Scientific Research Publishing

Open Journal of Rheumatology and Autoimmune Diseases, 2025, 15(4), 133-138
https://www.scirp.org/journal/ojra
ISSN Online: 2164-005X
ISSN Print: 2163-9914

Bilateral Peripheral Ulcerative Keratitis Revealing Granulomatosis with Polyangiitis: A Case Report from a Hospital in Ouagadougou

Kiswendsida Abdoul Aziz Zorome[1*], Hervé Tieno[1,2], René Bognounou[1,2], Lassane Zoungrana[2,3], Rosalie Kabore[1], Seydou Yameogo[1], Michel Bouda[1]

[1]Department of Internal Medicine, Endocrinology and Metabolic Diseases of the Bogodogo Teaching Hospital, Ouagadougou, Burkina Faso
[2]Health Sciences Training and Research Unit, Joseph Ki-ZERBO University, Ouagadougou, Burkina Faso
[3]Department of Internal Medicine of the Yalgado Ouedraogo Teaching Hospital, Ouagadougou, Burkina Faso
Email: *zoromeaziz115@gmail.com

How to cite this paper: Zorome, K.A.A., Tieno, H., Bognounou, R., Zoungrana, L., Kabore, R., Yameogo, S. and Bouda, M. (2025) Bilateral Peripheral Ulcerative Keratitis Revealing Granulomatosis with Polyangiitis: A Case Report from a Hospital in Ouagadougou. *Open Journal of Rheumatology and Autoimmune Diseases*, **15**, 133-138.
https://doi.org/10.4236/ojra.2025.154016

Received: August 5, 2025
Accepted: September 20, 2025
Published: September 23, 2025

Abstract

Introduction: Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a necrotising vasculitis affecting small and medium-sized vessels. It is a rare condition. **Observation:** This is a 40-year-old patient with a history of recurrent inflammatory episodes in both eyes of unknown cause, which have been progressing for a year. He consulted the ophthalmology department for eye pain, tearing and decreased visual acuity in both eyes, which had been progressing for two weeks. The initial ophthalmological examination revealed visual acuity of 1/50 in the left eye and "positive light perception" in the right eye. The diagnosis of bilateral peripheral ulcerative keratitis complicated by corneal perforation in the right eye was made. The patient was admitted to the ophthalmology department and an evisceration of the right eye was performed. In addition, an internal medicine consultation was requested to determine the cause. Paraclinical tests revealed a biological inflammatory syndrome with a C-reactive protein level of 20.15 mg/l; immunological tests were positive for ANCA with an anti-PR3 specificity of 182 AU/l (Antibody Unit/litre). The rest of the additional tests showed no abnormalities. Induction treatment was initiated with oral corticosteroid therapy (prednisolone 1 mg/kg/day) in combination with an immunosuppressant, namely methotrexate at a dosage of 0.3 mg/kg per week. The prognosis is favourable, with improved vision in the left eye and regression of the biological inflammatory syndrome. **Conclusion:** GPA is a serious condition that can affect visual prognosis in its purely ocular form. This atypical presentation should be consistently reported to raise awareness among clinicians about the early diagnosis of GPA.

Keywords

Ulcerative Keratitis, Granulomatosis with Polyangiitis, Ouagadougou

1. Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a necrotizing vasculitis affecting small and medium-sized vessels. It is part of a spectrum of conditions known as antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis [1]. It is a rare condition with an estimated global annual incidence of 10 to 20 cases per million, depending on geographical location [2].

Clinically, GPA is characterized in its complete form by otorhinolaryngological signs, pulmonary and renal involvement [3]. Ocular involvement is also common in GPA, with various manifestations depending on the different ocular tissues affected, but it mainly causes orbital mass, dacryocystitis, scleritis, conjunctivitis, and keratitis [4].

GPA remains a real diagnostic challenge, sometimes presenting with atypical symptoms such as eye involvement (scleritis) or systemic infection (candidemia) [5] [6]. A limited form may also occur, although the classic form of GPA involving the kidneys and respiratory tract is more commonly observed [7].

To illustrate these clinical features, we report an unusual case of GPA manifesting solely as bilateral ocular involvement at the Bogodogo Teaching Hospital in Ouagadougou, Burkina Faso.

