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New Onset Crescentic Ig a Nephropathy in Pregnancy: Case Report

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Abstract

IgA nephropathy (IgAN) (Berger's disease) is the most common primary glomerulonephritis in the world among patients undergoing renal biopsy. Due to complex pathophysiological changes that occur during pregnancy it is difficult to diagnose renal parenchymal diseases presenting for the first-time during pregnancy. This is a rare case of Ig A nephropathy presenting during pregnancy as preeclampsia with nephrotic syndrome. She also had renal dysfunction at presentation and her kidney biopsy performed in second trimester showed crescentic Ig A nephropathy. She delivered by preterm vaginal delivery which was complicated by community acquired pneumonia secondary to immunosuppression

Keywords: Ig A nephropathy, nephrotic syndrome, preeclampsia, preterm

Introduction

New onset renal parenchymal diseases with presentation first during pregnancy poses difficulty in diagnosis due to pregnancy related changes and pregnancy specific complications like preeclampsia which also presents with increased blood pressure and proteinuria. IgA nephropathy (IgAN), Berger's disease (described in 1968) is the most common primary glomerulonephritis in the world among patients undergoing renal biopsy.[1]

The most consistent clinical finding in IgA Nephropathy is the presence of hematuria; however, proteinuria, the presence or development of arterial hypertension, and reduced eGFR at diagnosis are the clinical factors that influence the development of CKD. The amount of proteinuria has been significantly related with worse renal outcomes.[2] Nephrotic syndrome usually occurs only in more advanced stages of the disease.[3] Here we are going to present a case where the patient presented during second trimester of pregnancy with severe preeclampsia, anasarca, new onset nephrotic syndrome with deranged renal function test later on diagnosed to be a case of new onset crescentic IgA nephropathy.

Case Report

23-year-old female, third gravida (G3P1L1A1) with first normal vaginal birth followed by spontaneous abortion in 1st trimester due to unevaluated cause presented at 24 weeks of gestation with severe hypertension BP of 170/98 mm hg and swelling of both lower limbs till mid-thigh, vulval edema and facial puffiness. Patient did not have any history of preeclampsia or any other medical complications in the first pregnancy. Patient was admitted in view of suspicion of early onset severe pre-eclampsia for further evaluation and management. Patient had undergone last ANC checkup at 16 weeks when she was asymptomatic and blood pressure was recorded normal. Initial investigations are summarized in Table 1.

Obstetric ultrasound done for fetal surveillance showed growth of fetus corresponding to the

gestational age. In view of presence of Hypertension, nephrotic range proteinuria, Microscopic Hematuria, acanthocyturia, deranged renal function a working diagnosis of glomerulonephritis presenting as nephrotic syndrome was made. There was no history of significant hair loss, oral ulcers, skin rashes, joint pain suggesting autoimmune disorders. Serology for Hepatitis B, Hepatitis C and HIV was negative. Serology for ANA, DsDNA, ANCA was negative with normal complement levels. APLA profile was normal. Renal Ultrasound showed normal sized kidneys. Kidney Biopsy of the patient was done at 24 weeks by positioning the patient in right lateral position in ultrasound guidance. Patient was started on oral diuretics, antihypertensives (tab Labetalol 200mg BD with tab nicardipine sustained release 20mg BD), LMWH thromboprophylaxis, water restriction, low salt (2g/day) and normal protein diet (0.8g/kg/day). Biopsy revealed crescentic IgA nephropathy with oxford MEST-C score of 2 (Figure 1). Since cyclophosphamide and mycophenolate could not be initiated in view of pregnancy patient received 3 pulse doses of 500mg methyl prednisolone and was later shifted to prednisolone 1mg/kg. Blood pressure of the patient was controlled with antihypertensives and vulval edema reduced. Patient was discharged after 16 days of hospitalization and advised to follow-up in OPD after 2 weeks. However, patient could not follow up due to second wave of Covid pandemic and presented at 32 weeks of gestation with NYHA grade 4 breathlessness and pain abdomen. There was no h/o fever. Patient was still taking 1mg/kg prednisolone (7 weeks after discharge). On examination BP was 170/90 mmhg, respiratory rate of 36/min, SPo2 – 86% on room air and 93% on 6L oxygen by face mask, pedal edema till mid-shin. Patient was in second stage of labour and delivered vaginally, a female child weighing 1.38kg. Baby cried immediately after birth with APGAR score of 7. Baby was shifted to NICU for observation. Rapid antigen test and RTPCR for COVID 19 was negative. On auscultation right infrascapular and interscapular area had bronchial breath sounds. Patient was shifted to ICU and Non-invasive ventilation was started. Steroid dose was reduced to 0.5 mg/kg and furosemide infusion was started with piperacillin tazobactam and azithromycin as cover for community acquired pneumonia. Oseltamivir was also initiated to cover influenza virus.

Patient x-ray showed right lower zone consolidation with air bronchogram. Serum procalcitonin was 39 ng/ml. Patient was treated as community acquired pneumonia. 24 hour urine protein was 1gram/gram creatinine and creatinine was 1.47 (Table 2) . Sputum and blood culture was sterile. Patient improved clinically and was weaned from non-invasive ventilation after 72 hours. Patient was discharged on 7th day in healthy condition with advice for regular follow up.

