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BMH Med. J. 2018;5(2):55-58 **Case Report**

A Rare Case Of Acute Lupus Nephritis In Pregnancy

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Systemic lupus nephritis is an autoimmune connective tissue disorder that commonly affects women in the reproductive age group. 80% of women with systemic lupus erythematosis develop lupus nehritis. Pregnancy in any class of lupus nephritis is associated with increase in adverse maternal and fetal outcomes. Disease should be quiescent for at least 6 months prior to conception for optimal maternal and fetal outcome. Rarely lupus can manifest denovo in pregnancy. We are reporting a case of acute class 4 lupus nephritis which was diagnosed for the first time in pregnancy in a young woman.

Case report

A young woman, primigravida at fifth month of amenorrhoea presented with generalised oedema and oliguria to the Emergency Department. Her complaints were generalised oedema for one and half months, followed by joint pain involving small joints of all four limbs associated with fever. Redness over the joints and difficulty in weight bearing were also noted her. She was initially seen in a local hospital and received symptomatic treatment. She started to develop red coloured urine and decreased urine output for 2 weeks. A diagnosis of systemic lupus erythematosus (SLE) with lupus nephritis was made and she was referred to here. She had headache, fever, breathing difficulty with pain and swelling in the perineal region. She had no past history of collagen vascular disease, hypertension or family history of renal disorders.

On examination she was pale with facial puffiness and generalised anasarca. Her pulse rate was 84 per minute and blood pressure 120/90 mm of Hg. Abdominal examination showed uterus corresponding to period of amenorrhoea (20 weeks of gestation). Local examination showed gross vulval oedema. She was provisionally diagnosed to have SLE with renal involvement. She was severely anuric and in renal failure.

On admission her lab investigations showed hemoglobin of 11.5gm% with leucocytosis and neutrophilia. Her blood urea was 102mg%, serum creatinine 2.73mg%, serum sodium 122meq/l, serum potassium of 4.8meq/l her liver function tests were normal except for total protein of 3.1, albumin of 1.0, globulin 2.1, A/G ratio of 0.5. Her ANA profile showed RNP/Sm+++ (strong positive), Sm ++, AMA M2 strong positive, Ribosomal-p-protein (RIB) borderline positive. Her C3 was low (64.88) and C4 normal (20.48).

Potential maternal and fetal complications were explained and she was given intensive care treatment with steroids and other supportive measures including human albumin infusion and fresh frozen plasma (FFP) transfusions. She was taken up for dialysis via right jugular vein. Owing to the poor maternal condition and acute renal shut down, she was planned for termination of pregnancy after explaining all complications. She was given Mifepristone orally followed 24 hours later by vaginal Cytotec. Meanwhile Glycerine Magsulf and saline dressing was given locally to reduce vulval oedema. After 2 days of vaginal Cytotec, Foley EAS induction was done and patient expelled a dead fetus weighing 280 grams.

She was continued on hemodialysis. Anemia was corrected with blood transfusion. Antibiotics, albumin infusion and other supportive measures continued. She was seen by Rheumatologist and three doses of injection Methylprednisolone were given. Renal biopsy was deferred for later date due to thrombocytopenia (platelet 40000). She received 20 hemodialysis and was discharged after three weeks of hospitalisation when her hemoglobin restored to 10.7 gm%, total count reduced to 6200, blood urea 53 and serum creatinine 2.56. Clinically she had improved a lot with better urine output and reduced anasarca.

Renal biopsy was done later (**Figures 1-3)** and was reported as class 4 lupus nephritis-rapidly progrssive type of renal failure. Microscopically about 21 glomeruli were seen, in that 3 were globally sclerosed, 8 with fibrocellular crescents, globally mesangial and endocapillary proliferation were seen and 2 wire loop lesions too were visualised. Immunoflourescence revealed mesangial and capillary wall deposits of IgG3 in 9 glomeruli with Kappa and Lambda positivity. IgM, IgA, C3 and C1q were absent. She was diagnosed as lupus nephritis, ISN/RPS class IV-G (A/C) activity score 5/24, chronicity score 4/12. She is on regular follow up and lupus controlled on chloroquine and steroids.

