

From presumed infection to systemic vasculitis: de novo granulomatosis with polyangiitis in pregnancy – a case report

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ARTICLE INFO

Keywords:

Granulomatosis with polyangiitis
Pregnancy
ANCA-associated vasculitis
Refractory sinusitis
Case report

ABSTRACT

Granulomatosis with polyangiitis (GPA) is a rare ANCA-associated vasculitis. De novo presentation during pregnancy is exceptionally rare and may mimic severe infection, delaying diagnosis. This report concerns a case of new-onset GPA in a 26-year-old primigravid woman presenting in the second trimester with progressive refractory sinonasal disease initially treated as severe bacterial sinusitis. Despite repeated antimicrobial therapy and multiple ENT interventions, her condition deteriorated with development of hemoptysis and cavitating pulmonary lesions. Histopathology demonstrated necrotizing granulomatous inflammation, and further evaluation revealed ANCA positivity with anti-PR3 antibodies, confirming GPA at approximately 22 weeks of gestation. Management was complicated by rapidly progressive multisystem disease, concern for evolving renal involvement, opportunistic pulmonary infection with *Pneumocystis jirovecii*, and the limited availability of pregnancy-compatible immunosuppressive therapy before fetal viability. Disease control was achieved with corticosteroids, intravenous immunoglobulin, azathioprine, and multidisciplinary reassessment, while rituximab was deferred until the postpartum period. Pregnancy was continued to 35 + 5 weeks of gestation, resulting in delivery of a live female neonate appropriate for gestational age. At 2-year follow-up, maternal remission was sustained and the child remained clinically well. This case highlights the diagnostic challenges of de novo GPA during pregnancy and demonstrates that favorable maternal and neonatal outcomes remain possible with timely multidisciplinary management.

1. Introduction

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis, is a rare antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis characterized by necrotizing granulomatous inflammation and small-vessel vasculitis, most commonly affecting the upper and lower respiratory tracts and the kidneys [1,2]. In Europe, the annual incidence is estimated at approximately 3–14 cases per million population [2,3]. GPA most often occurs in middle age, with peak incidence in the fifth and sixth decades of life [2,4].

Presentation of GPA for the first time during pregnancy is exceptionally rare. In a recent review, only 19 women were identified in whom GPA first developed during pregnancy [5]. Diagnosis may be delayed due to the rarity of the condition and its initial presentation resembling common infectious or obstetric conditions. This report describes a case of new-onset GPA in a 26-year-old primigravid woman presenting in the second trimester, requiring early recognition through

repeated reassessment and close multidisciplinary coordination.

2. Case Presentation

A 26-year-old previously healthy primigravid woman was hospitalized at 19 weeks of gestation for progressive refractory sinonasal symptoms initially treated as severe bacterial sinusitis. Symptoms began at 9–10 weeks of gestation with progressive rhinosinusitis, facial pain, nasal discharge, and headache. Despite repeated antibiotic therapy, her condition progressively worsened. Otorhinolaryngology evaluation initially suggested chronic ethmoidal sinusitis, and bilateral maxillary sinus punctures yielded *Escherichia coli* on culture.

Because of persistent symptoms and markedly elevated inflammatory markers (CRP 134 mg/L), she was admitted to the pregnancy pathology unit with presumed severe pansinusitis. Despite broad-spectrum antimicrobial therapy, her condition failed to improve, and she underwent endoscopic bilateral ethmoidectomy with conchoplasty, followed

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<https://doi.org/10.1016/j.crwh.2026.e00829>

Received 13 May 2026; Received in revised form 8 June 2026; Accepted 16 June 2026

Available online 17 June 2026

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by revision sinus surgery with bilateral tympanostomy due to ongoing clinical deterioration and worsening hearing. Histopathological examination of sinonasal tissue demonstrated granulomatous necrotizing sinusitis, raising suspicion of systemic vasculitis. During hospitalization, she also developed dry cough and hemoptysis. Chest imaging revealed multiple bilateral pulmonary nodules, some with cavitation, despite ongoing antimicrobial treatment (Fig. 1A).

