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Sjogren syndrome with hypokalemic quadriparesis diagnosed in pregnancy - a case report of the pregnancy outcome

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Abstract : Sjogren syndrome is one of the rarely reported autoimmune disorders in pregnancy. In this condition ,the individuals immune system mistakenly attacks its own moisture producing glands with a progressive degeneration of the exocrine glands. We report a case of a 29 yr old pregnant woman,G4P1L0A2, who presented with acute quadriparesis, distal renal tubular acidosis and severe hypokalemia in the twelfth week of her pregnancy. The patient was then evaluated and diagnosed with SJOGRENs SYNDROME with anti SS(RO) and antiSS(LA) POSITIVE and ANA POSITIVE. She was a case of bad obstetric history with 2 abortions and a neonatal loss due to fetal bradycardia attributed to congenital heart block. Patient was followed up regularly and has successfully delivered at term an alive boy baby weighing 2.450kg by LSCS. we report this case for its rarity and the good fetal outcome.

Keyword : Sjogren Syndrome, Congenital Heart Block

INTRODUCTION:

Primary SS(pSS) is an autoimmune disorder with a prevalence of 0.3-0.6% in general population [1], when defined strictly according to the American European Consensus Criteria(AECC)[2], and a female: male ratio of 9:1.pregnancy complications due to Sjogren's syndrome are neonatal lupus and congenital heart block[3]. SECONDARY SS, coexists with other auto immune disorders like rheumatoid arthritis and SLE[4]. 25% of these patients have general complications, one of which is renal involvement characterized by distal tubular acidosis and severe hypokalemia. Only 14 cases of distal tubular acidosis with hypokalemic paralysis have been reported so far, in association with Sjogren's syndrome[5,6].

CASE REPORT:

A 29 yr old G4P1L0A2 with 36 weeks of gestation was admitted in our hospital for safe confinement. She was a known case of Sjogren's syndrome diagnosed during the early weeks of the present pregnancy. She was married for 7 yrs , and degree consanguineous marriage and had conceived spontaneously.

Previous Obstetric history:

She had two 1st trimester spontaneous abortions and an early neonatal death due to ?congenital heart block in 2011.Patient was not evaluated then. The 3rd pregnancy was terminated by caesarean section at term for fetal bradycardia.

Present obstetric history:

She was booked and immunized in a private hospital. All 3 trimesters were uneventful. She was diagnosed in the third month of this pregnancy with Sjogren's syndrome on 13/6/2015, she presented to a private hospital with history of weakness and difficulty in using both upper and lower limbs for one day. She had swelling of both her legs and face for one day with vomiting and giddiness for one week, with neck pain for a day. She was a known case of hypothyroidism on tablet Eltroxin 75 microgram 1 od.

Course in the hospital:

She was admitted in the intensive care unit and her investigations showed

- Low potassium-1.9mg/dl
- Positive CRP-27mg/dl
- Bicarbonate -8mmol/l
- Urine ph-6.8
- d-dimer negative
- ECG–Right axis deviation, RV strain, st-t changes of severe hypokalemia

Spot urine studies showed -

- Spot cl-134 meq/l
- Spot potassium-25.50meq/l

She was diagnosed with hyperchloremic metabolic asidosis with type 1- RTA, and was treated with iv fluids, antibiotics and syrup potassium citrate. She was then evaluated further to rule out the cause of this hypokalemic quadriparesis with RTA. ANA SCREEN was done to rule out auto immune causes. She was found to be ANA-positive. SS-Apositive, RO-positive, SS-B-positive. All others were negative. She was started on tablet Prednisolone 5mg 1-0-0 and tablet Hydroxychloroquine 200 mg 0-0-1 since then.

Present pregnancy outcome:
She had regular checkups with the rheumatologist and the obstetricians. Her serum electrolytes was regularly monitored and fetal echo was done to rule out congenital AV block, which showed no abnormality. The pregnancy continued till term uneventfully and was terminated by lower segment caesarean section. She delivered an alive boy baby weighing 2.450kg.

- Neonatal Echo - Normal study.
- TFT of the baby –Normal.

So a successful pregnancy with good fetal outcome in a case of Sjogren's syndrome was obtained.

DISCUSSION:

SJOGREN'S SYNDROME: It is well known that the disease often starts in the fourth or fifth decade of life, thus most are postmenopausal. In a cross sectional analysis of the female patients with P(SS), only 13 % were less than 45 years[7]. Young onset P (SS) is usually associated with more systemic manifestations and high concentrations of autoantibodies. 59%-85%of them are noted to have an elevated ANA titer. ANA positivity is associated with higher prevalence of antiSS(A), antiSS(B) antibodies, antiphospholipid antibodies, rheumatoid factor and hypergammaglobulinemia. The presence of ANA in P(ss) is associated with a higher risk of cutaneous vasculitis, articular and renal involvement and therefore higher utilization of corticosteroids.

