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Case Report

Case Report of Morbidly Adherent Placenta in Association with SLE: Placental Mesenchymal Disease (PMD)

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Received: May 24, 2019; **Published:** July 11, 2019

Abstract

Recurrent pregnancy loss is associated with systemic lupus erythematosus (SLE) is a known fact [1]. Placental mesenchymal disease is a rare entity [2] and here it has obscured the diagnosis of morbidly adherent placenta leading to sudden presentation at the surgical table which was effectively managed by proper planning and prompt decisions and efficient team work.

Keywords: *Systemic Lupus Erythematosus (SLE); Placental Mesenchymal Disease (PMD)*

Introduction

Recurrent pregnancy loss is associated with systemic lupus erythematosus (SLE) is a known fact [1].

Case Report

A 39 years old SB, $G_5P_0A_4$ came for a second opinion at 8 weeks of gestation as she had been advised termination on account of intrauterine synechia which would eventually lead to foetal growth restriction. She was extremely keen on continuing the pregnancy despite the foreseen complications. She did not have any live issues and her obstetric record was as in table 1.

Pregnancy duration serially	Outcome	Procedure
5 W	Unwanted pregnancy as under therapy for vitiligo	Medicated termination
8 W	Missed	D & E
26W	IUD	Induction followed by D & E
11 W	Missed	D&E: incomplete evacuation Later hysteroscopic removal of POC
8 W	Present pregnancy @ 8 weeks	Natural conception

Table 1: *Obstetric history of Mrs SB.*

D&E: Dilatation and Evacuation; POC: Products of Conception; W: Weeks.

She was under treatment for systemic lupus erythematosus with hydro chloroquine [3] 300 mg daily and was now in remission. She had herself stopped taking these medications recently during first trimester. According to her the SLE was triggered due to ultraviolet therapy that she had undertaken for vitiligo.

Her investigations for anaemia, hypothyroidism, insulin resistance, toxoplasmosis, thyroiditis, APLA were all normal. About 25 - 50% of SLE have antiphospholipid antibodies (APLA) but only some patients suffer thrombotic or obstetric complications related to antiphospholipid syndrome (APS) [4].

She had mild urinary tract infection which was treated with cefixime as per the sensitivity report. We decided to continue this pregnancy with all the due risks to the mother and the foetus explained and her antenatal period continued.

During the first trimester hypoechoic cystic spaces were noted in the chorionic plate and molar pregnancy was not the considered diagnosis. Chorioangioma was ruled out as this is a focal lesion present more on the foetal side and also is not seen during the first trimester usually.

Later scans too ruled out this possibility. She had minimal spotting at 10 weeks and a review scan revealed increased cystic spaces which were hypoechoic. The first trimester scan at 12.1 weeks was within normal limits except for slightly less liquor. The uterine artery PI (Pulsatility index) and the biomarkers were within normal limits.