Further immunological evaluation showed ANCA positivity with anti-PR3 antibodies. Following multidisciplinary assessment involving an obstetrician-gynecologist, otorhinolaryngologist, pulmonologist, rheumatologist, nephrologist, infectious disease specialist, allergist and clinical immunologist, and clinical pharmacologist, the diagnosis of new-onset granulomatosis with polyangiitis (GPA) was established at approximately 22 weeks of gestation, based on the combination of refractory necrotizing granulomatous sinonasal disease, ANCA/PR3 positivity, and cavitating pulmonary lesions.

Given the severity of maternal disease, the uncertain short-term prognosis, and the possibility that pregnancy-compatible treatment might prove insufficient, the multidisciplinary team discussed the option of pregnancy termination before 22 weeks of gestation. Ultimately, as maternal status remained temporarily manageable and a pregnancy-compatible therapeutic strategy was considered feasible, the pregnancy was continued.

Initial treatment consisted of intravenous methylprednisolone followed by oral prednisolone. However, her course was complicated by worsening pulmonary disease (Fig. 1B-C) with concern for evolving renal involvement due to microscopic hematuria. Bronchoscopy and bronchial biopsy were performed, and bronchoalveolar lavage revealed *Pneumocystis jirovecii* DNA. Consequently, treatment with trimethoprim-sulfamethoxazole was initiated for *Pneumocystis jirovecii* pneumonia (PJP). Bronchial histopathology subsequently demonstrated necrosis with multinucleated giant cells and granulomatous inflammation, further supporting the diagnosis of GPA. Because management was complicated by the coexistence of active vasculitis, ongoing pregnancy, and opportunistic pulmonary infection, treatment required repeated multidisciplinary reassessment. After infectious control was achieved, she received intravenous immunoglobulin (IVIG) and additional high-dose corticosteroids, followed by azathioprine as a pregnancy-compatible steroid-sparing agent. Her course was additionally complicated by acute hypoxemic respiratory failure and severe anemia, which was managed with intravenous iron supplementation. Nevertheless, she gradually improved clinically and radiologically (Fig. 2), with a marked decline in inflammatory markers, and was discharged home on ongoing immunosuppressive therapy and antimicrobial prophylaxis.

The pregnancy was subsequently managed in a high-risk obstetric setting. At 35 + 5 weeks of gestation, she was rehospitalized with oligohydramnios and later developed preterm rupture of membranes, meconium-stained amniotic fluid, and concern for fetal wellbeing,

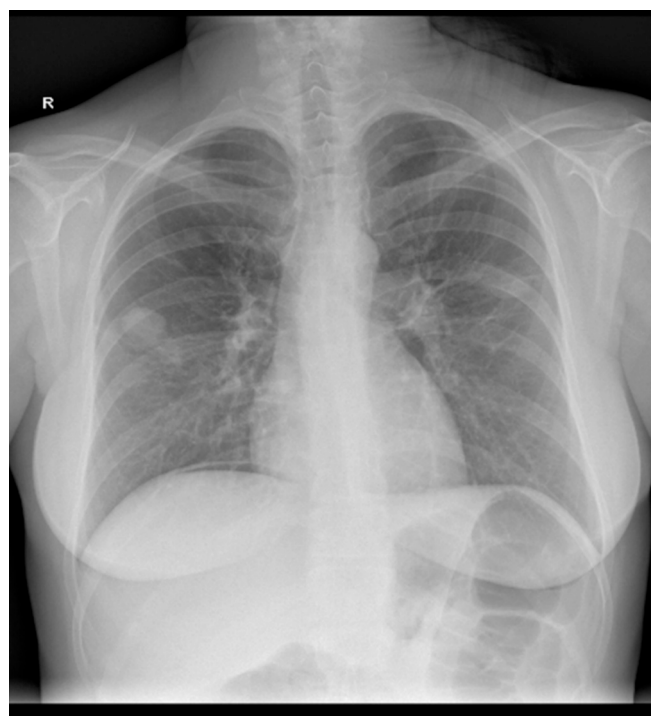


Fig. 2. Follow-up chest imaging demonstrating marked resolution of pulmonary lesions after treatment.

prompting emergency caesarean section. A live female neonate weighing 2350 g (appropriate for gestational age; approximately 13th percentile) was delivered, with Apgar scores of 9 and 9 at 1 and 5 min, respectively.

