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Case Report

A rare manifestation in anti-phospholipid syndrome

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ABSTRACT

Pregnancy per se is a state of immuno suppression and hypercoagulability. When associated with auto immune disorders, we encounter life threatening situations. Antiphospholipid syndrome still represents an important treatable cause of pregnancy morbidity. Known for its systemic arterial and venous thrombotic manifestations. There exist a complexity and heterogeneity of the mechanisms beyond the poor obstetric outcome in APS. Thrombotic events in these patients consist in deep vein thrombosis (32%), stroke (13%), superficial thrombophlebitis (9%), pulmonary embolism (9%) and transient ischemic attack (7%), among other clinical manifestations. Stroke is the most frequent arterial thrombosis. Rarely, APS can manifest in the form of hemorrhage. We encountered Subdural haematoma in pregnancy which is an infrequent manifestation. Our patient had gone through all bitter experiences on account of APS mentioned in domain 4-obstetric: prefetal death, fetal death, preeclampsia with severe features, central nervous system dysfunction, placental insufficiency with severe features. She has prosthetic aortic valve and on anticoagulants. This pregnancy she reported in emergency room with subdural haematoma and miscarriage with the multidisciplinary team work we were able to treat the uncommon manifestation successfully. Embarking on pregnancy in future after a critical episode is debatable. In certain situations, the decision making is vital to minimize the morbidity.

Keywords: Recurrent pregnancy loss, Antiphospholipid syndrome, Subdural haematoma, Venous thrombosis, Bad obstetric history

INTRODUCTION

Pregnancy per se is hyper-coagulable, immuno suppressive and hyper dynamic state and considered as a stress test for women of reproductive age. When auto immune disorders are associated with pregnancy it will lead to adverse perinatal outcome. Antiphospholipid syndrome is an auto immune disorder which contributes to recurrent pregnancy loss, preeclampsia and other prothrombotic conditions in pregnancy. Manifestations are owing to vascular thrombosis both arterial and venous.[1,2] The pathophysiology is antibodies are produced against phospholipid binding proteins. We have encountered a 38 years old lady married for 20 years, well preserved fertility but repeated fetal losses. She had 2 intra uterine fetal demise in late second trimester. Uncontrolled severe pIH which warranted termination at 20 weeks' gestation and one neonatal loss on account of gross prematurity. APS was diagnosed after her second pregnancy and all necessary measures taken in subsequent pregnancies. Her first wedlock broken after 4th pregnancy. In her second marriage (1 year ago) she conceived spontaneously and reported at 6 weeks' gestation and further follow up is narrated in this article.

CASE REPORT

Mrs S, 38 years old, G 5 P4 L0 A0 admitted with H/o 2 MA with severe headache and difficulty in speech since 1-day, sudden onset, no history of vomiting, convulsions,

syncope or bleeding per vaginum. She is a known case of anti-phospholipid syndrome diagnosed after recurrent pregnancy loss and on treatment. Undergone aortic valve replacement 12 years ago at our institute for aortic stenosis and on anticoagulants. She has periodical follow up with cardiologist and the prosthetic valve is functioning well. She was taking treatment for migraine on and off. She had two intra uterine fetal demises, medical abortion at 5 MA for severe PIH and one neonatal loss (gross preterm). She has undergone 3 vaginal deliveries and one caesarean section with no live issues. Present pregnancy was confirmed at 6 weeks as intrauterine and viable. She was on folic acid, supplementation, ecosprin and oral anti-coagulant. She was switched over to LMWH with the concurrent consultation with rheumatologist and cardiologist. Laboratory values at booking.

Table 1: 10 days after booking she landed in emergency room with headache.

LUPUS Anti-coagulant -DRVVT	222.5	Cardiolipin Antibody Ig M	16.84
Patient Plasma: PNP1:1 Mix value	137.9	Cardiolipin Antibody Ig G	>150
Screen ratio	6.23	Cardiolipin Antibody Ig A	11.91
Mix rate	3.86	CBC	normal
B2 Glycoprotein Ig G	>150	S.Creatinine	normal
B2 Glycoprotein Ig M	5.44	-	-
B2 Glycoprotein Ig A	15	-	-

Table 2: Vitals.

MRI brain			
Haemoglobin	8.4		
Platelet count	33,000/cc	Acute on chronic subdural haemorrhage in left cerebral convexity, anterior Falx –mass effect, mild midline shift chronic micro bleeds	
APTT	69.7		
PT	13.3		
INR	0.86		
Electrolytes	normal	USG Abdomen and Pelvis	Upper abdomen normal Pelvis- Failed Intrauterine gestation
LFT	normal		
Creatinine	1.3		

On admission, patient conscious, afebrile well oriented, vision normal and no other neurological deficit other than phonation difficulty. Systemic examination normal. Investigations revealed deranged coagulation parameters, anemia with thrombocytopenia (Table 2).

