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Case Report

Incidental diagnosis of antiphospholipid antibody syndrome (APS) in a trauma patient with thrombocytopenia & its anesthetic management

Neha Magoon *, Barkha Bindu, Harsh Sapra

Dept. of Neuroanaesthesia & Critical Care, Medanta, The Medicity Gurugram, India

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Introduction

Coagulopathy is frequently observed in acute phase of trauma. It is associated with greater mortality as well as transfusion requirements [1]. Coagulopathy with simultaneous thrombotic manifestations is a rare clinical presentation. On the other hand, Antiphospholipid antibody syndrome, an autoimmune disorder can present with both thrombosis and thrombocytopenia. The management of both the conditions is significantly different. Hence, it is important to differentiate between the two conditions. We encountered one such case of trauma with coagulopathy where diagnosis and management was a dilemma, until a bedside transthoracic echocardiography provided some essential hints to the underlying pathology.

Case report

A 30 year old, young hypertensive male, presented to our Emergency Department 17 h after a road traffic accident (RTA). His injuries included multiple bilateral rib fractures, fracture of midshaft of right clavicle, bilateral hemothorax and D12 burst compression fracture (Fig. 1). Neurologically, his Glasgow Coma Score (GCS) was E4V5M6 with immediate onset paraplegia after RTA. In view of hemothorax, an intercostal drain (ICD) was inserted in left side of chest (Fig. 2). After initial management, the patient was shifted to our ICU for monitoring and further workup for spine surgery. Baseline investigations revealed an isolated raised APTT (53.7 s) with normal PT (12.7 s) and INR (1.23), hemoglobin (Hb) % of 13.4 g/dl and a platelet count of 110×10^9/l. A total of 1750 ml blood drained from the ICD within 10 h of insertion. His Hb dropped to 7.4 g/dl and platelet count was 90×10^9/l. He was transfused two units packed red blood cells (PRBC). Bedside transthoracic echocardiography (TTE) performed as part of preoperative evaluation revealed an echogenic mass attached to atrial aspect of A2 scallop with normal ejection fraction (EF), likely nonbacterial thrombotic

* Corresponding author.
E-mail address: neha_magoon@yahoo.com (N. Magoon).

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endocarditis (NBTE). A transesophageal echocardiography (TEE) could not be performed due to coagulopathy. In the subsequent days, his platelet counts continued to decline and dropped to 40×10^9/l over next 2 days. The simultaneous presence of thrombocytopenia, NBTE and deranged APTT prompted an evaluation for Antiphospholipid Syndrome (APS). His IgM and IgG Anti Cardiolipin antibodies were positive and Lupus anticoagulant test was strongly positive (Normalised LA ratio 2.9). Hence, a diagnosis of primary Antiphospholipid Syndrome was made. Accordingly, patient was started on Hydroxychloroquine and steroids. In view of upcoming spine surgery, anticoagulant was not started preoperatively. Preoperative lower limb venous Doppler did not show any evidence of deep vein thrombosis (DVT). Patient was subsequently taken up for surgery on a platelet count of 53×10^9/l and APTT of 55.1 s, with high risk consent in view of coagulopathy and high likelihood of perioperative thrombotic events. An uneventful surgery (D12 bilateral laminectomy and decompression with D10-D11-L1-L2 percutaneous pedicle screw and rod fixation) (Fig. 3) in prone position was performed with intraoperative transfusion of 2 units single donor platelet concentrate (SDPC). Intraoperatively, DVT prophylaxis with sequential compression devices, prevention against hypothermia, strict input/output charting & adequate hydration was maintained. Invasive blood pressure monitoring and optimal analgesia were continued in postoperative period. Patient received his first dose of anticoagulant 24 h after surgery. His platelet counts increased gradually and reached normal limits. He was later shifted to ward and subsequently discharged. He had a Barthel Index Score for Activities of Daily Living of 20 at discharge. At 1 month follow up, he was

