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CASE REPORT

Rare Presentation of Catastrophic Antiphospholipid Syndrome with Myocarditis in Post-partum Period: Case Report and Review of Literature

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Introduction

Catastrophic antiphospholipid syndrome (CAPS) is a disorder characterized by multi-organ failure caused due to multiple small vessel thrombosis associated with thrombotic microangiopathy. In more than half the cases, CAPS is triggered by an identifiable factor like infection, trauma, surgery, anticoagulation withdrawal, lupus flares or pregnancy [1]. It is suggested that APS is not only a thrombotic disease but is also associated with microangiopathic features which can explain the greater incidence of HELLP (haemolysis, elevated liver enzymes and low platelet count) syndrome in pregnant women developing this dreaded complication [2].

Case Report

A 25-year-old third gravida woman with previous history of two abortions was referred to our centre at 27-week and 3-day period of gestation with complaints of shortness of breath and uncontrolled hypertension. She had a history of previous two missed abortions at 6–8-week gestation, and during the workup for recurrent pregnancy loss she was diagnosed to have APS with aCL IgG and IgM positive. She was started on aspirin in inter-conceptional period and on low molecular weight (LMW) heparin as soon as she became pregnant. She developed pre-eclampsia at 25-week gestation, and blood pressure (BP) was inadequately controlled on alpha-dopa, labetalol and nifedipine. She was also given dexamethasone for foetal pulmonary maturation. On admission, she was breathless at rest, i.e. New York Heart Association (NYHA) Class 4 with tachycardia, tachypnoea and blood pressure of 160/100 mmHg. Laboratory data at admission included haemoglobin of 11.5 gm/dl, platelet count of 1.5 lakh per mm^3, urea of 34 mg/dl and creatinine level of 0.9 mg/dl. Liver transaminases were mildly raised with SGOT being 62 IU/ml, SGPT being 79 IU/ml, and alkaline phosphatase was 139. Her fundus examination showed grade 1 hypertensive retinopathy, and 2D echocardiography showed moderate to severe mitral regurgitation with the presence of an eccentric posterolateral jet and normal biventricular function. Initial chest X ray was suggestive of pulmonary oedema. Obstetric sonography showed a single live foetus in breech presentation with an estimated foetal weight of 865 g. A provisional diagnosis of pre-eclampsia with severe features with partial HELLP syndrome was made. She was admitted in the obstetric high dependency unit for stabilization. On the third day of her hospitalization, she developed signs and symptoms of imminent eclampsia and pregnancy was terminated by emergency caesarean section with the delivery of a live born female with weight of 1020 g and APGAR scores of 7 and 9 at one and 5 min of life. She received magnesium sulphate for eclampsia prevention till 24 h after delivery. Anticoagulation with LMW heparin was restarted 24 h after delivery. On the post-partum day 2, she developed shortness of breath with NYHA class IV symptoms, and chest auscultation revealed diffuse bilateral basal chest crepitation. Chest X ray was suggestive of pulmonary oedema, and diuretics were administered. Her course progressively worsened, and she developed hypotension and cardiogenic shock and was shifted to the cardiac care unit. Echocardiography was repeated which showed severe mitral regurgitation with global left ventricular hypokinesia and ventricular ejection fraction of 20–25%. A provisional diagnosis of CAPS with myocarditis was made, and she was given intravenous immunoglobulin (IVIg) in a dosage of 1 g/kg body weight. Subsequently, repeat echocardiography showed a worsening ejection fraction of 10–15%. On the post-partum day 5, she developed cardiac arrest and expired.

Discussion

Pregnancy and the post-partum period are well-known predisposing factors for venous thrombosis due to procoagulation state. Thrombophilic disorders notably increase gestational vascular complications, leading to pre-eclampsia, foetal growth retardation, abruptio placentae and recurrent pregnancy loss. In our patient, APS was diagnosed during workup of recurrent pregnancy loss which showed aCL positivity. In most of the reported cases of CAPS, previous obstetric history was usually unsuccessful with Gomez-Puerta et al. [3] reporting only 1 patient with a previous successful pregnancy and 9 patients with previous abortions or foetal losses and Hanouna et al. [4] reporting only one successful pregnancy among their series of 13 patients.

HELLP syndrome complicates around 0.6% of all pregnancies and is thought to occur due to activation of microvascular coagulation followed by formation of microthrombi in multiple organs including the liver. In the series of 8 women by Gomez-Puerta, they found a correlation between severe HELLP syndrome and development of CAPS in the obstetric period [3]. Hanouna et al. [4] also reported that in 12 out of 13 women, CAPS was preceded by HELLP syndrome which was associated with pre-eclampsia in six cases and with eclampsia in three and occurred at a mean gestational age of 26.6 weeks (range 13–37 weeks) [4]. Both these authors reported a significant association between HELLP syndrome and CAPS (92 and 53%) [3, 4].

CAPS is an unusual (<1%), but life-threatening variant of the APS, characterized by an aggressive form of the disorder in which there is formation of small vessel thrombi at multiple locations over a short period of time causing multi-organ failure. There have been more than 280 reported cases in the international web-based registry for this condition "CAPS registry" (http://www.med.ub.es/MIMMUN/FORUM/CAPS.HTM) though there is a paucity of the literature from India. CAPS during pregnancy or post-partum period represents 6% of cases and can be triggered by numerous factors including infections such as endometritis, caesarean wound or episiotomy wound infection, mastitis and anticoagulation withdrawal during labour. In our patient, CAPS developed on the post-partum day 2 and main manifestation was cardiac involvement in the form of severe myocarditis leading to cardiac failure. In

the series by Gomez-Puerta et al. [3], cardiac involvement was seen in only 3 (20%) patients in the form of myocardial infarction, valve disease and myocardial TMA in one case each. Hanouna et al. reported CAPS occurring in postpartum or post-abortal period in 12 patients. There was a mean interval of 9.1 days between development of HELLP syndrome and CAPS. In their series, cardiac involvement was seen in 8 women with severe cardiac insufficiency in 4, one of whom required extracorporeal membrane oxygenation (ECMO) and another had a cardiac arrest and was successfully resuscitated [4].

Conclusion

Patients with APS who develop HELLP syndrome in antenatal period should be carefully followed up in the post-partum period for development of CAPS. Early resumption of anticoagulation with aspirin and LMW heparin may be warranted in these patients. Prompt initiation of steroids may also be a useful strategy in these patients to improve outcomes. Possibility of this rare diagnosis should be kept in mind while managing such patients.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interests.

Informed Consent All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5). Informed consent was obtained from all patients for being included in the study.

References

1. Cervera R. Update on the diagnosis, treatment, and prognosis of the catastrophic antiphospholipid syndrome. Curr Rheumatol Rep. 2010;12:70–6.
2. Appenzeller S, Souza FH, Wagner Silvia de Souza A, et al. HELLP syndrome and its relationship with antiphosphplipid syndrome and antiphospholipid antibodies. Semin Arthritis Rheum. 2011;41:517–23.
3. Gomez-Puerta JA, Cervera R, Espinosa G, et al. Catastrophic antiphospholipid syndrome during pregnancy and puerperium: maternal and fetal characteristics of 15 cases. Ann Rheum Dis. 2007;66:740–6.
4. Hanouna G, Morel N, Le Thi Huong D, et al. Catastrophic antiphospholipid syndrome and pregnacy: an experience of 13 cases. Rheumatol (Oxford). 2013;52(9):1635–41.