CASE REPORT OPEN ACCESS

A Case of Granulomatosis with Polyangiitis in Pregnancy

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ABSTRACT

Background: Granulomatosis with polyangiitis, a rare autoimmune disease affecting organs such as the respiratory system, kidneys, and lungs, poses unique challenges in pregnancy. A 25-year-old woman with chronic liver disease, vasculitis, protein C and S deficiency, hypothyroidism, and gestational diabetes presented at 24 weeks gestation with shortness of breath, fever, and vomiting. Her thrombocytopenia was managed with transfusions, and regular prenatal ultrasounds were conducted. At 36 weeks, an elective cesarean delivered a healthy 2.4 kg baby. This case highlights the critical role of multidisciplinary care in achieving favorable outcomes in complex pregnancies affected by granulomatosis with polyangiitis.

Keywords: Granulomatosis with polyangiitis, Wegener' Granulomatosis, Granulomatous vasculitis, ANCA-associated Vasculitis, Pregnancy

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INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis (WG), is a rare, chronic systemic disorder marked by granuloma formation and vasculitis[1,2]. As an autoimmune condition, GPA causes inflammation in blood vessels, predominantly targeting the upper respiratory tract, lungs, and kidneys[3]. GPA is the most prevalent type of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), with proteinase 3(PR3) antibodies present in 85–95% of cases[4].The antineutrophil cytoplasmic antibodies (ANCAs) are detected in most patients with active disease, the cause, however, remains unknown. Treatment involves immunosuppressive therapy, with cyclophosphamide (CYC) and corticosteroids[5]. The disease manifests in the fourth and fifth decades of life and is rarely linked to pregnancy. During pregnancy, it is a therapeutic challenge that calls for individualized care[6].

CASE PRESENTATION

A 25-year-old patient with G2P1+0 known case of NBNC chronic liver disease, ANCA +ve vasculitis (Wegener granulomatosis), protein C and S deficiency, hypothyroidism, and gestational diabetes mellitus presented on 24 weeks of gestation with complaint of SOB and fever for 1 month and vomiting for 3 to 4 days. Her labs showed thrombocytopenia for which she received 2 PCV transfusions.

During 1st, 2nd, and 3rd-trimester ultrasound scans

were done. She felt quickening in 5th month. On the 36th week of gestation, she presented to the ER with a complaint of leaking. LSCS was planned due to a previous C-section and leaking. After all aseptic measures, the abdomen was opened by Pfannenstiel incision, rectus sheath cut, muscles were separated, the peritoneum was opened and the lower segment of the uterus was opened by transverse incision. A live baby weighing 2.4 kg was delivered by vertex presentation placenta and membranes were expelled out completely the uterus was closed back in layers and homeostasis was secured. Bilateral tubal ligation (BTL) was also done. Operative findings: liquor was scanty, adhesions were present and there was blood loss of about 800 ml, 4 platelets were transfused during the C-section. Treatment was done via prednisolone (5mg), azathioprine (200mg), and hydroxychloroquine (200mg).

LAB FINDINGS

Hb	11.2g/dl	Protein S	66%
TLC	7.7microlitre	Protein C	46%
Platelets	96	P ANCA	Negative
Creatinine	0.7mg/dl	C ANCA	453
Cardiolipin IgM	0.45MPL U/ml	Cardiolipin IgG	0.07 GPL U /ml
Anti-thrombin	91%	Cardiolipin IgM	0.45 MPL U/ml
Creatinine	0.7mg/dl		

DISCUSSIONS

Wegener's granulomatosis is an autoimmune disorder characterized by granulomatous inflammation and necrotizing vasculitis, which can lead to significant morbidity and mortality if left untreated[7]. According to the available literature disease's presentation during pregnancy is particularly challenging, as the treatment options are limited due to the potential effects on the fetus and the associated risks for both the mother and the developing child. Wegener's granulomatosis that manifests during pregnancy may exhibit a more aggressive clinical course and may necessitate more intensive treatment compared to cases that arise at other times[8].