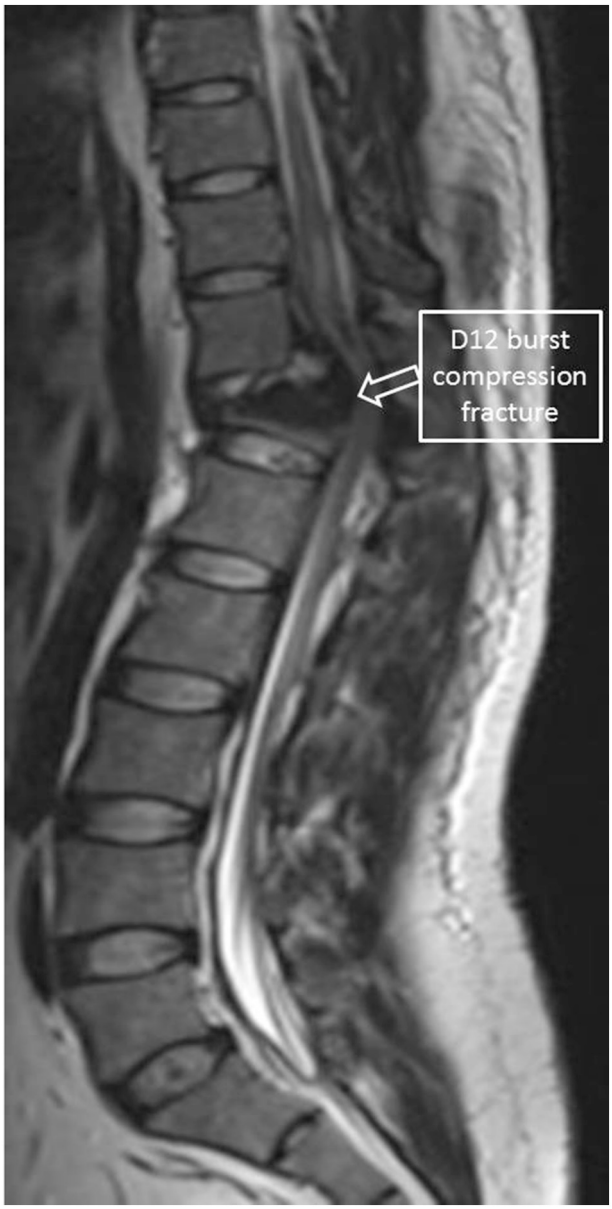

Fig. 1. Preoperative MRI of the spine showing D12 burst compression fracture.

being managed as a case of non-criteria APS by Rheumatology division. He did not report for subsequently advised follow up visits, hence, long-term follow up was not done.

Discussion

We have discussed in this report, a patient of trauma who came to us with signs of trauma-induced coagulopathy, but was diagnosed with an unusual and somewhat contrary condition. We have tried to bring out two notable points based on our experience. First is the fact that not all thrombocytopenia and coagulopathy in trauma are due to DIC, and second is the significance of TTE in preoperative workup.

Bleeding and coagulopathy are common presentations of patients with trauma. This could be either due to the primary injury itself or due to trauma induced coagulopathy. Management of bleeding and coagulopathy involves multiple transfusions to maintain hemoglobin percentage and platelet counts within acceptable limits. Surgery, if required in such patients, involves high risk. A timely and rapid diagnosis of APS in our patient led to critical clinical decisions and helped us to manage the patient better. A timely diagnosis of APS also helped us prevent adverse postoperative thrombotic complications.