2. Observation

A concise timeline of key events to clarify the chronology is as follows:

- **Week 0:** A 40-year-old patient with a history of recurrent inflammatory episodes in both eyes, which had been developing for a year, presented with a flare-up of ocular symptoms consisting of eye pain, tearing, and decreased visual acuity in both eyes.
- **Week 2**: He consulted the ophthalmology department at Bogodogo Teaching Hospital due to persistent symptoms.
- **Day 0 (Week 2):** The initial ophthalmological examination revealed a visual acuity of 1/50 in the left eye and "positive light perception" in the right eye.
- **Day 4 (Week 2):** The patient was admitted to the ophthalmology department, and an evisceration of the right eye was performed.
- **Week 4:** The patient was seen in an internal medicine consultation with the results of the paraclinical assessment.
- **Week 6**: The patient is seen for a follow-up appointment after starting etiological treatment.

On day 0, in the right eye, the examination reveals eyelid oedema, diffuse conjunctival hyperaemia with chemosis, a perforated cornea in the upper part with

iris herniation, and the retrocorneal structures are not visible. The examination revealed pseudoptosis in the left eye, diffuse conjunctival hyperaemia, three corneal ulcerations, one located in the superotemporal region and two punctiform ulcerations located in the nasal and superior regions, respectively (**Figure 1**), and an iris hernia (**Figure 2**). The diagnosis of bilateral peripheral ulcerative keratitis complicated by corneal perforation in the right eye was made. The postoperative course was uneventful. In addition, an internal medicine consultation was requested during hospitalization for the etiological assessment. Upon re-evaluation, the extraocular clinical examination revealed no specific abnormalities. The diagnostic hypotheses considered were infectious causes (syphilis, viral infections: HCV, HIV); systemic causes (rheumatoid arthritis, relapsing polychondritis, systemic vasculitis). Paraclinical tests were prescribed, and the patient was discharged on day 6 pending the results of the etiological tests. Treatment with moxifloxacin eye drops was prescribed for the left eye (1 drop 4 times per day).

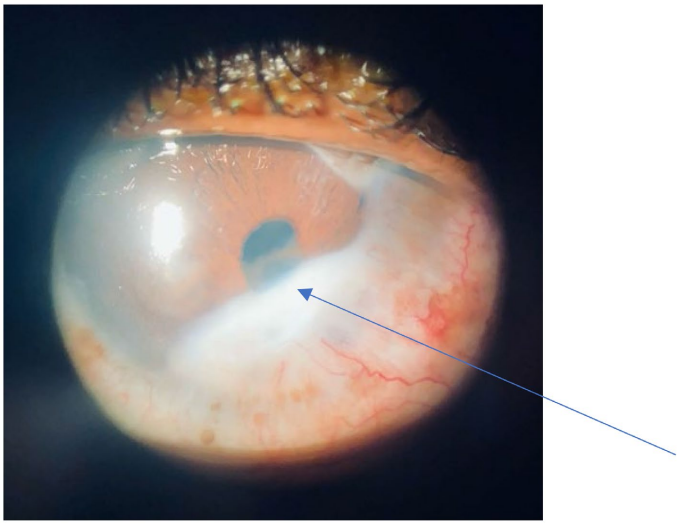

Figure 1. Corneal ulcer on the right eye seen with a slit lamp.

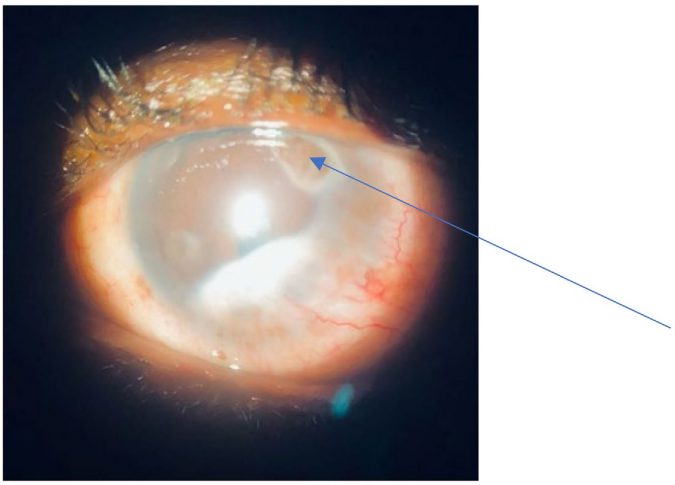

Figure 2. Corneal ulcer with iris herniation in the right eye seen with a slit lamp.