One of the earliest reported cases of Wegener's granulomatosis in pregnancy was published in 1983, where the disease appeared during the postpartum period and subsequently led to the interruption of a subsequent pregnancy[9]. The literature frequently highlights adverse pregnancy outcomes in women with active disease at the time of conception. The Vasculitis Clinical Research Consortium Patient Contact Registry found that pregnancy loss was more common in women who conceived after being diagnosed with systemic vasculitis, including AAV, compared to those who conceived before diagnosis (33.8% vs. 22.4%). Increased disease activity during pregnancy was also linked to a higher likelihood of preterm delivery. Additionally, a recent French study documented 19 pregnancies in 11 women with AAV, reporting elevated rates of complications[10].However, there is geographic and ethnic variation in GPA epidemiology. Among pregnancies complicated by GPA, approximately 71% resulted in live births. However, adverse outcomes were noted, including fetal loss in 11.5% of cases and premature births in 17.2% of pregnancies[11]. The management of GPA during pregnancy often involves immunosuppressive treatments such as cyclophosphamide and corticosteroids, which can complicate maternal health. One case highlighted a woman who experienced a flare of GPA at 34-35 weeks but ultimately delivered a healthy baby.6 Although there are no specific preventive measures, understanding risk factors can be crucial. GPA is more common in individuals of northern European descent and typically affects adults aged 40-65. Genetic predispositions and environmental factors may also play a role in the development of the disease[12].

CONCLUSION

Managing pregnancy in GPA patients demands preconception planning, precise clinical assessment, and aggressive treatment of active disease. Ideally, conception should be delayed for at least six months after achieving remission. A multidisciplinary team is essential for effective diagnosis and treatment.

DECLARATION

Not applicable

CONFLICT OF INTEREST

The author(s) declared no potential conflicts of interest concerning the research, authorship, and/or

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ETHICAL APPROVAL

The study was approved by the IRB, Liaquat College of Medicine & Dentistry.

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AUTHORS CONTRIBUTION

All authors equally contributed.

REFERENCES

1. Kubaisi B, Abu Samra K, Foster CS. Granulomatosis with polyangiitis (Wegener's disease): An updated review of ocular disease manifestations. Intractable Rare Dis Res. 2016 May;5(2):61-9. doi: 10.5582/irdr.2016.01014.
2. Garlapati P, Rout P, Qurie A. Granulomatosis with Polyangiitis. 2024 Aug 31. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 32491759. Available from: https://www.ncbi.nlm.nih.gov/books/NBK557827/
3. Tee QX, Wong A, Nambiar M, Lau KK. Granulomatosis with polyangiitis: Common and uncommon presentations. J Med Imaging Radiat Oncol. 2022 Dec;66(8):1089-1096. doi: 10.1111/1754-9485.13471.
4. Banerjee P, Jain A, Kumar U, Senapati S. Epidemiology and genetics of granulomatosis with polyangiitis. Rheumatology International. 2021 Dec;41(12):2069-89.
5. Tuin J, Sanders JS, de Joode AA, Stegeman CA. Pregnancy in women diagnosed with antineutrophil cytoplasmic antibody-associated vasculitis: outcome for the mother and the child. Arthritis Care Res. 2012;64(4):539–545. doi: 10.1002/acr.21556.
6. Verma A, Rajbhar S, Thakur P, Agrawal S, Pradhan S. Favorable Pregnancy Outcome in a Granulomatosis With Polyangiitis Patient With Renal Insufficiency. J Med Cases. 2021 Jan;12(1):27-31. doi: 10.14740/jmc3610.
7. Irfan O, Khan H, Khan Z, Ashraf A, Ahmed R, Khan JA, et al. Granulomatosis with polyangiitis: a 17 year experience from a tertiary care hospital in Pakistan. BMC Res Notes. 2018 May 16;11(1):303. doi: 10.1186/s13104-018-3434-2.
8. Devakumar VN, Castelino M, Chow SC, Teh LS. Wegener's granulomatosis in pregnancy: a case report and review of the medical literature. BMJ Case Reports. 2010 Jan 1;2010:bcr0920092296. doi: 10.1136/bcr.09.2009.2296.
9. Koukoura O, Mantas N, Linardakis H, Hajiioannou J, Sifakis S. Successful term pregnancy in a patient with Wegener's granulomatosis: case report and literature review. Fertil Steril. 2008 Feb;89(2):457.e1-5. doi: 10.1016/j.fertnstert.2007.03.054.
10. Croft AP, Smith SW, Carr S, Youssouf S, Salama AD, Burns A, et al. Successful outcome of pregnancy in patients with anti-neutrophil cytoplasm antibody-associated small vessel vasculitis. Kidney Int. 2015 Apr;87(4):807-11. doi: 10.1038/ki.2014.329.
11. Daher A, Sauvetre G, Girszyn N, Verspyck E, Levesque H, Le Besnerais M. Granulomatosis with polyangiitis and pregnancy: A case report and review of the literature. Obstet Med. 2020 Jun;13(2):76-82. doi: 10.1177/1753495X18822581.
12. Zhao WM, Wang ZJ, Shi R, Zhu YY, Zhang S, Wang RF, et al. Environmental factors influencing the risk of ANCA-associated vasculitis. Front Immunol. 2022 Sep 2;13:991256. doi: 10.3389/fimmu.2022.991256.