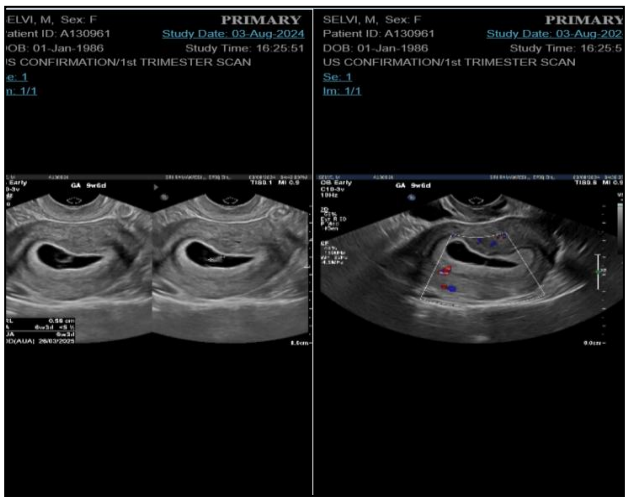

Figure 1: USG-failed IU gestation.

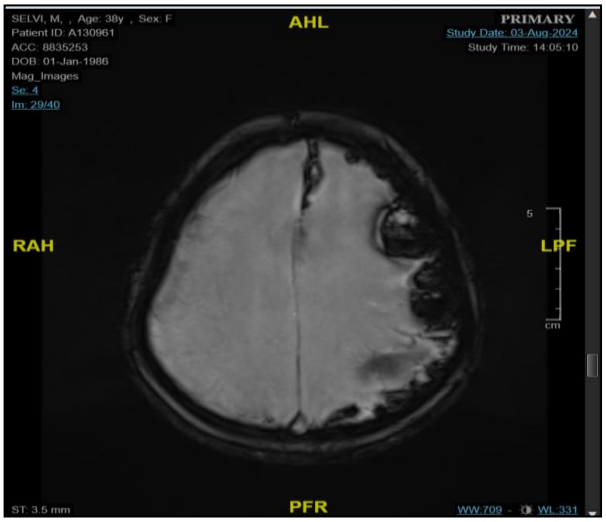

Figure 2: MRI brain-Subdural haematoma.

USG Abdomen and pelvis-failed intrauterine gestation and Upper abdomen normal (Figure 1). MRI brain revealed acute on chronic subdural haemorrhage in left cerebral convexity, anterior falx–mass effect, mild midline shifts with chronic micro bleeds (Figure 2).

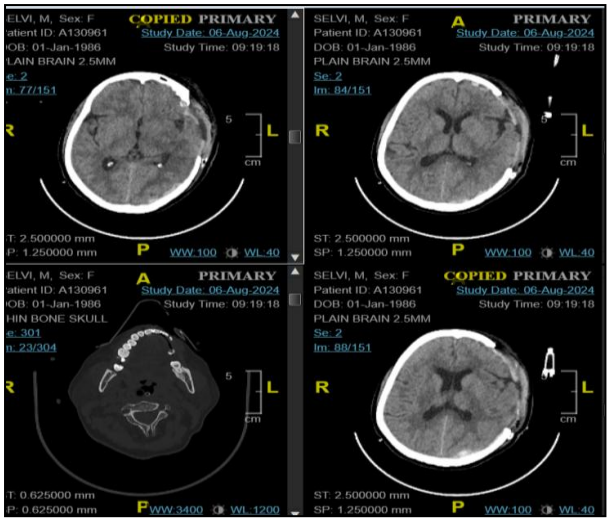

Figure 3: Postoperative imaging.

Multi-disciplinary team of obstetrician, neurologist, neuro surgeon, hematologist and cardiologist involved in the management of patient. Surgical intervention of SDH and Missed miscarriage was planned. Patient and family members counseled regarding the risk factors involved during surgery and anaesthesia and the guarded prognosis. Measures taken to improve the coagulation parameters by blood components transfusion since her general condition was stable. Under general anaesthesia, craniectomy with subdural haematoma evacuation done. Clots impinging on the cerebral surface removed and diffuse oozing tackled meticulously.