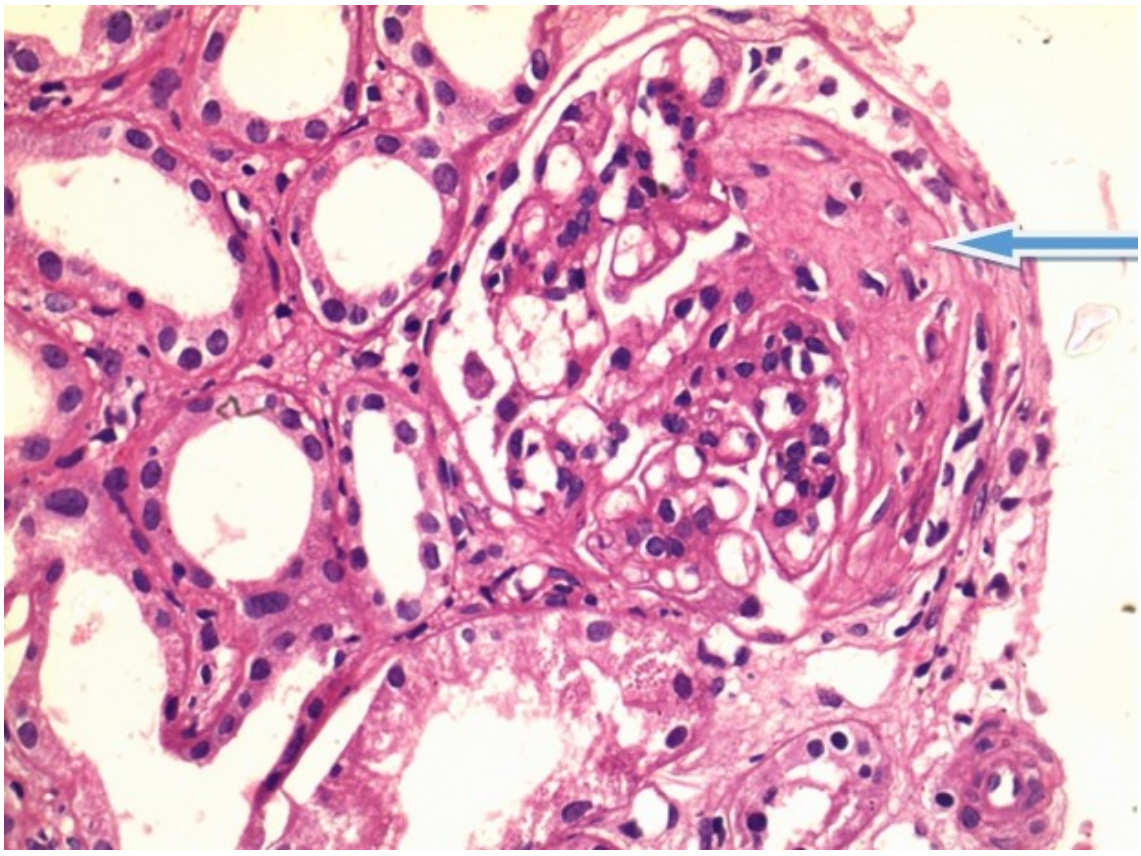

Figure 1: Extracapillary proliferation (crescents) - pathognomonic lesion indicating activity an classification as class IV lupus nephritis

