

Case Series

PREGNANCY WITH ADDISON DISEASE; A CASE SERIES

Saima Najam, Syeda Shehla Batool and Ali Raza Haider

Abstracts: Addison disease or primary adrenocortical insufficiency is rare and potentially lethal endocrine disorder, in pregnancy it is even more rarely seen. We report two cases of Patients with known addisons disease were taking treatment before the pregnancy. Both patients fortunately were of same age and conceived spontaneously but had different associated illnesses. Both were very compliant and co operative and they both delivered at around 37 weeks , and were managed in collaboration with the endocrinologist and dietitian. Both patients had uneventful recovery after normal delivery and cessarian section respectively. Therefore we conclude that Patient education, monitoring and titration of her steroids are the only ways to avoid complications associated with the under or over treatment with steroids and should be managed by the multidisciplinary team.

Keywords: addison disease, labor, prednisolone, hydrocortisone.

Introduction

Primary adrenocortical insufficiency is rare because more than 90% of total gland volume must be destroyed for symptoms to develop. It is a long term potentially severe chronic disease of the adrenal cortex associated with the insufficient production of the glucocorticoids and mineralocorticoids. Auto immune adrenalitis is the most common cause in the developed world while tuberculosis has been recognized as a frequent etiology in the developing world. There is increased incidence of concurrent Hashimoto thyroiditis, premature ovarian failure , type 1 Diabetes and graves disease.¹

As Addison disease is a very rare chronic condition , with prevalence of 4-11/100,000, So the precise prevalence of the disease in pregnancy is unknown. (2)One of the leading reports, which was done in Norway over 12 years, showed an Estimated incidence of 1/3000 births.⁴ Women with Addison disease show reduced parity, the reason being ,firstly they are reluctant to get pregnant because of risk of complications and other concomitant autoimmune diseases, secondly autoimmune thyroid disease can lead to reduce fertility and thirdly the loss of adrenal androgen can play a role in the fertility of this patient, although this is not clear.² Addison disease has deleterious effects on pregnancy outcome., it can lead to abortion , intrauterine growth restriction , intrauterine fetal death and postpartum adrenal crisis.³ Considering this together with the rarity of the condition a pregnant Addison patient is a very infrequent scenario in the clinical practice. We

report two cases with Addison disease who got pregnant immediately after marriage and her pregnancy, labor and delivery remained uneventful.

Case Report One

A 31years old house wife , married for two months, known case of Addison disease and Hypothyroidism for last ten years presented to emergency room (ER) eleven months ago with the complaint of fatigue , nausea and frequency of urine before 3 days of her expected period. Her electrolytes (sodium 136mmol/L, reference range 136-145mmol/L, potassium 4.00mmol/L, reference range 3.5-5.1 mmol/L) and thyroid stimulating hormone (TSH 1.52miu/L, reference value 0.27-4.2miu/L) were normal. Her bhcg was done and was found positive and urine culture showed no uropathogen growth after 72 hours. She was booked and managed in collaboration with the endocrinologist with the same dossed of the steroids which she was taking earlier. She was a very compliant patient and visited regularly, she was counselled on every visit about the medication , life style and precautions to be taken in case of stress, infection and surgery. At 7 weeks once she presented with dizziness was found to have hypotension her blood pressure dropped to 80/60 mm of Hg, she was hydrated with normal saline and given Injection Hydrocortisone 50 mg stat. Her blood sugars and electrolytes were normal at that time. Her rest of the pregnancy remained un eventful. She was co managed by the obstetrician and the endocrinologist. She was advised to take increase salt and her serum electrolytes were kept with in the normal range by increasing dose of glucocorticoids

.Before conception she was taking tab prednisolone 5.0mg at 7.00am and 2.5 mg at 3.00pm. With fludrocortisone 0.1mg OD, thyroxine 75MCG* OD. By term her prednisolone was increased to 10.0 mg at 7.00 AM and 2.5 mg at 3.00PM with the same dose of fludrocortisone and thyroxine. She presented at 37+6 weeks with spontaneous rupture of membranes(ROM) for 5 hours, and labor pains in the emergency room. Her amniure was positive which confirmed ROM, her bishop score was 6, and cardiotocography (CTG)was reactive so she was admitted for the induction of labor. She was induced with 2.0 mg prostaglandin E2 intravaginal gel after 6 hours of ROM .In active phase of labor she was started with Inj hyrocortisone 100mg intravenous 6 hourly till delivery of the baby and the placenta in collaboration of the endocrinologist.She was kept hydrated with normal saline throughout the labor and delivery.. Her labor progressed smoothly and after 5 hours of active phase she delivered a 2.4 kg , female baby with Apgar score of 6/10 and 8/10 as vertex, the length of her second and third stage of labor was 20 min and one minute respectively. Her post natal recovery was also Uneventful. After delivery her prednisolone was doubled for 48 hours and then tapered to the pre pregnancy dosage before discharge. She was discharged in stable condition on her 5th post natal day. One week follow up showed normal electrolytes and good healing of her episiotomy and successful breast feeding. The progress of the baby was also normal and her electrolytes were normal as well.