P(ss) WITH RENAL INVOLVEMENT:

DISTAL RENAL TUBULAR ACIDOSIS is a condition characterized by inability of the distal nephrons to acidify the urine. Immunohistochemical analysis of the tissue obtained by renal biopsy showed complete absence of the HTPase pump in the intercalated cells in the collecting tubules that is responsible for the distal portion secretion. Therefore the DTA is common , which is mild and in some cases, hypokalemic paralysis has been the only presenting dominant symptom, as in our case. Potassium citrate is the preferred therapy to correct the acidemia in distal RTA. Therefore in patients presenting as hypokalemic paralysis, a clinical suspicion can unmask a subclinical Sjogren's syndrome.

P(SS) AND PREGNANCY OUTCOME:

Pregnancy outcome in P(SS) has not been extensively studied but has been considered to be associated with unfavourable fetal outcome[8]. Only two studies have reported risk of spontaneous abortion and fetal loss in pregnancies in Sjogren's syndrome[9,10] (as in our case history) . However, both premature deliveries and spontaneous abortions were significantly higher when SLE was associated with SS. There is a high risk of neonatal lupus and congenital atrio-ventricular block associated with high perinatal morbidity and mortality. As in our case the two abortions of unknown cause and a neonatal death due to ?CHD could be possibly attributed to the undiagnosed P(SS). This pregnancy had a successful outcome after she received treatment for the same. Fetal outcome: Term boy , Birth weight: 2.450 kg. Screening for CHD: Echo -normal study. Therefore a meticulous fetal screening for CHD & IUGR is needed and maternal monitoring for the renal complications is mandatory with RFT and pH values in a case of Sjogren's syndrome in pregnancy. These efforts will give a successful pregnancy outcome.

CONCLUSION:

Although Obstetricians may encounter this disorder rarely, awareness of the varied forms of this disease is essential for the early diagnosis and a good fetal outcome. These patients should be referred at the earliest to a tertiary care centre, where a multidisciplinary approach along with a rheumatologist and a neonatologist is possible. Last but not the least, a good neonatal intensive care unit is necessary for the backup.

REFERENCES:

1. Bowman SJ, Ibrahim GH, Holmes G, Hamburger J, Ainsworth JR. Estimating the prevalence among Caucasian women of primary Sjögren's syndrome in two general practices in Birmingham, UK. Scand J Rheumatol 2004;33:39-43.

2. Vitali C, Bombardieri S, Jonsson R, et al., Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 2002;61:554-8.

3. Koopman W, Moreland LJonsson R, Bowman S, Gordon T., Sjögren's syndrome. In: KoopmanW, MorelandL, editors. Arthritis and allied conditions – A textbook of Rheumatology. 15th edition. Philadelphia: Lippincott Williams & Wilkins; 2005. p. 1681-705.

4. Theander E, Jacobsson LT., Relationship of Sjogren's syndrome to other connective tissue and autoimmune disorders. Rheum Dis Clin North Am 2008;34:935-47.

5. Siamopoulos KC, Mavridis AK, Elisaf M, Moutsopoulos HM, Kidney involvement in primary Sjögren's syndrome. Scand J Rheumatol Suppl. 1986;61:156–60.[PubMed: 3473626]

6. Chang YC, Huang CC, Chiou YY, Yu CY. Renal tubular acidosis complicated with hypokalemic periodic paralysis. Pediatr Neurol. 1995;13:52–4.[PubMed: 7575850]

7. Lee LA, Transient autoimmunity related to maternal autoantibodies: neonatal lupus. Autoimmun Rev 2005;4:207-13.

8. Mecacci F, Pieralli A, Bianchi B, Paidas MJ, The impact of autoimmune disorders and adverse pregnancy outcome. Semin Perinatol 2007;31:223-6.

9. Siamopoulou-Mavridou A, Manoussakis MN, Mavridis AK, Moutsopoulos HM, Outcome of pregnancy in patients with autoimmune rheumatic disease before the disease onset. Ann Rheum Dis 1988;47:982-7.

10. Julkunen H, Kaaja R, Kurki P, et al, Outcome in women with primary Sjogren's syndrome. A retrospective case-control study. Clin Exp Rheumatol 1995;13:65-71.