JGPEMN
ISSN 2091-279X
eISSN 2362-1168

CASE REPORT

Rare case of hypertension in pregnancy in rural setup - neurofibromatosis in pregnancy: a case report

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Date of submission: 10 Nov 2024

Date of acceptance: 16 Feb 2025

Date of Publication: 5 Mar 2025

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How to cite:
Limbu A. Rare case of hypertension in pregnancy in rural setup - neurofibromatosis in pregnancy: a case report. J Gen Pract Emerg Med Nepal. 2025 Jun;12(19):59-62.

Online information
DOI:
https://doi.org/10.59284/jgpeman309

Abstract

A 23-year-old woman, pregnant with her first child at 15 weeks, came in with severe headaches and dangerously high blood pressure (220/130 mmHg). Despite initial treatment with labetalol, her blood pressure remained unstable, even after adding methyldopa and nifedipine. During her exam, signs of Neurofibromatosis type 1 (NF1) were evident, including multiple neurofibromas, café-au-lait spots, and axillary freckling. Due to limited resources, tests for secondary causes of hypertension, like renovascular disease or pheochromocytoma, could not be performed. The patient chose not to be referred to a higher-level facility, citing financial concerns, and opted for voluntary discharge. Later, she delivered a premature baby at 34 weeks via cesarean section at a different hospital. While NF1 itself doesn't directly increase maternal mortality, it complicates the management of hypertension in pregnancy, contributing to a higher risk of adverse outcomes for both mother and baby. In rural areas with limited healthcare resources, these challenges are even more pronounced, and patient preferences can further complicate care. This case highlights the urgent need to improve access to healthcare, especially in rural settings, and to provide better training for healthcare providers managing complex pregnancies.

Keywords: Hypertension, Pregnancy, Neurofibromatosis

INTRODUCTION

Hypertension in pregnancy is a significant concern due to its potential complications for both mother and fetus. Identifying the underlying causes, particularly in young primigravida patients, is essential for effective management.[1]

Neurofibromatosis is a rare neurocutaneous disorder with a prevalence of 1 in 3000 for its type 1 variant and 1 in 25,000 for its type 2 variant.[1,2] The prevalence of neurofibromatosis type 1 (NF1) with hypertension during pregnancy is not explicitly quantified in the medical literature. However, several studies and guidelines provide insights into the association between NF1 and hypertensive disorders in pregnancy.

A population-based study by Terry et al. found that among 19 million pregnancy-related admissions, 1,553 were associated with NF1, indicating a prevalence of 0.008% for NF1 in pregnancy.[3]

This report describes a unique case of a pregnant woman with chronic hypertension and neurofibromatosis, aiming to educate readers about the diagnostic challenges and management options for future reference.

CASE REPORT

A 23-year-old female from Mahakali-8, Darchula, presented to the outpatient department for a general checkup at 15 weeks of gestation. She reported intermittent headaches lasting for a few hours. The headaches were dull, pressing type, particularly involving her forehead. It was non-radiating and not associated with any photophobia or phonophobia. It was not associated with nausea or vomiting, not aggravated by a change in position of the head, it was not associated with vision changes. The headaches disappeared on their own without intervention after a few hours. Her past medical history was not significant for any chronic medical illness, and she was not taking any long-term medications.

On Examination, she was alert and well-oriented to time, place, and person; there were no features of anemia, jaundice, lymphadenopathy, edema, cyanosis, or dehydration. Her Glass Coma Scale was 15/15, her Pulse was 75 beats per minute, her Respiratory rate was 14 per minute, her blood pressure measured on the right arm was 220/130 mmHg, and her blood pressure was recorded on the one hand. She had an oxygen saturation of 98% in the room air, and her blood glucose level at presentation was 130 mg/dl. Respiratory, Cardiovascular, Abdominal, and Central nervous systems revealed no gross abnormality.

The patient was immediately transferred to the emergency room, where she received IV Labetalol (10 mg stat) for hypertensive crisis.

Laboratory investigations were sent, and they showed a total count of 5 x 10^9 million/mm^3 with Neutrophils-50% and Lymphocytes 10%. A renal function test at the time revealed a sodium level of 142mmol/liter (135-145 mmol/l) and a potassium of 3.2mmol/liter (3.5-5.5 mmol/l); creatinine could not be measured due to unavailability. Her urine routine and microscopy findings did not reveal gross abnormality except for trace albumin. Her serology was non-reactive for HIV, Hepatitis B, and Venereal Disease Research Laboratory(VDRL) test was non-reactive. The liver function test was within normal limits. Electrocardiography examination was normal, with a sinus rhythm and a rate of 70 beats per minute. The retinal examination was not done due to the unavailability of an Ophthalmoscope at our center.

