CASE REPORT

Schmidt Syndrome in Pregnancy: A Hormonal Potpourri to a Happy Pregnancy

Vinu Vilashini[1], Rohini G[2], Tharaka Senathirajah[3]

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Abstract

Autoimmune polyglandular syndrome type 2 (APS-2) or Schmidt syndrome is defined as a coexistence of autoimmune thyroid disease, primary adrenal insufficiency, and/or type 1 diabetes mellitus. The presentation of APS-2 is rarely encountered during pregnancy since there is reduced fertility owing to the adrenal and thyroid insufficiency individually and can present with multiple complications. We present a case of a 25-year-old G2A1, who is a known case of Schmidt syndrome diagnosed preconceptionally when presented with nausea, vomiting, diffuse and progressive hyperpigmentation spots over palmar creases, soles with elevated ACTH and decreased level of cortisol, and resolution of symptoms when started on steroid replacement therapy. The patient was followed up regularly throughout the pregnancy and delivered by cesarean section and developed gestational hypertension, which was treated. We present this case to conclude that pregnancy with Addison's disease can be managed successfully with proper monitoring and multidisciplinary management approach.

Keywords: Addison's disease, Autoimmune polyglandular syndrome, Case report, Schmidt syndrome.

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Introduction

Autoimmune polyglandular syndrome type 2 (APS-2) or Schmidt syndrome is defined as a coexistence of autoimmune thyroid disease, primary adrenal insufficiency, and/or type-1 diabetes mellitus. The presentation of APS-2 is rarely encountered during pregnancy since there is reduced fertility owing to the adrenal and thyroid insufficiency individually and can present with multiple complications. Very few cases have been reported so far.[1] If the diagnosis of APS-2 is missed during pregnancy or postpartum period, it can cause a significant maternal and fetal morbidity.[2] With glucocorticoid therapy and improved obstetric care, there is an immense reduction in maternal mortality.[3]

Case Description

A 21-year-old female was diagnosed to be a case of autoimmune polyglandular syndrome type 2 when she presented with nausea, vomiting, diffuse and progressive hyperpigmentation spots over palmar creases and soles (Table 1).

The patient was started on steroid replacement therapy—T. fludrocortisone 100 mcg once daily (OD), T. prednisolone 5 mg OD, and T. thyronorm 50 mcg OD. The patient's condition improved and was monitored with serum ACTH, serum cortisol, serum TSH, and serum electrolytes regularly.

The patient got married 4 years later. The patient conceived spontaneously 3 months later but had a spontaneous abortion at 8 weeks, managed medically. The patient conceived spontaneously 6 months later and was admitted at 8 weeks + 4 days with excessive nausea and vomiting. The patient was found hypokalemic and corrected with 40 mEq of potassium chloride. Patient and attenders were counseled for stress management before discharge. Fludrocortisone was stopped in the first trimester, and the dosage of hydrocortisone was increased. Fludrocortisone was restarted in the second trimester. All trimesters were uneventful, serial electrolytes, RBS, blood pressure monitoring, serum ACTH, serum cortisol, and serum TSH was done and found to have within normal limits. Anomaly scan and growth scan were done—normal morphology and biometric measurements. Estimated fetal weight (EFW) – 50th centile (Fig. 1).

[1–3]Department of Obstetrics and Gynecology, Saveetha Medical College and Hospital, Chennai, Tamil Nadu, India

Corresponding Author: Vinu Vilashini, Department of Obstetrics and Gynecology, Saveetha Medical College and Hospital, Chennai, Tamil Nadu, India, Phone: +91 9894349344 e-mail: vinu.vilashini@gmail.com

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Table 1: Investigations done at first presentation

Investigation	*Values*
Sr. ACTH	1080 pg/mL
Sr. cortisol	< 0.5 µg/dL
TSH	24.6 mIU/mL
Anti-TPO	36.8 IU/mL
Sr. sodium	131 mEq/L
Sr. potassium	3.9 mEq/L
Sr. calcium, parathormone, vitamin D, vitamin B12, folic acid, fasting and postprandial blood sugar	Within normal limits