Antiphospholipid syndrome is a multisystem autoimmune condition, characterized by clotting in arteries & veins. The revised Sapporo criteria defines it by the clinical evidence of arterial/venous/small vessel thrombosis in any tissue/organ &/or pregnancy morbidity, along with persistently positive Antiphospholipid antibodies (aPL) measured with lupus anticoagulant (LA) test, Anticardiolipin antibody (aCL) enzyme linked immunosorbent assay (ELISA), and/or anti-β2-glycoprotein-1 antibody test, on 2 occasions at least 12 weeks apart [2,3]. when APS occurs without any underlying autoimmune disease, it is called primary APS. APS also has certain "non-criteria" clinical features, thrombocytopenia being one of the most important among them. Infections (borrelia, treponema, HIV, HCV), drugs (quinidine, phenytoin, valproic acid), environmental factors (trauma, low serum vitamin D) are all considered triggers for APS [4].

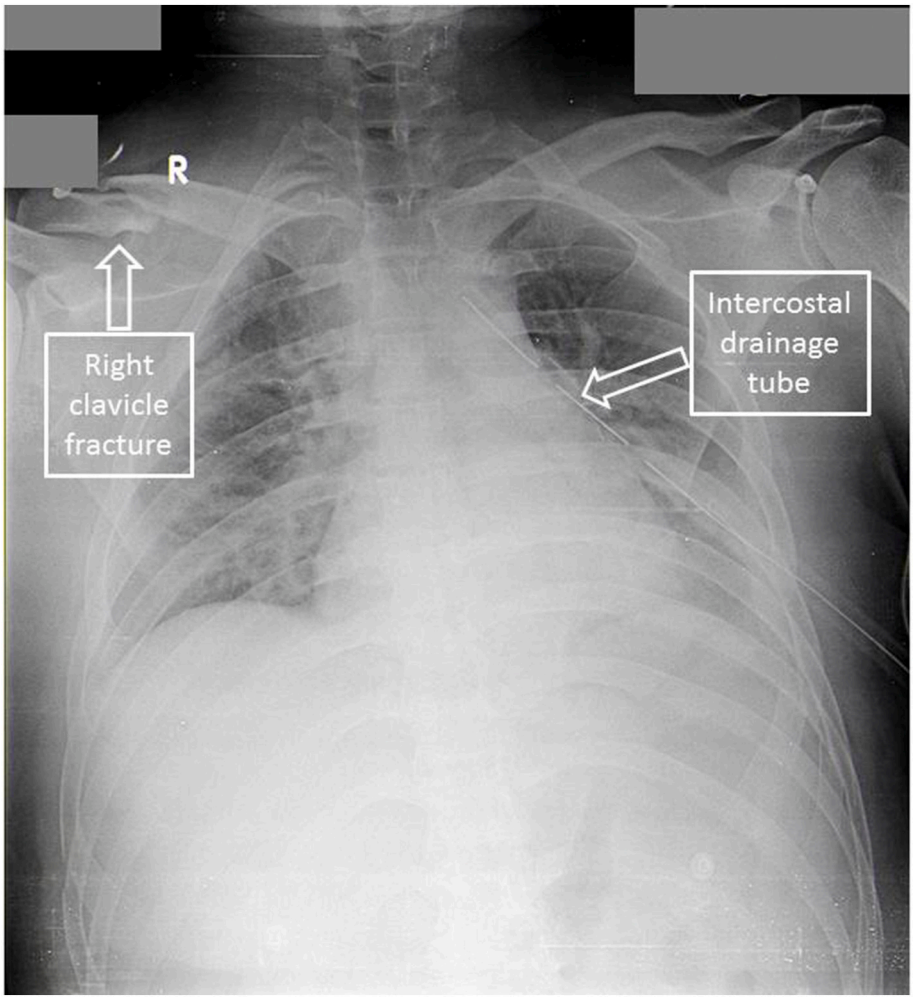

Fig. 2. Preoperative chest radiograph showing left sided intercostal drain and fracture of midshaft of right clavicle.