Chronic hypertension in pregnancy was suspected, and the patient was admitted for observation and monitoring.

During her hospitalization, the patient was prescribed:
1. Inj Labetalol 5 mg IV TDS
2. Tab Nifedipine 10 mg TDS

The following morning, her blood pressure measured 190/110 mmHg after completing the injectable dose of Labetalol.

While preparing for an ultrasound, it was noticed that the patient had multiple pedunculated masses ranging from 1x1 cm to 2x3 cm across her abdomen, chest, and back, which was initially missed during the OPD checkup. She also had numerous freckles around her armpit and groin area of a few millimeters in size. At least five hyperpigmented macules were present in her torso, and one smaller hyperpigmented macule a few millimeters in size in her lower back(Figure 1). She reported that these lesions had been present since childhood, and her sister had a similar condition, including facial lesions. Obstetric ultrasound revealed a Single live fetus with an unstable lie. It was 15 weeks of gestation in size, with a fetal heart rate of 134 beats per minute. Ultrasound of the abdomen did not reveal any significant findings.

A provisional diagnosis of Primigravida at 15 weeks of gestation with NF1 with chronic hypertension was made based on the characteristic findings of neurofibromas and café-au-lait spots. The patient was counseled about the complexities associated with NF1 and the potential implications for her pregnancy.

Despite recommendations for referral to a higher center, the patient expressed financial constraints and declined referral to a higher center. She also refused the option to terminate the pregnancy, stating it was her first.

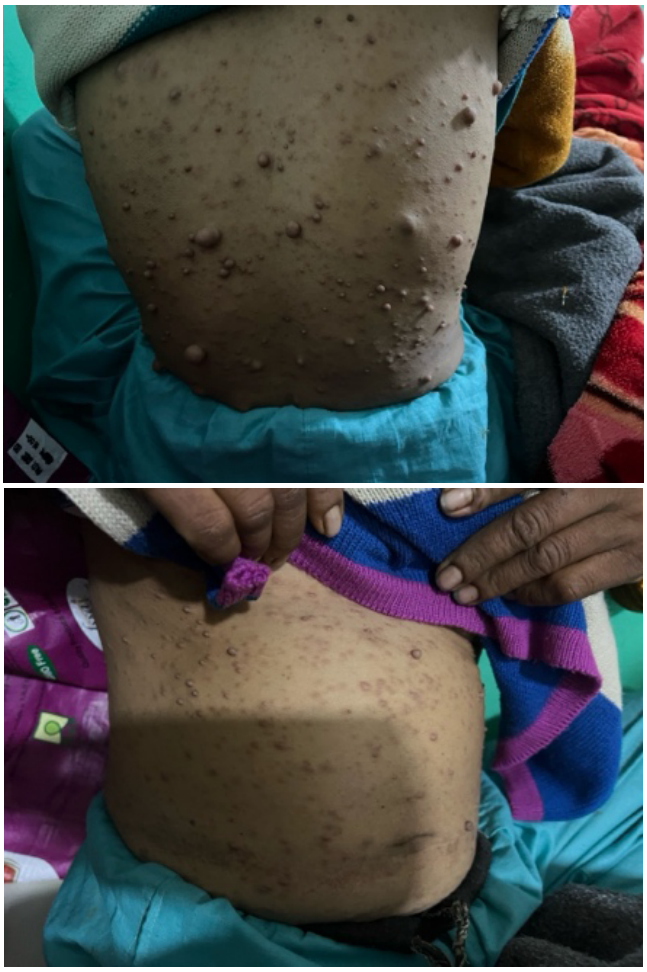

Figure 1. Numerous cutaneous lesions of varying sizes are observed on the back (above) and abdomen area (below). The lesions include dome-shaped nodules consistent with fibromatous growths. Additionally, hyperpigmented macules are scattered across the affected area.