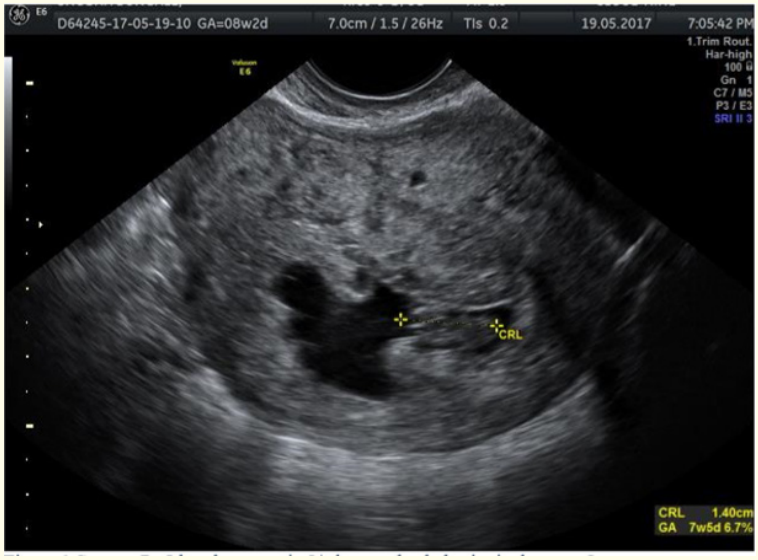

Figure 1: *Scan at 7w5d: adenomyotic/abnormal subchorionic lacunae?*

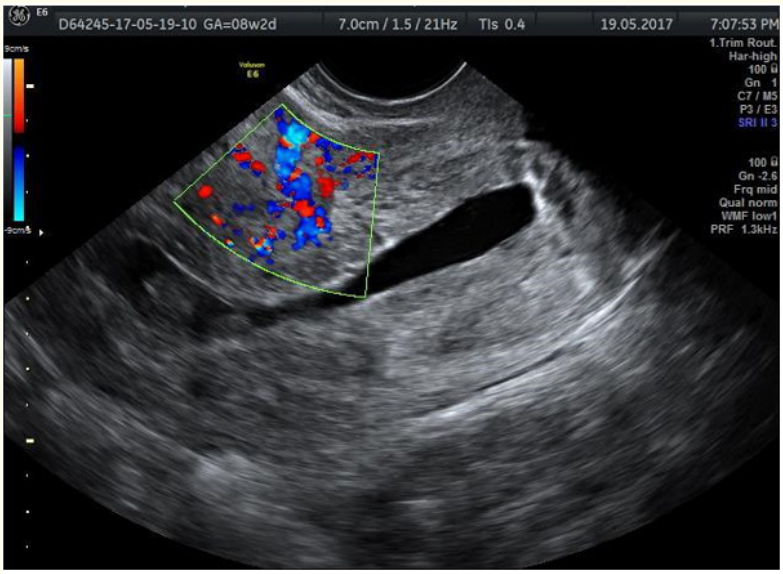

Figure 2: *Chorioangioma and partial mole was ruled out.*

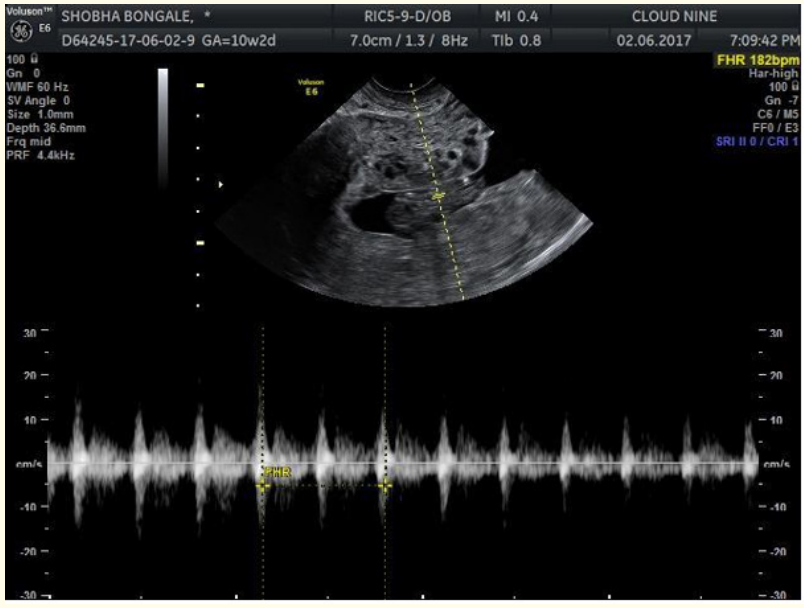

Figure 3: *Review at 10w+2 days cystic lesions were more in number and size and hypoechoic.*

The patient continued antenatal care with usual issues such as acidity, emesis and these were tackled with symptomatic treatment. At 17 weeks sonographic review the cystic lesions seemed to have enlarged and it was considered to be a placental mesenchymal disease. In addition, the baby had developed CTEV (Congenital Talipes Equinovarus). After this the maternal and foetal surveillance continued at regular intervals.

