Case report

A rare case of acute onset vasculitis in pregnancy

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Abstract

Vasculitis in pregnancy is a rare condition. When a patient presents, it is usually a pregnant woman with a previous diagnosis of vasculitis. We present a case of vasculitis presenting for the first time during pregnancy with a remarkable recovery following the delivery.

A 32 year old, previously healthy primi gravida presented at a gestation of 28 weeks and four days with a history of fever for three days. She developed a vasculitic type rash over both lower limbs. Investigations revealed urinary sediments, proteinuria, haematuria, and pulmonary consolidation, suggesting skin, renal and pulmonary involvement. Patient did not improve despite adequate treatment with antibiotics and antiviral treatment. Not only the investigations done to identify a possible aetiology leading to a secondary vasculitis but also the tests done to classify a primary vasculitis, became negative. Skin biopsy suggested leucocytoclastic small vessel vasculitis.

Patient symptomatically improved with methylprednisolone pulse therapy and plasmapheresis. Termination of pregnancy was considered with a view of commencing cytotoxic therapy to prevent permanent pulmonary and renal injury.

On day 18th of the illness, live baby was delivered by Caesarean section at a gestation of 31 weeks. Patient improved dramatically following the delivery. Patient became completely asymptomatic over few days, proteinuria and pulmonary fibrosis reversed in about one months' time with steroids.

Key words: pregnancy, vasculitis, small vessel vasculitis, leucocytoclastic

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Introduction

Vasculitides are a heterogeneous group of autoimmune diseases, all characterized by inflammation of blood vessels (vasculitis) and subsequent ischemia and damage to the organs supplied by these vessels[1]. It is a rare condition to occur in pregnancy. Vasculitis can be classified according to the size of the vessel involved as small, medium or large vessel vasculitidies (Chapel Hill nomenclature)[1]. Vasculitis can result as a secondary manifestation of other diseases or as a primary phenomenon[1]. Outcome of pregnancy has shown to be better with this vasculitis first presenting during the pregnancy in contrast to preexisting vasculitis[2].

Case report

A 32 year old, previously healthy primi gravida admitted at a gestation of 28 weeks and four days with a history of fever of 39 °C for three days and bilateral knee joint arthralgia. Clinical examination on admission was unremarkable with a blood pressure of 100/60 mmHg. Full blood count done on admission showed marginal neutrophil leukocytosis with normal platelets. The urine full report (UFR) was positive for proteins and there were sediments with red cells. Serum Creatinine was 46 umol/L (Normal Range 35-71 umol/L) and urine protein creatinine ratio was 0.89. Ultrasound scan (USS) of the abdomen and kidney showed normal findings with an adequately grown live fetus. Patient was commenced on intravenous (IV) Coamoxyclav 1.2g, 8 hourly and IV Metronidazole 500mg, 8 hourly after sending urine for culture and antibiotic sensitivity with a working diagnosis of vasculitis.

On day five of the illness patient developed a non-blanching macular papular vasculitic type rash over both lower limbs more over the ankle (Figure 1) which became coalescent with time (Figure 2).