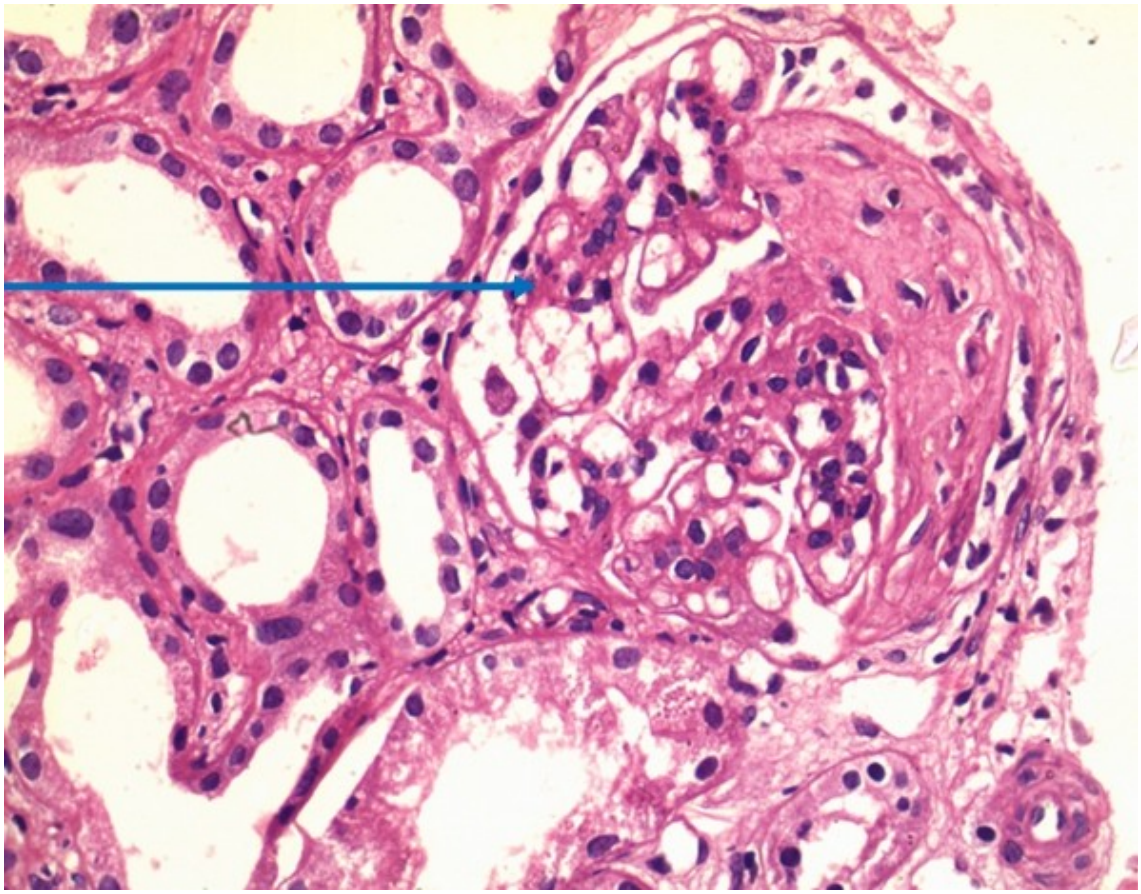

Figure 2: Wire loop lesions-glomerular capillaries stain deep red and appear acellular and thickened due to heavy deposition of subendothelial immune complex deposits

