CASE REPORT

Anesthetic management of the parturient with combined protein C and S deficiency

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ABSTRACT

Protein S is a vitamin K de pendent co-factor of protein C. Deficiency of Protein C and S results in a hypercoagulable state, which is treated with anticoagulation. This is the first report of a patient with combined protein C and S deficiency, who underwent a Cesarean section under spinal anesthesia in KRL Hospital Islamabad (Pakistan).

Key words: Protein C and S deficiency; spinal anesthesia; caesarean section

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INTRODUCTION

Protein C is a 62-KD, Vitamin K dependent glycoprotein synthesized in the liver, and circulates in the blood as an inactive zymogen at a concentration of 4μg/ml. Its activation into activated protein C (aPC), is catalyzed b y thrombin when it is bound to the endothelial proteoglycan thrombomodulin.This activated form exerts its anticoagulant activity primarily through inactivation of factors Va and VIIIa[1],which are required for factor X acti vation and thrombin generation. The catalytic activity of activated protein C is enhanced by protein S. The results of deficiency of these can result in dee p vein thrombosis, pulmonary embolism, thrombophlebitis, neonatal purpura fulminans, liver cir rhosis and w arfarin-induced skin necrosis .[2]

Protein S is also a vitamin K-de pendent plasma protein discovered in 1977. Patients, deficient in either protein S or C, or who express a dysfunctional protein S , are at a risk for repetitive thrombosis.1 Under normal circumstances, pregnancy is associated with a h ypercoagulable state.[2] Venous thromboembolism is among the leading causes of maternal death in developed countries.[3-5] We describe the use of neuraxial anesthesia for cesarean section in a parturient who presented with both protein C and S deficiency.

CASE REPORT

A 34 years old primigravida, with a past medical history of epilepsy controlled on two antiepileptic medications, had developed deep venous thrombosis of left leg at 30 weeks of gestation and was on anticoagulant therapy i.e . low molecular weight heparin according to her w eight. Her work up for hereditary thrombophilia was done and she was found to have deficiency of both protein C and S. She had developed bursitis of left knee joint and mumps at 36 weeks of gestation. Her elective C- section was decided at 37 weeks of gestation. A coagulation profile obtained prior to surgery showed her PT 14, APTT 40, INR 1.0 and platelets 205000.

Anticoagulants were stopped 24 hours prior to her delivery. Spinal anesthesia was chosen for the surgery. The patient was preloaded with inj. Ring er's lactate solution in the operating room and placed in the sitting position. A spinal anesthesia was given at the L3-L4 level using a 25 gauge pencil point needle. The patient received 2.0 cc of 0.5% (hyperbaric) bupivacaine to obtain a T4 level of anesthesia. The patient was placed in the left uterine displacement position and had an uneventful Cesarean section. She was monitored in the post anesthesia care unit for two hours. She had by this time recovered motor and sensory functions

bilaterally and was discharged from the recovery room and sent to the high dependency unit. In the immediate post op period she was given two fresh frozen plasmas and one unit of blood. Her repeat coagulation profile was carried out. The results were in normal range. She was restarted on aspirin and heparin on 2nd post operative day and was discharged on day 7 after shifting to oral warfarin as an anticoagulant for further management.

DISCUSSION

Protein S is a vitamin K dependent co-factor of protein C. Protein C acts by neutralizing activated factors V and VIII and by an inhibitory action on plasminogen activator.[6] Deficiency of both protein C and S results in increased incidence of venous thrombosis. Deficiency of these may be hereditary or acquired. Hereditary disease is an autosomal dominant disorder, with homozygotes generally dying in infancy due to massive thrombosis. Heterozygotes generally have their first thrombotic event by their mid-twenties[7]. Acquired disease is usually due to hepatic disease.[7]

The frequency of venous thromboembolism from protein C and S deficiency ranges from 7 to 17%.[6] There is also a risk of spontaneous abortion. Pregnant patients with protein C and S deficiency can be managed with either a combination of aspirin and subcutaneous heparin or low molecular weight heparins (LMWHs).[8] LMWHs do not cross the placenta[8-9] and thus have a fetal safety profile equivalent to that of unfractionated heparin. Patients should not be offered neuraxial anesthesia unless the PTT is within the normal range if they are managed with aspirin and heparin.[10] The effect of low molecular weight heparin cannot be determined by a lab test, so the patient must be off it for at least 24 hours prior to neuraxial anesthesia. Tachycardia, hypotension, and hypothermia increase the likelihood of thrombosis and should also be avoided[10]. The risk of DVT among healthy pregnant women undergoing elective cesarean section is low and general medical thromboprophylaxis is probably not justified.[11]

To the best of our knowledge, this is the first report of a patient with combined protein C and S deficiency, who underwent a Cesarean section under spinal anesthesia. Our search revealed three previous case reports of patients with only protein S deficiency, who presented in labor.[12,13] out of these, two patients delivered by caesarean section done under spinal anesthesia without complication. The third patient had been on heparin, also. She had had a combined spinal/epidural anesthetic vaginal delivery.

Spinal anesthesia has an added benefit of being associated with a lower incidence of DVT. It is also preferred as pregnant patients are at increased risk for aspiration prior to intubation for general anesthesia and there is an increased risk of difficult intubation. A regional anesthetic technique is thus preferred whenever possible, and general anesthesia should be avoided.

CONCLUSION

In summary, neuraxial techniques can be used safely in patients with protein C and S deficiency as long as an appropriate laboratory workup is done and the patient has been off anticoagulants for an adequate period of time.

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