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Labor

The patient was induced with 1st dose of PGE2 gel at 38 weeks. At the time of induction, her modified Bishop's score was 4. Inj. hydrocortisone 100 mg IV 8th hourly was started. Intermittent CTG monitoring was done and found reactive. Hourly BP monitoring was done after 4 hours after PGE2 gel induction. The patient had an elevated blood pressure of 180/100 mm Hg. Urine albumin – 2+. No imminent symptoms of eclampsia. Blood pressure management was started with two doses of IV labetalol 20 mg IV. However, repeat blood pressure was persistently high. The patient underwent an emergency cesarean section in view of severe preeclampsia with an unfavorable cervix and delivered a male baby of 2.70 kg with a good Apgar. The intraoperative period was uneventful (Fig. 2).

Postpartum

The patient was administered with IV hydrocortisone 100 mg IV 8th hourly for 48 hours postoperative period and restarted on twice the usual dose for the next 48 hours and tapered to T. Fludrocortisone 100 µg OD and T. Hydrocortisone 10 mg OD (Fig. 3).

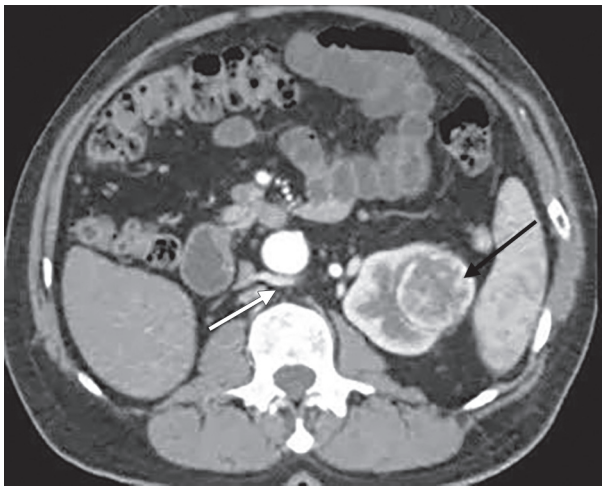

Fig. 1: CECT abdomen and pelvis – B/L hypoplastic adrenal glands

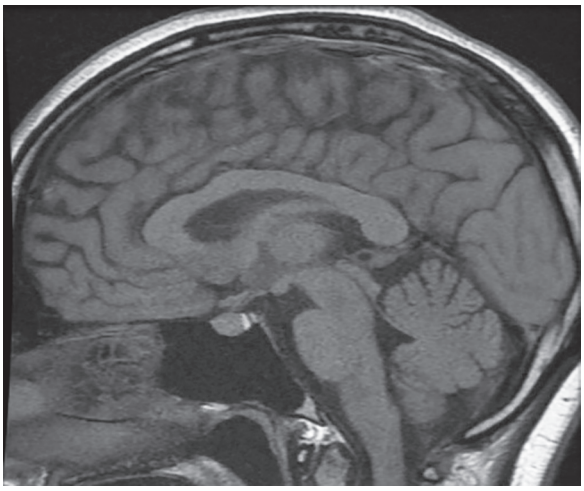

Fig. 2: MRI brain – normal study

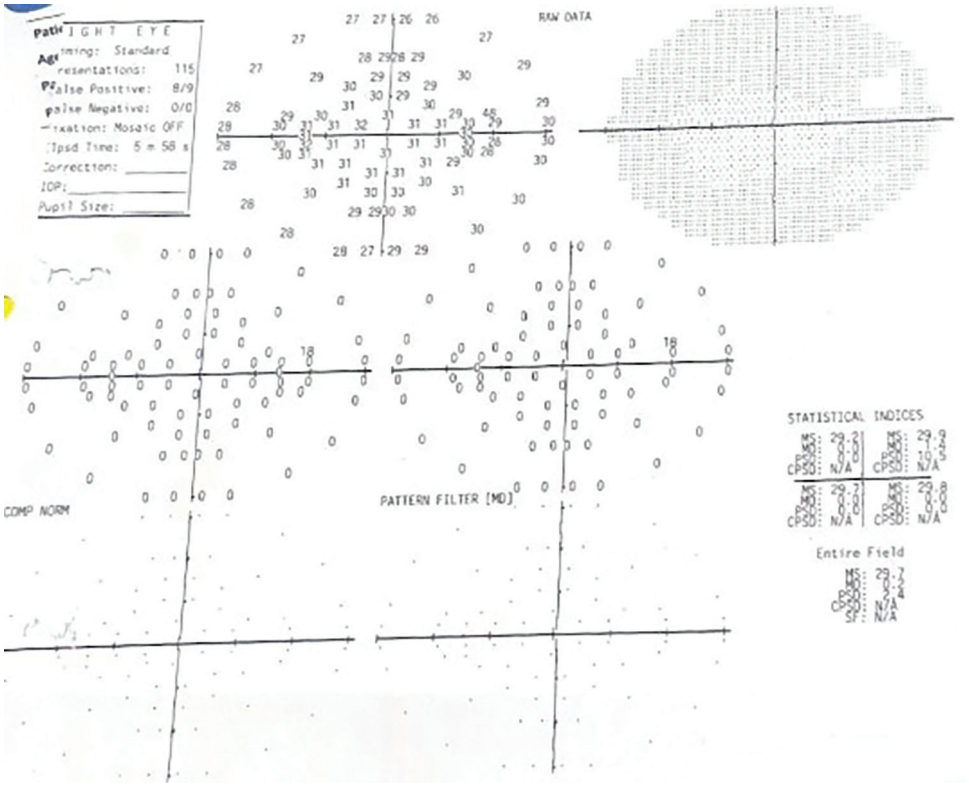

Fig. 3: Vision perimetry – normal study

The baby was evaluated, and TSH was within normal limits. The baby's external appearance was normal with good activity, bowel, and bladder movements. Blood pressure monitoring was done 2 hourly and was found adequately controlled with T. Labetalol 100 mg twice daily. Preeclampsia workup was within normal limits. The patient got discharged on the 10th postoperative day and was followed up on outpatient basis with home BP monitoring charts, and hypertension resolved by 4 weeks postpartum. The patient continued on T. Fludrocortisone 100 µg OD, T. Hydrocortisone 10 mg OD, and T. Thyronorm 50 µg OD and is on regular follow-up with an endocrinologist.

Discussion