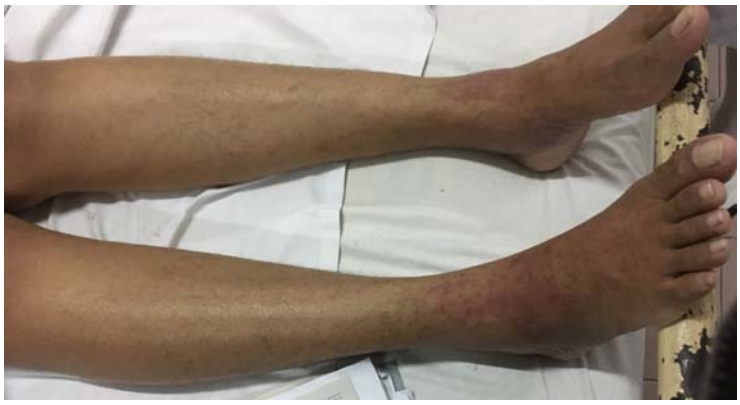

Figure 1. **Rash on day 5.**

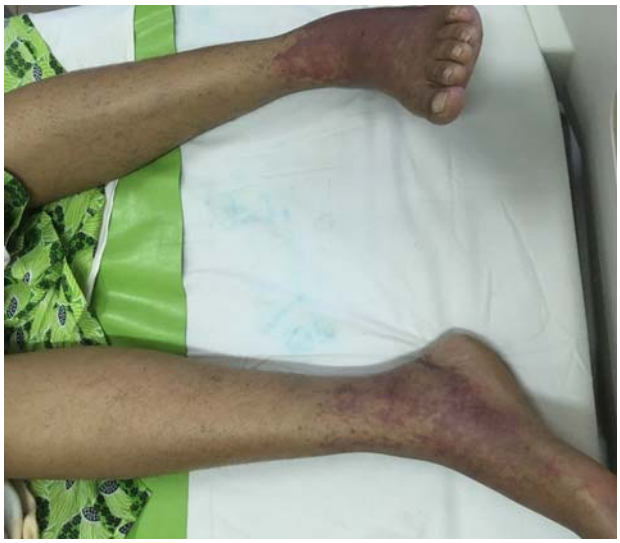

Figure 2. **Rash on day 7.**

The patient was referred to the rheumatology team, who suggested to commence oral Prednisolone 10 mg mane along with IV Ceftriaxone 1g two times daily from day 7, considering the possibility of a vasculitidies. However Predisolone was deferred as the repeat USS of the kidney showed evidence of pyelonephritis.

Day 8 of the illness patient became tachypnoeic with a 92% of arterial oxygen saturation on air. There were clinical evidence of left lung consolidation and the arterial blood gas (ABG) showed a Type I respiratory failure. Chest X ray postero-anterior view (CXR PA) confirmed the left sided massive lung consolidation (Figure 3).

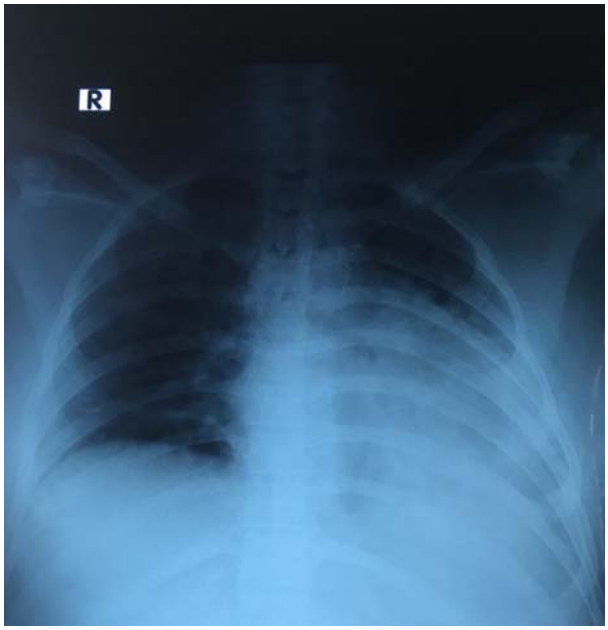

Figure 3. **CXR PA showing a massive lung consolidation.**

Patient was admitted to the Intensive Care Unit (ICU) on the same day. She was started on IV Meropenem, IV Ceftazidime, IV Teicoplanin, oral Clarithromycin, Tamiflu, Prednisolone 10 mg daily dose and subcutaneous Enoxaparin. Adequate respiratory support was given with Oxygen via face mask to maintain the saturation above 96%.

Liver transaminases were marginally elevated with a C-reactive protein (CRP) level of 117 mg/L (Normal less than 8 mg/L). Anti-Nuclear Antibody (ANA), Rheumatoid factor, urinary and blood cultures were negative. Complement 3 and 4 levels were within normal range. Electro-cardiogram (ECG) and 2D Echocardiogram were normal. Low procalcitonin levels (0.01 ng/ml) suggested that it is unlikely to be bacterial infection.

High Resolution Computed Tomography (HRCT), C-Anti-neutrophil cytoplasmic antibody (ANCA), P-ANCA, immunoglobulin A (IgA), and investigations for retrovirus, cytomegalovirus (CMV), Mycoplasma, Rickettsia, Melioidosis and Pneumocystis jiroveci Pneumonia (PJP) infections were arranged. The above investigations were negative.

Irrespective of the antibiotics and antiviral treatment, high spiking fever continued. She became more dependent on oxygen and the respiratory rate increased to 40/min. HRCT showed evidence of diffuse pulmonary hemorrhage.

Multidisciplinary team meeting (MDT) was arranged with the collaboration of all the relevant disciplines. MDT concluded that, even after adequate covering of any atypical infection, not responding to therapy raise the suspicion of a primary vasculitis. Thus a decision to start IV Methylprednisolone 1g/daily for 3 days after performing a Broncho Alveolar Lavage (BAL) was made.

Bronchoscopy was inconclusive of pulmonary hemorrhage and the samples from BAL were sent for, cytology, testing for Pneumocystis Jiroveci, influenza A and B, CMV, cultures including tuberculosis, Aspergillus, other bacteria and fungi. All were negative.

Skin biopsy also was performed which suggested leucocytoclastic small vessel vasculitis. All other investigations done for aetiological identification became negative including C-ANCA and P-ANCA.