On week 4, the patient was seen in internal medicine consultation with the results of the paraclinical assessment. According to the baseline laboratory values, the tests revealed a biological inflammatory syndrome with a C-reactive protein level of 20.15 mg/l, mild anaemia at 12.8 g/dl with hyperleukocytosis at 10.49 G/l, predominantly neutrophils at 67%, and thrombocytosis at 484 G/l. The erythrocyte sedimentation rate (ESR) was not measured. The prescribed immunological assessment was positive for ANCA with anti-PR3 specificity at 182 AU/l (Antibody Unit/litre); rheumatoid factors, ACPA and antinuclear antibodies were negative. Serological tests for HCV, HIV and syphilis were negative.

The differential diagnosis discussion of PUK was excluded:

- Rheumatoid arthritis was ruled out as the patient did not have any joint symptoms or a positive ACPA.
- Relapsing polychondritis was excluded due to the absence of auricular, nasal, or laryngeal cartilage inflammation.
- Syphilis was excluded based on a negative serological test.

The assessment was completed by a clinical ENT examination, which was normal (no evidence of sinusitis or recurrent chondritis). Renal function, including creatinine at 81 µmol/l (GFR CKD-EPI = 115.5 ml/min) and urinalysis without proteinuria or haematuria, was normal. The chest X-ray was consistent with bilateral interstitial pneumonia, with no nodules observed. A chest CT scan was considered to rule out occult nodules but was not performed due to financial constraints.

The final diagnosis was ANCA-associated vasculitis of the granulomatosis with polyangiitis type, revealed by bilateral ulcerative keratitis.

In collaboration with ophthalmologists and internists, induction treatment is initiated based on oral corticosteroid therapy with prednisolone 1 mg/kg/day in combination with an immunosuppressant, notably methotrexate at a dosage of 0.3 mg/kg/per week with folic acid supplementation of 10 mg/week. A tapering of corticosteroid therapy is recommended, with a target dose of 5 to 7.5 mg/day at 4 months following induction and withdrawal at one year.

In week 6, the current visual acuity at the latest follow-up shows significant improvement in the left eye (visual acuity at 4/10), with resolution of the biological inflammatory syndrome (C-reactive protein level of 0.57 mg/l) and normalization of haemoglobin and platelets.

3. Discussion

The significance of this case report lies in the fact that granulomatosis with polyangiitis can manifest itself simply as bilateral ocular involvement. Furthermore, this ocular involvement can also compromise the visual prognosis in cases of delayed diagnosis.

Granulomatosis with polyangiitis (GPA) is characterized by extravascular necrotizing granulomatous inflammation, usually affecting the respiratory tract. In most cases, the clinical presentation is dominated by a pulmonary-renal syndrome with alveolar haemorrhage and rapidly progressive glomerulonephritis [8]. Other

organ damage is also observed. Ocular involvement is common in GPA, with ocular manifestations varying widely and reported in 16% to 78% of cases. In 27% of cases, they are the first sign of undiagnosed GPA [9].

Corneal involvement is a common ocular manifestation of GPA. It can be primary or secondary. The inflammation characteristic of GPA can specifically affect the cornea, leading to peripheral ulcerative keratitis (PUK) [10]. PUK is generally unilateral and sectoral but can also occur bilaterally in 40% of patients [11]. The collapse of the corneal epithelium makes the corneal stroma thinner and, if keratolysis progresses, it can cause perforation. Matrix metalloproteinases (MMPs) may contribute to the keratolysis observed. PUK in GPA is also linked to the presence of an autoantibody against cytokeratin 3 [12].