Discussion

This is a rare case of Ig A nephropathy presenting during pregnancy as crescentic glomerulonephritis causing nephrotic syndrome. In our patient during evaluation of preeclampsia we found she had nephrotic range proteinuria and raised serum creatinine which led us to doing further evaluation. Due to complex pathophysiological changes that occur during pregnancy it is difficult to diagnose renal parenchymal diseases presenting for the first-time during pregnancy. Preeclampsia is the most common cause of nephrotic syndrome in pregnancy.[4] Primary renal disease presenting as nephrotic syndrome is known to occur only in 0.028% of pregnancies.[5] Gold standard for diagnosing any renal parenchymal disease is kidney biopsy which was diagnostic of Ig A nephropathy. Sadly, there are no serum markers or radiological investigation for diagnosis of IgA nephropathy and it is diagnosed only by histopathological examination.

Pathognomonic of the condition are prominent, globular deposits of IgA (often accompanied by C3 and IgG) in the mesangium and less prominently along the glomerular capillary wall.[6]—Nephrotic syndrome without hypertension or renal impairment is reported to have good outcomes.[7]Elevated creatinine is an independent risk factor for adverse pregnancy outcomes.[8] The pathological determinants are the presence of mesangial hypercellularity, endocapillary proliferation, segmental sclerosis and tubular atrophy.[9] Additionally, mesangial C3 deposition and decreased serum C3 levels are associated with disease progression and poor kidney outcomes in patients with IgAN.[10] presence of cellular crescent is also a marker of poor prognosis. Incidence of preterm delivery and delivery by

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caesarean section and low birth weight baby is increased in pregnancy with nephrotic syndrome.[8] Our patient had renal impairment and hypertension, codominant C3 deposition with IgA in the mesangium and presence of cellular crescents as poor prognostic factors. Patient had preterm delivery and low birth weight baby as adverse events. There are no Management guidelines of Crescentic IgA nephropathy in pregnancy. Low dose Pulse cyclophosphamide therapy has shown good results in crescentic IgA nephropathy with nephrotic syndrome, however the same could not be started in our patient due to teratogenic effects of cyclophosphamide in pregnancy.

Conclusion

Any pregnant women coming with raised BP should be worked up thoroughly and all the differential diagnosis should be kept in mind. Even though IgA nephropathy presenting for the first time during pregnancy is very rare it should always be kept in mind that renal parenchymal diseases is a differential in a patient with raised BP and anasarca even in pregnancy.

Table 1 – Summary of investigations during first admission

Complete blood count	Hb -9.4 mg/dl T L C – 12300/ml Platelet – 2.9 lakh/ml
Renal function test	Blood Urea – 37mg/ dl, creatinine – 1.66mg/dl, sodium 133 mmol/L, potassium 4.0 mmol/L.
Liver Function Test	Total Bilirubin – 0.27 mg/dl, Direct Bilirubin 0.02 mg/dl, AST 15 U/L, ALT 9 U/L, Albumin 1.8 mg/dl, Globulin 2.3 mg/dl
Urine routine microscopy	Protein – 3+, RBC – 5-10, Pus cell 5-10, Dysmorphic RBC- Present (8/HPF) No Casts
Lipid Profile	Total Cholesterol – 395mg/dl, Triglycerides-379 mg/dl, LDL – 243 mg/dl.
24 urine protein	5.6 g /g of creatinine

Table 2 – summary of investigations at the time of second admission

Complete blood count	Hb 9.9, TLC 20000 (92%neutrophils), platelet – 1.75 lakh
Renal function test	Blood urea- 80, Creatinine- 1.47, Sodium 136
Liver Function Test	Total Bilirubin- 0.59, AST – 33, ALT- 24, Albumin 2.3, Globulin – 2.3
Urine microscopy	Protein – 1+, RBC- not detected, Pus cell-occasional

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Figure 1

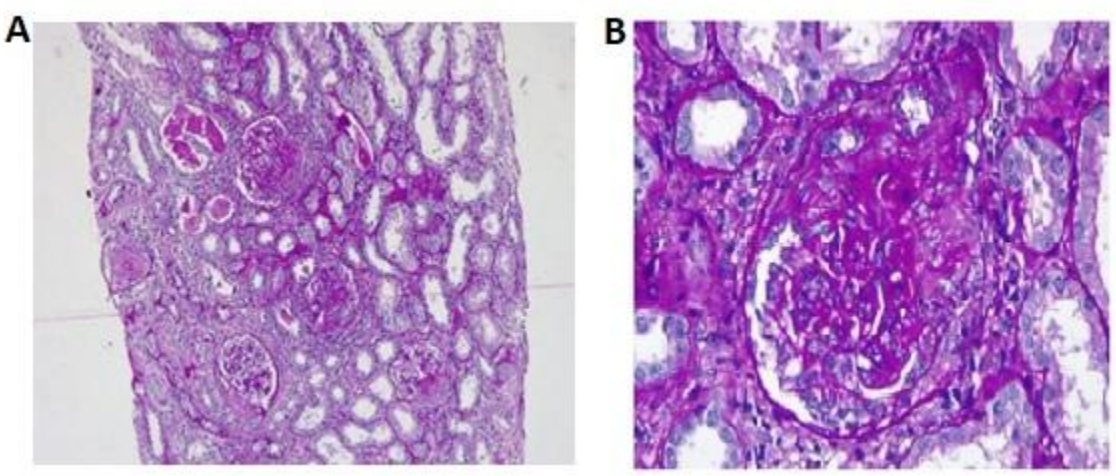

Figure 1 – (A)light microscopy showing 5 glomeruli with 1 showing complete sclerosis and 3 glomeruli showing proliferation and significant increase in mesangial matrix. Multifocal chronic inflammation is noted in the interstitium (B) magnified image showing segmental increase in mesangial matrix and a cellular crescent.

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