The pathophysiology of autoimmune polyglandular syndrome involves a characteristic feature of lymphocytic infiltration into the affected glands, producing organ-specific autoantibodies and having an association with the HLA gene linkage.[4] Its diagnosis is often overlooked due to heterogeneous symptoms and slow progressive nature. The life-threatening acute adrenal crisis can be elicited by hyperemesis gravidarum, infections and delivery (both vaginal and cesarean), or failure to adequately adjust substitution therapy doses, which can be due to poor adherence, lack of education or insufficient medical monitoring, miscarriage in the first trimester, preterm birth or fetal growth restriction in the later trimesters and sometimes the development of preeclampsia too triggers the crisis.[5] An ideal glucocorticoid replacement dosage during pregnancy is not defined but is expected to be adjusted through the pregnancy, usually after the 24th week to mimic the normal physiological elevation of cortisol during pregnancy.[6] Hydrocortisone is preferred as it does not cross placenta. Fludrocortisone (category c) dose is uptitrated if there is hypotension or hyperkalemia even after increasing in hydrocortisone dosage. The delivery involves a multidisciplinary approach with IV glucocorticoid 100 mg bolus followed by 200–300 mg/24 hrs in active labor and is given twice the usual dose given during 48 hours postpartum and slowly tapered thereafter.[7]

Conclusion

The current evidence of management of APS-2 disease during pregnancy is not well-defined. The patients can present with a wide spectrum from nonspecific complaints to life threatening sequel of of DKA, vasomotor shock with Addison's, and myxedema coma with AITD if left untreated. Successful management involves a proper monitoring for recurrence of symptoms and adjusting the dosage of medication. The real challenge lies in monitoring the patients as they are more of clinical assessment.

Orcid

Vinu Vilashini https://orcid.org/0000-0001-6402-9935
Tharaka Senathirajah https://orcid.org/0000-0003-4561-3305

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