In the early postpartum period, persistent disease activity prompted escalation to rituximab. At 2-year follow-up, the patient had achieved sustained clinical remission under maintenance therapy with rituximab, low-dose prednisolone, and trimethoprim/sulfamethoxazole prophylaxis. Follow-up chest radiography showed complete resolution of pulmonary lesions and normal inflammatory markers, renal function remained preserved, and urinalysis showed no hematuria or proteinuria. Residual morbidity was limited to persistent hearing impairment and chronic sinonasal colonization without evidence of invasive infection. At 2-year follow-up, the child remained healthy, with no clinically apparent impairment in growth or neurodevelopment. Table 1 presents a timeline of the clinical events.

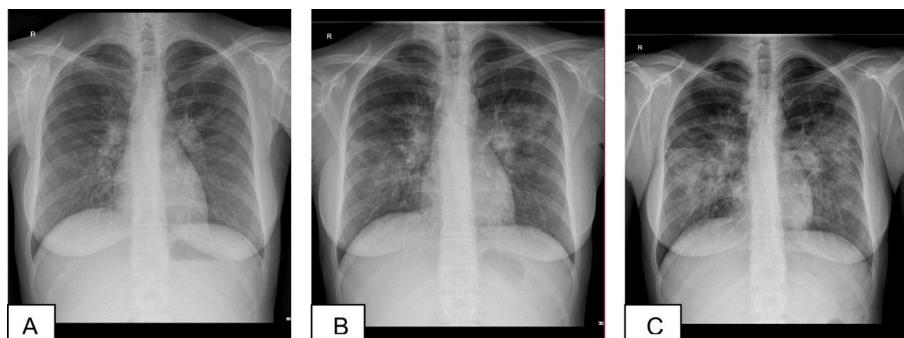


Fig. 1. Serial chest imaging demonstrating progressive pulmonary involvement during hospitalization. (A) April 4: bilateral pulmonary nodules. (B) April 9: progression in the size and number of lesions. (C) April 17: further progression with cavitory transformation.

Table 1
Timeline of clinical presentation, diagnosis, treatment, and follow-up.

Gestational age	Clinical event
9–10 weeks	Onset of progressive rhinosinusitis symptoms
19 weeks	Hospitalization for presumed severe pansinusitis
19–21 weeks	Progressive pulmonary disease and repeated ENT surgery
~22 weeks	Diagnosis of GPA confirmed (ANCA/PR3 positive) and corticosteroid therapy initiated
During hospitalization	<i>Pneumocystis jirovecii</i> pneumonia diagnosed and treated
Second and third trimester	IVIG and azathioprine initiated
35 + 5 weeks	Emergency caesarean section
Postpartum	Rituximab initiated because of persistent disease activity
2-year follow-up	Sustained maternal remission; child clinically well

3. Discussion

ANCA-associated vasculitis (AAV) includes three major subtypes: granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis (MPA). Among these subtypes, GPA is considered the most common [11]. However, ANCA-associated vasculitis in pregnancy is rare and typically represents pre-existing disease, whereas de novo presentation is exceptionally uncommon [5–7].

In 2019, Daher et al. reviewed cases of GPA in pregnancy published from 1970 to 2017. Among 87 pregnancies in 72 women, only 19 patients presented with GPA for the first time during pregnancy, and almost all cases involved respiratory tract manifestations like those observed in the patient presented here [5]. To the authors' knowledge, only approximately 20 cases of de novo GPA during pregnancy have been published to date. Clinical experience with GPA during pregnancy remains limited, particularly in women presenting with de novo disease, who often pose greater diagnostic and therapeutic challenges than those conceiving in remission. [5–7].

Available literature suggests that pregnancies complicated by AAV are associated with higher rates of maternal disease activity, preterm delivery, and fetal loss, although outcomes appear better when diagnosis is made promptly and management is coordinated across specialties [6,7]. In the review by Daher et al., 79% of pregnancies complicated by new-onset GPA resulted in live birth [5]. As in the present case, this suggests that favorable maternal and fetal outcomes are possible with timely diagnosis and appropriate treatment.

