	65.5	8.37	Transfused 2 units FFP
Day 16	>180	49.5	5.78	
Day 17	146.4	46.4	5.31	Started on IV Methylprednisolone 50mg OD
Day 18	>180	69.5	9.05	
Day 19	116.4	45.2	5.12	Plasmapheresis x1
Day 20	98.4	51.1	6.03	Plasmapheresis x2
Day 21	>180	53.5	6.4	
Day 22	>180	75.4	10.08	Went in for operation AKA
Day 23	75.1	37.4	4	Day 1 post operation

APTT: Activated Partial Thromboplastin Time; PT: Prothrombin Time; INR: International Normalized Ratio; FFP: Fresh Frozen Plasma; AKA: Above Knee Amputation

with transient improvement in the coagulation profile. However, the levels became deranged again with APTT>180 sec, PT 75.4 sec, and INR 10.08.

With the impression of acquired LA, AKA was performed with preoperative tranexamic acid, desmopressin, and platelet transfusion for platelet dysfunction in ESRD. The surgery was successfully carried out with an estimated blood loss of 400 ml. Day 1 post-operative, his APTT was 75.1 sec, PT 37.4 sec, and INR 4.00.

LA confirmatory test revealed a positive Staclot LA and a prolonged Dilute Russell's viper venom time of 181.2 sec.

Discussion

A deranged coagulation profile can be due to factor deficiency, presence of specific factor inhibitor or LA. Identification of the underlying cause is important as some disorders increase the risk of bleeding while the others are prothrombotic. The initial approach to a prolonged APTT or PT is to rule out an artefactual abnormality due to a mixture of heparin solution with blood, use of anticoagulant, or due to liver disease [1]. An additional test that can be done is a mixing test to differentiate between a coagulation factor deficiency and the presence of an inhibitor.

LA is an inhibiting antibody which causes prolonged APTT in vitro despite its prothrombotic activity. In sepsis, DIVC is the foremost important diagnosis of coagulopathy. However, persistently deranged coagulation profiles in a non-bleeding patient, which are not corrected with FFP transfusion or vitamin K should raise the suspicion of LA. Although rare, acquired LA is one of the possible differential diagnoses and has been reported in literature to be frequently developed in critically ill patients [2].

Lupus Anticoagulant-Hypoprothrombinemia Syndrome (LAHS) should be considered in LA with a prolonged PT and INR. An acquired factor II deficiency in association with the acquired lupus anticoagulant complicates the management further as it may predispose the patient to both thrombosis and severe bleeding [3]. PT is usually relatively insensitive to LA; however, a strong LA might give rise to a falsely increased PT [4]. A factor II level and an uncorrected PT mixing test can help to differentiate a strong lupus anticoagulant from LAHS.

Managing a sick patient with the presence of LA and deranged coagulation profile who needs a surgical intervention may pose a big dilemma for the surgeons and physicians. In the present case, surgical intervention is the definitive management in acquired LA associated with severe sepsis from the infected stump. Improvement of the coagulation profile post-operatively indicated the infected stump was the most likely source of LA formation. Therefore, it is imperative to diagnose acquired LA as they differ in the coagulability state especially in the decision for surgical intervention and in critically ill patients whom the risk of thromboembolic event is high.

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