Case Report Two

A 31years old house wife , married for 7months , known case of diabetic from last 5 years, and known case of Addison disease from last 2 years , was booked at 7 weeks of pregnancy. She was taking insulin (regular)10 units three times a day and was on hydrocortisone tab 10 mg at 8.00pm and 5.0 mg at 4.00pm. She has been very compliant and did regular visits. She only needed increments in the dosage of insulin which was done in collaboration with the endocrinologist, the insulin was increased to 20 units twice daily of Glargine insulin and 20 units of Aspart three times a day at 29 weeks and was continued in the same dosage till delivery. Her hydrocortisone was continued in the same dosage and no increments were required. she was counselled on every visit about the medications , life style , diet ,exercise and precautions to be taken in case of stress, infection and surgery. She

presented at 37+1 weeks with irregular contractions in the emergency room. She was in latent phase of labor at the time of admission, after evaluation cessarian section was planned because of the good size of the baby. Injection hydrocortisone 100mg was given before induction of anesthesia then under spinal anesthesia her cessarian was done . A female baby with Apgar score 8/10 and 9/10 was delivered as cephalic and the weight of the baby was 4.7 kg. Her post op recovery was un eventful. The baby was shifted to NICU for the blood sugar monitoring.Injection hydrocortisone 100 mg intravenous three times a day was continued for 24 hours, then reduced to 50 mg intravenous three times a day for next 24 hours and then was given 50 mg intravenous two times a day for next 24 hours. After that tab hydrocortisone was started, 10.00mg was given at 6.00am and then 5.00 mg was given at 12.00 pm and 5 mg at 6.00pm was continued for 48 hours. The insulin was given acc to sliding scale for first 48 hours and then pre regnancy dosage resumed that is 10mg three times a day. She was discharged on her 6th post op day on hydrocortisone tab 10 mg at 8.00am and then 5.00mg at 4.00pm and the same dose of insulin in stable condition. On follow up visit after a week the patient showed uneventful recovery with the normal healing of her abdominal wound and the baby who was doing well on breast feeding along with bottle feeding. .

Discussion

Managing a patient with Addison disease in pregnancy and labor is a challenging clinical scenario. The replacement of the glucocorticoids and the mineralocorticoids should be continued throughout the pregnancy, delivery and lactation. The dose depends on clinical scenario and serum electrolytes. Two third of the total daily dose is given when the patient awakens and one third is given in the after noon as in the normal circadian rhythm of cortisol.⁴ The increment is usually needed in third trimester. Mineralocorticoid treatment is a concern as well. Ambrosi et al in their review has suggested that reduction in the dose of the Fudrocortisone is required if the patient has hypertension and hypokalemia.⁵ how ever as our patient did not develop the hypertension so we continued with the same dose 0.1 mgm of Fludrocortisone as she was using before pregnancy. Our patient delivered normally, During labor adequate hydration with continuous infusion of normal saline and glucocorticoids was required. As the second patient required cessarian section so she received 100mgm of hydrocortisone IV before

surgery begins , which was then continued every eight hours and then tapered to the previous oral dose in 48 hours.⁶

Both babies came out to be healthy and no teratogenic effect was noticed because of the Glucocorticoid and mineralocorticoid treatment

through out the pregnancy ,as observed by Lindsay et al.⁷

*Department of Obst. & Gynae
Dr. Sulaima Alhabib Hospital,
Sweidi, Riyadh, KSA
www.esculapio.pk*

References

1. Melmed S, Polonsky KS, Larsen PR and Kronenberg HM. Williams Endocrinology, 13TH edition, 2016. Section iv, Adrenal cortex and endocrine hypertension.
2. Oliveria D, Lagas A, Palva S and Carrilho F. Treatment of Addison's disease during pregnancy. Endocrinol Diabetes Metab Case Rep. 2018;2018:17-0179
3. Banu NA, Begum K, Islam NI. Addison disease in pregnancy- A case report. Bang Med J 2011; 40:48-50.
4. Albert E, Dalakar K, Jorder R, Berge LN. Addison's disease and pregnancy. Acta Obstet Gynecol Scand 1989;68:185-7.
5. Ambrossi B, Barbetta L, Morricone L. Diagnosis and management of Addison's disease during pregnancy. J Endocrinol Invest 2003;26:698-702.
6. Otta CF, Mereshian PS, Iraci GS, Ojeda MR. Pregnancies associated with primary adrenal Insufficiency. Fertility and sterility 2008;90:17-20.
7. Lindsay JR, Nieman LK. The hypothalamic pituitary adrenal axis in pregnancy: Challenges in disease and treatment. Endocr Rev 2005;361:1881-93.