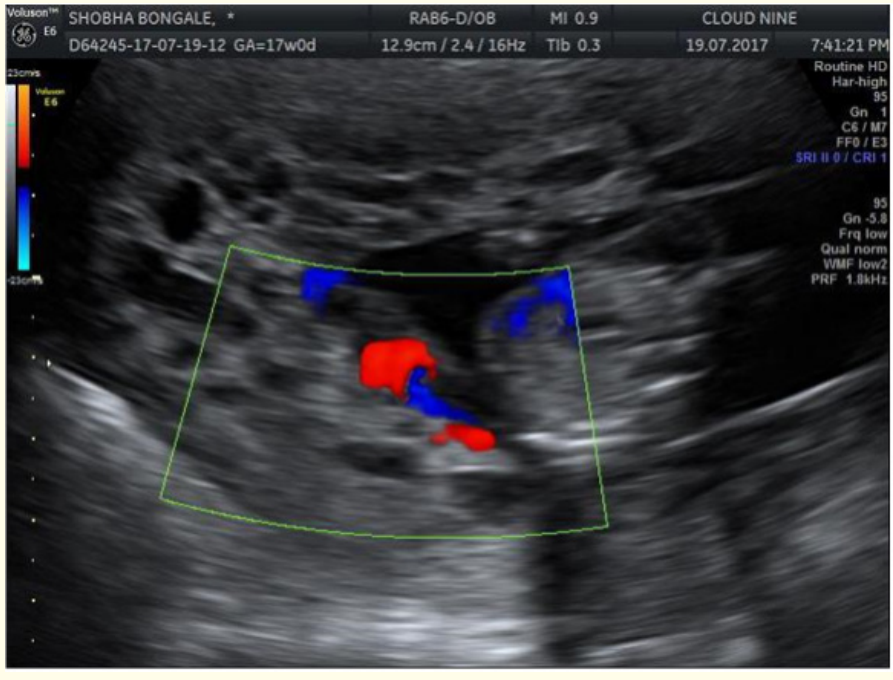

Figure 4: *Placental mesenchymal disease.*

The pregnancy period continued with 2 - 3 episodes of brisk vaginal bleedings and she was hospitalised and stabilised. Each time the sonography could not reveal any source of bleeding or evidence of placental separation or low placed placenta. At 24 weeks scan SGA (small for gestational age) baby was observed on biometric evaluation but the uterine artery and foetal dopplers were normal. The colour dopplers at 28+6 weeks showed umbilical artery resistance and close surveillance was undertaken. At 33 weeks 4 days gestation a decision of elective caesarean delivery was taken due to absent end diastolic flow in the umbilical artery. The expected birthweight was 1.2 kg and with magnesium sulphate for neuroprotection and antenatal steroids for lung maturity the patient was posted for an elective section. Preparedness for prematurity and possibilities of massive PPH (postpartum haemorrhage) and obstetric hysterectomy were kept in mind and essential provisions made.

Spinal anaesthesia was given after accurate preload and tranexamic acid was administered prophylactically. Uterus looked vascular and many surface vessels and myometrial sinuses were noted to be prominent. Baby was delivered and handed to the neonatology team. The 1 kg baby girl cried at birth and the necessary cord blood samples collected. Oxytocics were given and on palpation of the uterine cavity it was observed that the placenta was firm to hard in consistency, globally occupying the entire uterus and the myometrium was thinned out considerably at the posterior surface (Figure 5). The patient was oozing from areas where the fragmented placenta was trying to separate. Thus, it was a patchy placenta adherent at some places and loosely attached at others. A decision of obstetric hysterectomy was taken in view of morbidly adherent placenta, global placenta, massive haemorrhage and thinned out myometrium. Simultaneously massive blood transfusion protocol was initiated.