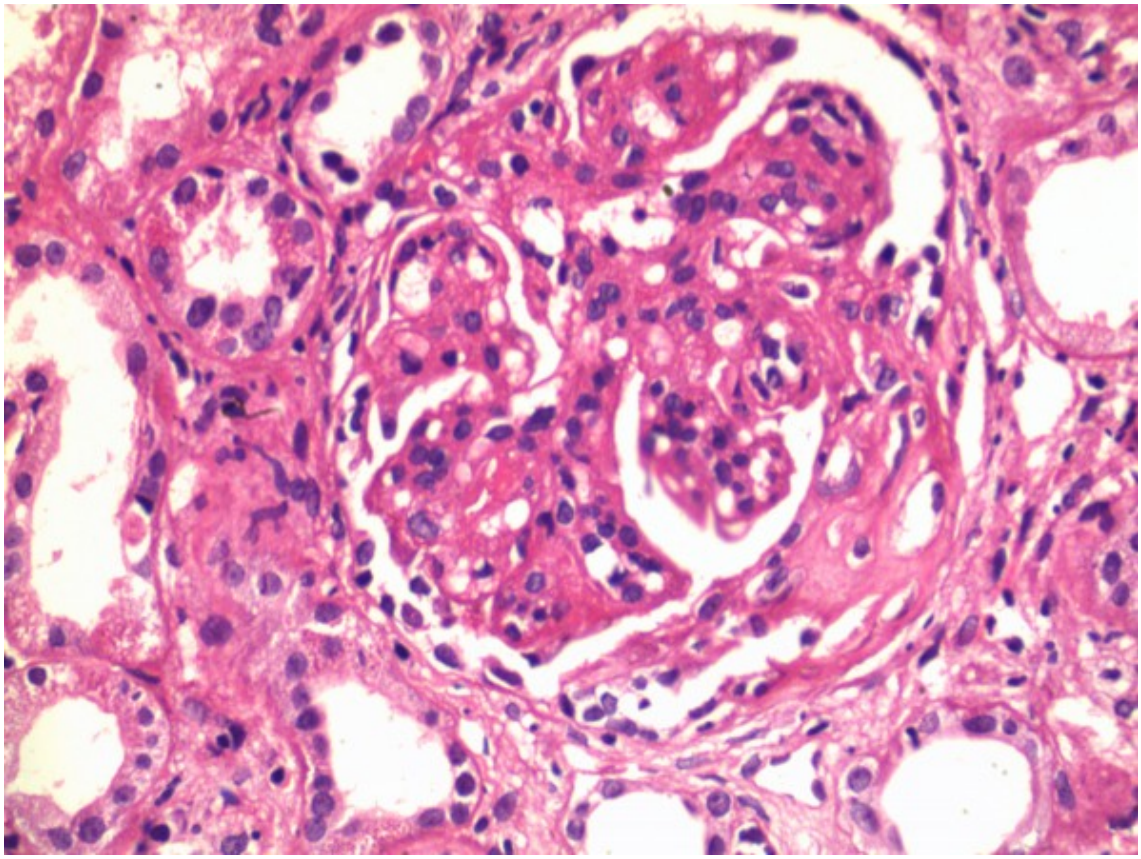

Figure 3: Mesangial proliferation

Discussion

Here lupus flare and SLE was first diagnosed in pregnancy and nothing could be done to save the fetus. When diagnosed prior to pregnancy, as mentioned, lupus need to be controlled with drugs at least 6 months prior to conception [1]. SLE in pregnancy with acute flare can lead to fetal loss, preterm birth, intra uterine fetal growth restriction, neonatal lupus syndromes and maternal complications like gestational diabetes, osteoporosis, avascular necrosis, hypertension, pre eclampsia, eclampsia, stroke, HELLP syndrome and even maternal death [2]. Pre eclampsia mimics the disease symptoms of lupus nephritis and can confuse the diagnosis.

The risks involved may be minimised by appropriate timing of pregnancy and optimization of therapy prior to conception. NSAIDs [3] and steroid [1] exposure has to be limited to the minimum during pregnancy. However in case of disease flares, short courses of high doses and/or intravenous pulse methylprednisolone can be used. Hydroxychloroquine should be continued in all pregnant women with SLE [1]. Azathioprine is one of the few immunosuppressive agents that has documented safety in pregnancy [4]. In cases of recurrent pregnancy loss associated with SLE, aspirin in combination with prophylactic doses of heparin significantly reduce the risk [1].

Advanced technology and better understanding of the disease has significantly improved outcomes in lupus pregnancies over the last few years. Pregnancy can be successful in most women with lupus nephritis. Pregnancy should be planned and management strategy should be agreed with the patient prior to conception. Women with SLE need treatment throughout pregnancy. Women whose disease is well controlled can conceive even though infertilty can occur as a part of the disease or due to cytotoxic agents. During pregnancy lupus commonly flares after the 32nd week, even in ladies on steroids and immunosuppressants. Steroids are commonly increased in the peripartum period and continued till the post partum period is over.

It is essential that maternal disease is well controlled prior to, during and after pregnancy to ensure the best possible outcome for the mother and child. SLE requires a multi disciplinary approach for its diagnosis and successful management.

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