Patient became fever free following the first methylprednisolone pulse (Figure 4). Plasmapheresis was also commenced simultaneously. CRP came down to 9.2 mg/L, but the respiratory distress and proteinuria were persistent. Repeat HRCT showed evidence of possible lung fibrosis. Decision was taken to start IV Cyclophosphamide 750mg/m^2 after termination of pregnancy to prevent permanent damage to maternal organs.

On the 18th day of the illness, live baby weighing 1475g, was delivered by Caesarean section at a gestation of 31 weeks and one day. Baby was provided with neonatal ICU care due to prematurity. Plasmapheresis and oral Prednisolone 60 mg daily continued.

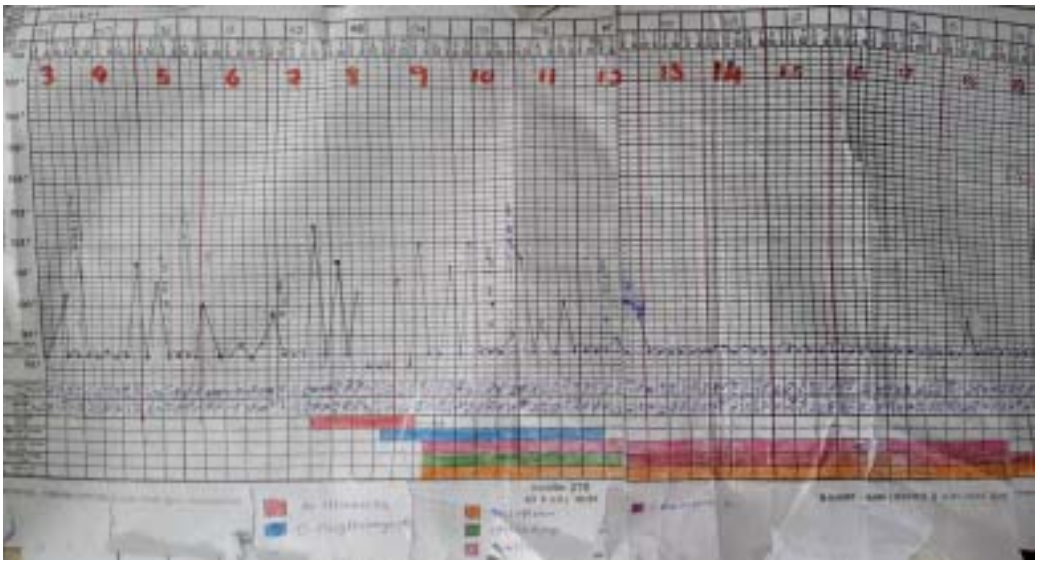

Figure 4. **Temperature monitoring chart.**

Mother improved dramatically following the delivery. Plasmapheresis was stopped after 6 cycles. Oral high dose Prednisolone was continued for one month and then tailed off over a period of two months. Patient became completely asymptomatic over few days, proteinuria and pulmonary fibrosis reversed in about one months' time.

Discussion

Our patient developed systemic vasculitis for the first time during the pregnancy. There are few cases reported in the literature who present with systemic vasulitis for the first time during pregnancy[3]. Skin biopsy confirmed small vessel leucocytocstlaic vasculitis. Owing to the rarity of this condition there was a delay in making the diagnosis and providing the definitive care.

Acute onset of this condition led to the suspicion of an infective aetiology leading to a secondary vasculitis. Low procalcitonin levels and the condition not responding to antibiotics excluded the possibility of a bacterial infection. Other causes which could give a similar presentation include infections such as hepatitis B, C, retrovirus, mycobacteria, connective tissue diseases such as systemic lupus erythematosus, rheumatoid arthritis, neoplasms and some drugs[4]. Extensive investigations performed excluded the possibility of the above causes leading to secondary vasculitis.

Small vessel vasculitis which could give rise to a similar picture are ANCA associated vasculitis, and Henoch-Schönlein purpura[5,6]. But neither the clinical presentation nor the investigations supported these either. Management of vasculitis during pregnancy and their outcome has been evaluated in several studies[3,7,8]. These studies have shown increase incidence of preterm delivery. As in the case described sometimes it is iatrogenic[3,7,8]. In this patient there was renal involvement with acute life threatening lung involvement. This required aggressive therapy with high dose steroids and plasmaphoresis. Pregnancy was terminated at 32 weeks with a view of commencing Cyclophophamide treatment.

In about 35% of cases small vessel systemic vasculitis flare up during pregnancy[3]. Our patient improved dramatically following the delivery indicating that her condition was pregnancy related. She required only two months of steroids after delivery. This is more in keeping with diagnosis of idiopathic leucocytoclastic vasculitis occurring in pregnancy.

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