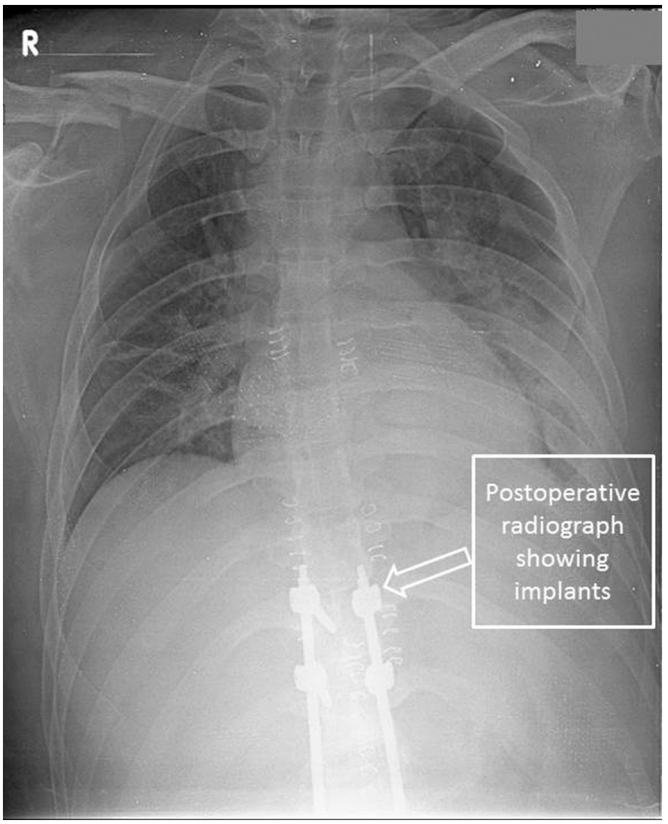

Fig. 3. Postoperative radiograph showing surgical implants.

Literature search shows reports of occurrence of catastrophic APS that is precipitated by trauma in patients with APS [5,6]. However, a diagnosis of primary APS in the setting of trauma is relatively rare.

Thrombocytopenia in APS is reported in 30–46% of patients [7], which is generally mild and requires no intervention. Severe thrombocytopenia ($<50 \times 10^9/l$) is seen in <10% of cases [8]. Binding of antiphospholipid antibodies to platelets promotes platelet activation and aggregation causing thrombus formation [9,10]. Thrombocytopenia in APS is therefore associated with thrombotic, and sometimes hemorrhagic complications. Missing the diagnosis of APS in our patient could therefore have been catastrophic. It would have led to multiple transfusions preoperatively in order to correct coagulopathy, and avoidance of anticoagulants postoperatively due to thrombocytopenia. Either of these interventions could have had adverse sequelae. Thrombotic complications were the most common cause of death in APS with a mortality rate of 5.3% in a large European cohort during a 5 year study period [11].

The presence of NBTE on echocardiography prompted us to look for alternative causes of thrombocytopenia. Had the TTE not been performed, the patient would most likely have been diagnosed as a case of trauma related DIC and administered multiple transfusions. A TTE is not routinely performed as part of preoperative evaluation at several centres. Based on this patient experience, we feel that 2D echocardiography is a convenient and feasible bedside modality that is easy to perform and must be incorporated into practice, especially for patients with clinical presentations that are not entirely explainable.

Intraoperative management of such patients must aim at preventing hypothermia and dehydration [12]. Hypothermia can directly damage enzymes in coagulation cascade & hamper platelet function. Dehydration & hypotension increase blood viscosity. Therefore, use of body and fluid warmers with adequate hydration is imperative. Transfusion threshold in such patients must be kept high as well. Our patient was transfused platelets intraoperatively in view of spine surgery.

Conclusion

Thrombocytopenia with thrombosis in a case of trauma is an unusual presentation. We must consider APS in the differential diagnosis of such patients. Transfusion threshold must be kept high and reserved for overt thrombocytopenia. Incorporating TTE in preoperative evaluation of such patients is not only feasible, but can prevent catastrophic management errors.

Fundings

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Declaration of competing interest

None.

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