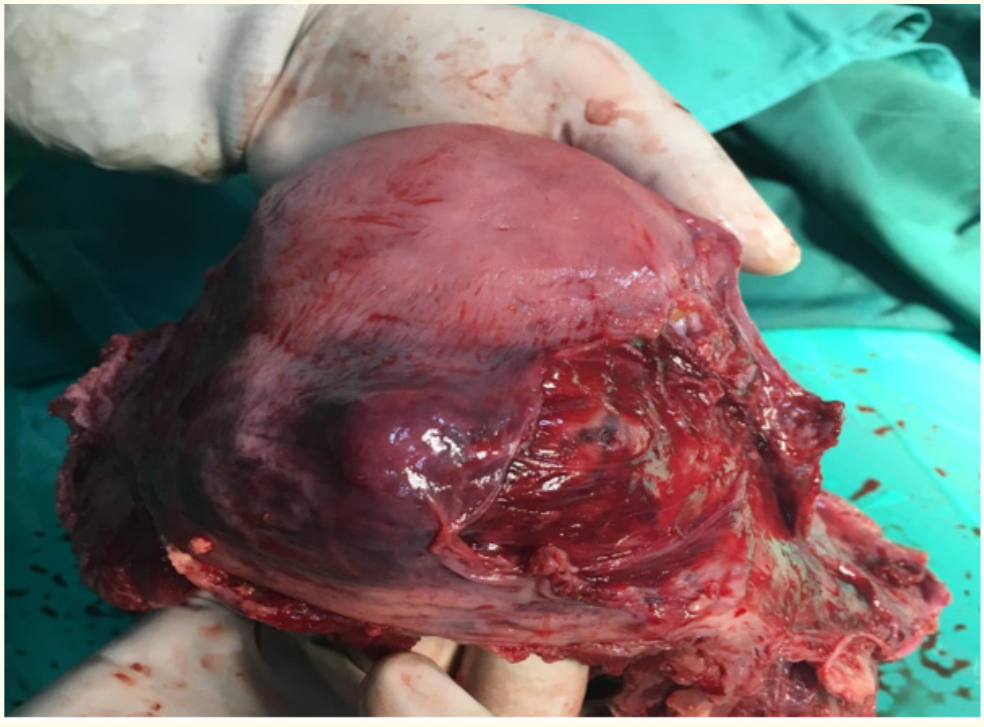

Figure 5: *Posterior uterine surface showing thinning: hysterectomy.*

Haemostasis was achieved and central line was initiated for CVP monitoring. In all 36 blood components were administered over a period of 32 hours and patient continued to remain hemodynamically stable. TRALI (Transfusion Related Acute Lung Injury) in early stages was treated with non-invasive positive pressure ventilation for 24 hours.

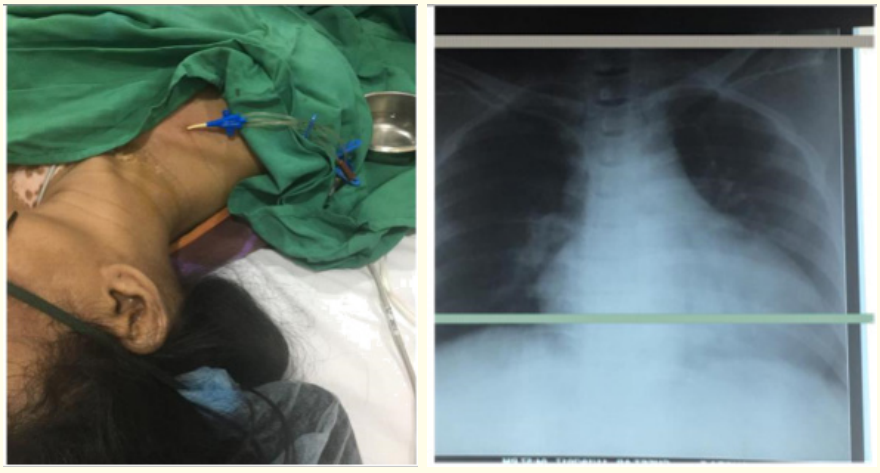

Figure 6: *Central line and XRC: TRALI.*

Post-delivery patient developed severe abdominal wall ecchymosis (Figure 7) for which expectant management was given Post hysterectomy the patient was stable and discharged from obstetric side after 12 days. The baby continued to be in the NICU and fared well.