Bone not replaced. Skin flap closed with drain insitu. In lithotomy position, suction evacuation to empty the products of conception undertaken. There was difficulty in visualizing the cervix and explored after careful dissection. Under USG Guidance Products of conception (POC) removed and uterine cavity empty with no undue bleeding PV. POC sent for HPE Patient withstood the surgery reasonably well. Patient received 10 units of FFP, 6 units of platelets, 2 units of PRBC and one unit of Single donor platelet perioperatively. Immediate post operative period patient had one episode of convulsions and managed efficiently, subsequently recovered smoothly. Follow up CT Scan brain on 2 occasions-post operative D1, D3 normal and drain removed (Figure 3). Slowly regaining her speech with stable vitals and no other neurological deficit. Physiotherapy, oral fluids and antibiotics continued. HPE confirmed the products of conception. Patient discharged on 7th postoperative day.

DISCUSSION

Antiphospholipid syndrome (APS) is an autoimmune disorder that is associated with adverse pregnancy outcome. Pregnancy complications associated with are preeclampsia, thrombosis, autoimmune thrombocytopenia, fetal growth restriction, and fetal loss. It is estimated that the true incidence of the syndrome can be up to 1–2% or more in the general population.[3] antiphospholipid syndrome still represents an important treatable cause of recurrent pregnancy loss and known for its systemic arterial and venous thrombotic manifestations, like other autoimmune disorders, APS does not have a known etiology.

The diagnosis of APS requires at least 1 clinical criterion and 1 laboratory criteria. Women with the clinical features of APS should be tested for 3 antiphospholipid antibodies that have proven association with the diagnosis of APS: lupus anticoagulant (LA), anticardiolipin (aCL) antibody, and anti-beta-2glycoprotein I antibody. In a prospective PROMISSE study, LA was the main predictor of adverse pregnancy outcomes in APL carriers.4

There exist a complexity and heterogeneity of the mechanisms beyond the poor obstetric outcome in APS These antibodies interact with endothelial structures during trophoblast syncytium formation and disturb thrombaxane-prostacycline balance, upregulation of platelet aggregation and dysregulation of complement activation.

In women with well-recognized obstetric APS, anticoagulant prophylaxis is recommended during pregnancy and the postpartum period. Up to 5% of healthy individuals are known to have aPL antibodies. Non-traumatic Subdural haematomas (SDH) are uncommon during pregnancy and post-delivery. Thrombotic events in these patients consist in deep vein thrombosis (32%), stroke (13%), superficial thrombophlebitis (9%), pulmonary embolism (9%) and transient ischemic attack (7%), among other clinical manifestations.[5] Stroke is the most frequent arterial thrombosis.[3] The rate for thrombosis or stroke is 5-12%. These observations suggest that women with documented APS should not take estrogen-progestin combination oral contraceptives.

Our patient had gone through all bitter experiences on account of APS mentioned in domain 4-obstetric: Prefetal death, Fetal death, Preeclampsia with severe features, central nervous system dysfunction, placental insufficiency with severe features

Catastrophic antiphospholipid syndrome (CAPS) is a is a rare phenomenon with nearly 50% mortality and definite criteria to be fulfilled to fit into the diagnosis.[6] It is a life-threatening variant of APS with rapid onset of thrombosis and involves multiple organ systems. CAPS in pregnancy is very rare. Triple therapy, which consists of anticoagulants, corticosteroids, plasmapheresis and intravenous immunoglobulins, is typically administered to pregnant patients with CAPS.[7,8] Rapid deterioration can occur at any time warranting prompt early initiation of treatment. Landry-Guillain-Barré-Strohl syndrome (LGBSS) of acute inflammatory demyelinating polyradiculoneuropathy, although exceedingly rare in

pregnancy, can occur in patients with APS and lupus. Recurrent thrombotic events and recurrent CAPS are separate entities.[6]

In patients with SLE (Systemic lupus erythematosus) and APS, headaches may arise from diverse etiologies, including vascular diseases such as cerebral venous sinus thrombosis (CVST) and vasculitis, as well as infectious conditions like meningitis, and spontaneous intracranial hypotension.[9] This phenomenon could rarely mix with a severe neurological complication known as subdural hematoma (SDH).

Pregnancy with proven APS is always a challenging and the outcomes are unpredictable. Definitive treatment protocol exists with close vigilance of both mother and fetus. The course of pregnancy can deviate at any point of time.

CONCLUSION

With the multidisciplinary team work, we were able to treat the uncommon life-threatening manifestation successfully. Embarking on pregnancy in future after a critical episode is debatable. In certain situations, the decision making is vital to minimize the morbidity.

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