One of the greatest diagnostic challenges is that the majority of pregnant women with de novo GPA present with respiratory tract symptoms that mimic infection. In the present patient, the presence of *Escherichia coli* in sinus aspirate cultures raised additional concern because of its rarity in sinusitis pathogenesis [12]. However, the diagnosis became clearer only after progressive deterioration despite antimicrobial treatment, destructive sinonasal disease on pathology, and the subsequent appearance of cavitating pulmonary lesions. A previously published case of de novo GPA at 21 weeks of gestation also described an ENT-dominant presentation with bilateral sudden hearing loss and erosive rhinitis [5], which closely mirrors the diagnostic difficulty seen here. GPA should remain in the differential diagnosis of severe or refractory sinonasal disease during pregnancy, particularly when pulmonary abnormalities emerge.

This case also highlights a therapeutic dilemma that is not easy to capture in guidelines alone. Outside pregnancy, organ-threatening AAV is generally treated with high-dose glucocorticoids in combination with rituximab or cyclophosphamide [8]. During pregnancy, the range of acceptable options is narrower, and treatment decisions must balance maternal disease severity against fetal exposure. Current British Society for Rheumatology guidance supports the use of prednisolone and considers azathioprine compatible with pregnancy and breastfeeding, whereas cyclophosphamide is generally avoided during pregnancy whenever safer alternatives are available [9]. Rituximab can be

considered in selected circumstances, but its use later in gestation raises concerns about fetal and neonatal B-cell depletion and therefore usually requires individualized risk–benefit assessment [9]. In the present patient, one of the most challenging decision points occurred at approximately 21 weeks of gestation, when her clinical condition was rapidly deteriorating; however, this had to be balanced against her strong desire to continue the pregnancy and the documented risk of impaired fertility associated with cyclophosphamide exposure in women with GPA [13]. In practice, disease control was initially achieved using a pregnancy-compatible bridging strategy that included high-dose corticosteroids, intravenous immunoglobulin (IVIG), and azathioprine, while rituximab was deferred until the postpartum period once persistent disease activity became evident.

The coexistence of active vasculitis and suspected opportunistic pulmonary infection added another layer of complexity. Infection remains a major cause of early morbidity and mortality in AAV, particularly during periods of intensive immunosuppression [10]. In patients receiving rituximab, cyclophosphamide, and/or prolonged high-dose glucocorticoids, contemporary recommendations support consideration of trimethoprim–sulfamethoxazole prophylaxis because of the risk of *Pneumocystis jirovecii* pneumonia [8,10]. In this case, pulmonary deterioration could not be assumed to represent vasculitic progression alone, and the detection of *P. jirovecii* DNA required treatment of a competing diagnosis before immunosuppression could be escalated further. This sequence is clinically important, because in pregnancy the consequences of either under-treating active vasculitis or overlooking infection may be substantial for both mother and fetus.

This case demonstrates that favorable maternal and neonatal outcomes remain possible even when de novo GPA presents before fetal viability and treatment options are constrained by pregnancy. Despite severe multisystem disease, pregnancy was continued into the late preterm period, maternal remission was achieved, and both mother and child remained clinically well at follow-up.

Contributors

Gerda Deveikienė contributed to patient care, conception of the case report, acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Jelena Volochovič contributed to patient care, interpreting the data and revising the article critically for important intellectual content.

Gintautas Domža contributed to interpreting the data and revising the article critically for important intellectual content.

All authors approved the final submitted manuscript.

Declaration of generative AI and AI assisted technologies in the writing process

During the preparation of this work the authors used ChatGPT (OpenAI) solely for English language editing and stylistic refinement. The authors reviewed and edited the output as needed and take full responsibility for the content of the publication.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Funding

This work received no specific grant from any funding agency in the

public, commercial, or not-for-profit sectors.

Declaration of competing interest

The authors declare that they have no competing interest regarding the publication of this case report.

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