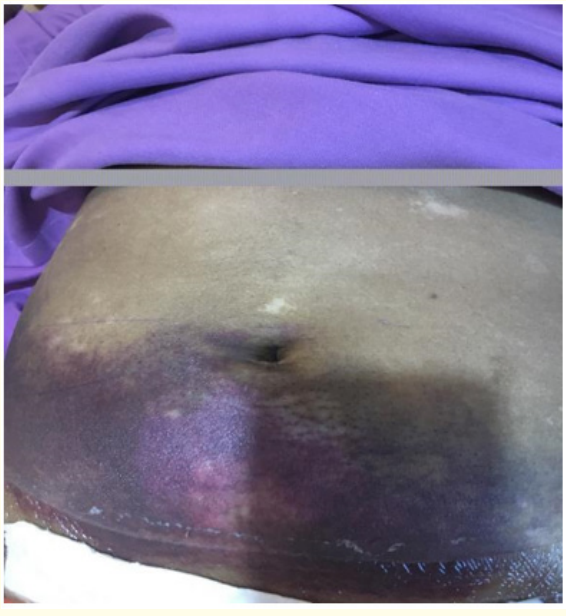

Figure 7: *Day 6 postop severe ecchymosis.*

Neonatal management

Baby cried after birth but soon developed grunting for which facial PEEP (positive end-expiratory pressure) administered. Subsequently baby settled and was transferred to neonatal intensive care unit. On clinical examination infant was small for gestational age due to growth restriction and had a bilateral CTEV of moderate degree. Infant started on feeds after 72 hours and gradually graded up to full feeds. There were no cutaneous, cardiac, haematological and hepatic manifestation of neonatal lupus [5,6]. After tolerance for oral feeds was confirmed the baby was shifted to the mother (11 days) and discharged afterwards. At follow up infant had serial plasters and tendon release surgery for CTEV [7,8]. Infant is growing well and developed milestones according to corrected age.

Histopathology: The specimen of the uterus and the attached placenta was sent for histopathology. Gross examination revealed placentomegaly with cystic spaces with patches of normal trophoblastic tissue. The cystic structures represent cystic dilatation of the stem villi. Occasionally, there is thrombosis and aneurysmal dilation of stem villous vessels. Vessels on the foetal surface may be tortuous and dilated and may be thrombosed. Histologically, oedematous stem villi with dilated vessels and absent trophoblastic proliferation, The vessels were thick walled, with fibromuscular hyperplasia. Terminal villi were normal The report mentioned placenta accrete as the final diagnosis with thinning of the posterior wall. The invasion was only up to the serosa surface and did not reach beyond. It also mentioned the possibility of placental mesenchymal dysplasia.

Discussion and Conclusion

Placental mesenchymal dysplasia (PMD) is a radiological entity first described in 1993 [9]. It is an uncommon vascular anomaly of the placenta characterized by placentomegaly with multicystic placental lesions on ultrasonography and mesenchymal stem villous hyperplasia on histopathology. Placental mesenchymal dysplasia is considered in the differential diagnosis of cases of multicystic placental lesion such as molar pregnancy, chorioangioma, subchorionic hematoma, and spontaneous abortion with hydropic placental changes [9]. PMD should also be considered as an early sign of placenta accrete as is known to be associated with vascular abnormalities. However, lack of high-velocity signals inside the lesion and a normal karyotype favour a diagnosis of PMD. PMD must be differentiated from gestational trophoblastic disease. Texture of the placenta is recommended to be evaluated sonographically at 16 to 17 weeks of gestation and such cystic spaces, rounded hematomas and infarcts should be evaluated and have been known to be associated with preterm births, growth restriction and preeclampsia. Scarred uterus may initiate abnormal placentation and the entire uterine cavity may be involved in this. PMD is associated with intrauterine growth restriction and intrauterine foetal demise and early neonatal death [10]. The cause of foetal death in many cases is foetal vascular obstructive pathology causing longstanding, severe foetal hypoxia characterized by chorionic vessel thrombosis.