To manage her hypertension, Methyldopa (250 mg TDS) was prescribed alongside Nifedipine (20 mg BD). On the third day, this regimen was titrated to Methyldopa (250 mg TDS) and Nifedipine (20 mg TDS). During her hospital stay, the lowest blood pressure measurement obtained was 160/80 mm Hg on the left arm and 170/90 mm Hg on the right arm, which was achieved within 24 hours of the hospital stay. However, a significant fluctuation ranged from 200-160 systolic and 120-60 diastolic blood pressure. The patient was required to procure medications herself when the stock within the pharmacy dried out. Unfortunately, she was only able to obtain Methyldopa before taking voluntary discharge from the hospital on the fourth day of admission. The patient delivered a premature male child at 34 Weeks of gestation via cesarean delivery at another center.

DISCUSSION

This case highlights the challenges of managing hypertension in pregnancy within a rural setting. Especially when underlying conditions such as NF1 are present. Patients' limited resources and financial constraints in these areas significantly affect the available management options. Blood pressure goals in pregnancy do not differ in a patient with Neurofibromatosis .[4] However, two of the most common causes of hypertension in NF1, namely, renovascular disease and pheochromocytoma, could not be properly evaluated in our setup. Though ultrasound was done, its sensitivity for diagnosing overall adrenal region lesions varies from 70-97%. The procedure is highly operator-dependent, and variability can be affected by the patient's body habitus, the size and functional status of the lesion, and the quality of the ultrasound machine.[5]

Testing for 24-hour urine metanephrine level or plasma metanephrine level could have ruled out pheochromocytoma, while other invasive tests would have been needed to rule out renovascular disease further. Furthermore, a diagnosis of pheochromocytoma requires prompt initiation of alpha blockade therapy such as phenoxybenzamine, primarily if the blood pressure cannot be controlled with a multidrug regimen with a plan for resection of the tumor in the near future.[6]

A study by L Dugoff, et al. presented data on pregnancy outcomes in 105 women with NF-1. It did not show an increased incidence of preeclampsia, preterm delivery, intrauterine growth restriction, pregnancy-induced hypertension, stillbirth, spontaneous abortion, or perinatal mortality.[7] Similarly, a retrospective cohort study by Terry et al. which included a nationwide sample from 1988-2009 and 1553 patients with NF1, concluded that although NF1 in pregnancy was associated with increased risk of maternal morbidity (preeclampsia, Intra-Uterine Growth Retardation (IUGR), preterm labor, cesarean section), increased risk of hypertension, and cerebrovascular complications, it was not associated with an increased risk of maternal mortality.[3]

It is safe to say that due to the associated risk to both fetus and mother, anticipation and planning of delivery require the involvement of obstetricians, radiologists, and pediatricians to minimize the risk and improve delivery outcomes.

The guidelines emphasize the importance of timely and effective management of hypertension in pregnancy, especially in the context of underlying medical conditions like NF1.[8] American College of Obstetricians and Gynecologists recommends the use of antihypertensive medications such as Methyldopa (3000 mg), Labetalol (2400 mg), and Nifedipine (120 mg) at maximum daily dosages for managing chronic hypertension during pregnancy.[9] The literature suggests targeting a blood pressure of 110–135/85 mmHg during pregnancy to reduce the risk of severe maternal hypertension while minimizing the risk of impaired fetal growth.[10] Regular blood pressure and fetal well-being monitoring and counseling regarding potential complications is crucial.

In the case of NF1, special attention must be paid to the risk of associated complications such as pheochromocytoma, which can present as secondary hypertension.[3] Comprehensive care ideally involves a multidisciplinary approach, including obstetricians, neurologists, and genetic counselors. Educating patients about their condition and its implications is essential to empower them to make informed decisions about their care.

CONCLUSION

This case underscores the importance of thorough clinical evaluation and individualized management of hypertensive disorders in pregnancy. In our case, unfortunately, due to the unavailability of medications for the management of hypertension in pregnancy, the patient opted to take a voluntary hospital discharge rather than get treated. The outcome, however, was favorable for both mother and child, as suggested by previously mentioned studies. This case serves as a reminder that there is a need for strategies to improve access to care and medication in these settings, along with enhanced training for healthcare providers on managing complex cases such as NF1 in pregnancy.

DECLARATIONS

Acknowledgement

I would like to thank Dr. Rajesh Awasthi (Medical Officer, District Hospital Darchula) for collecting the photographs and hospital staff for support during the management of this case.

Conflict of Interest

None

Funding

None

Consent of the Study

Informed consent was given by the patient for publication of this case report.

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