PMD is postulated be a result of the maternal nondisjunction error during the first meiotic division this leads to chimerism due to the formation of diploid as we; as paternal isodisomic cell lines [11]. Apart form FGR, fetal demise, PMD is associated with transient neonatal diabetes, paternal disomy 6, trisomy 13 and Klinefelter syndrome. PMD has a female predominance.

Retrospective analysis: The 4 D scan at 17 weeks (Figure 8) was actually revealing and it did show a global placenta. However, accrete was not appreciated at any antenatal scanning proving the less sensitivity of the sonography to identify myometrial invasion.

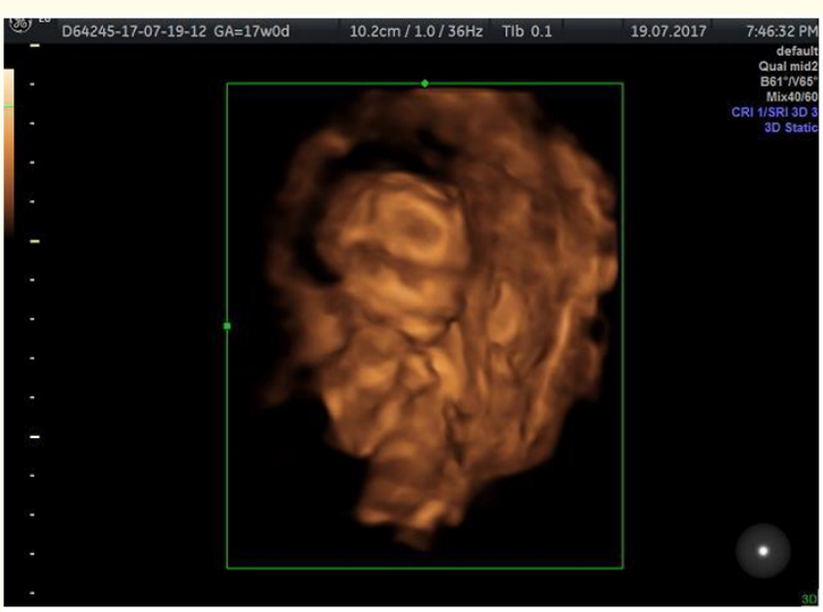

Figure 8: *17 weeks 4 d scan.*

What helped?

1. Cooperative patient and her husband amenable to counselling and compliant with a lot of trust in our treatment.
2. Fantastic team work: quick decisions by the obstetrician and the anaesthesiologist and the nursing assistance and continuous support from the blood bank and swift initiation of transfusion and resuscitative protocols.
3. Prenatal counselling sessions with the entire team of obstetricians, neonatologist, anaesthesiologist and the radiologist.
4. Infrastructure: with all the necessary monitoring and supportive gadgets.
5. Availability of round the clock support staff, continuous vigilance by duty doctors and guidance and timely interventions by the intensivist.

Lessons learnt: SLE and previous uterine surgeries and not only caesarean deliveries predispose the mother to morbidly adherent placenta where a hysterectomy may be necessary and massive blood transfusion and good infrastructure is necessary for delivery.

Careful antenatal risk assessment, radiological evaluation can help suspect MAP or adverse outcomes and help to be on guard.

All pregnancies are at risk of sudden turn of events and proper evaluation, vigilance and proper patient cooperation helps achieve good results despite all odds.

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Volume 8 Issue 8 August 2019