GPA has one essential element for diagnosis and monitoring: the presence of ANCA, with diffuse cytoplasmic fluorescence, directed against PR3 in 75% of cases and much more rarely against myeloperoxidase. They are present in approximately 90% of systemic forms and 50% of localized forms of the disease. They are highly specific and therefore have great diagnostic value. Under certain conditions, in combination with suggestive clinical signs, their presence may be sufficient for diagnosis [3].

Currently, the combination of glucocorticoids and immunosuppressive agents has changed the prognosis for patients with GPA, with a 5-year survival rate of 95% and a 10-year survival rate of 80% [3]. In secondary progressive or localized forms that require more "aggressive" treatment than cotrimoxazole, the treatment often combines:

- Corticosteroid therapy is initiated at a dose of 0.5 to 1 mg/kg/day of prednisone equivalent, without exceeding 60 mg/day. Bolus administration of methylprednisolone is exceptional in limited/localized forms. After 3 weeks of glucocorticoid treatment, the dose is gradually reduced [3] [13]. There is no internationally accepted tapering schedule. It varies from 6 months (North American protocols) to 18–24 months (European protocols) [14].
- An immunosuppressant, preferably methotrexate (at a dose of 0.3 mg/kg/week orally or subcutaneously), but cyclophosphamide or rituximab may also be used if necessary, according to standard procedures [3] [13].

4. Conclusion

GPA is a serious condition that can affect visual prognosis in its purely ocular form. Adequate management requires appropriate technical facilities and multidisciplinary collaboration. This atypical mode of presentation deserves to be reported consistently to raise awareness among clinicians about the early diagnosis of GPA.

Conflicts of Interest

The authors declare that they have no conflict of interest.

Acknowledgments

The authors would like to express their sincere thanks to the entire medical team of the Department of Internal Medicine, Endocrinology, and Metabolic Diseases at Bogodogo Teaching Hospital for their open collaboration.

References

[1] Rout, P., Garlapati, P. and Qurie, A. (2025) Granulomatosis with Polyangiitis. StatPearls Publishing. http://www.ncbi.nlm.nih.gov/books/NBK557827/

[2] Ntatsaki, E., Watts, R.A. and Scott, D.G.I. (2010) Epidemiology of ANCA-Associated Vasculitis. *Rheumatic Disease Clinics of North America*, **36**, 447-461. https://doi.org/10.1016/j.rdc.2010.04.002

[3] Puéchal, X. (2020) Granulomatosis with Polyangiitis (Wegener's). *Joint Bone Spine*, **87**, 572-578. https://doi.org/10.1016/j.jbspin.2020.06.005

[4] Mei, L., Wang, L. and Yan, H. (2022) Updates of Ocular Involvement in Granulomatosis with Polyangiitis. *Graefe's Archive for Clinical and Experimental Ophthalmology*, **261**, 1515-1523. https://doi.org/10.1007/s00417-022-05918-w

[5] Kharel, R., Maskey, H.M.S., Chaudhary, S. and Jha, S. (2022) Scleritis as the Harbinger of Granulomatosis with Polyangiitis. *Annals of Medicine & Surgery*, **84**, Article ID: 104908. https://doi.org/10.1016/j.amsu.2022.104908

[6] Zeroual, C., Moudatir, M., Echchilali, K., Barakat, L. and Kabli, H.E. (2024) A Bronchopulmonary Onset of Candidemia Revealing a Granulomatosis with Polyangiitis. *Case Reports in Clinical Medicine*, **13**, 147-154. https://doi.org/10.4236/crcm.2024.135016

[7] Rogaczewska, M., Puszczewicz, M. and Stopa, M. (2019) Exclusively Ocular and Cardiac Manifestation of Granulomatosis with Polyangiitis—A Case Report. *BMC Ophthalmology*, **19**, Article No. 139. https://doi.org/10.1186/s12886-019-1148-4

[8] Klapa, S., Arnold, S. and Lamprecht, P. (2024) Granulomatose mit Polyangiitis und mikroskopische Polyangiitis. *Laryngo-Rhino-Otologie*, **103**, 490-499. https://doi.org/10.1055/a-2217-4457

[9] Říhová, E., Svozílková, P., Brichová, M., Klímová, A., Kuthan, P. and Diblík, P. (2019) Ocular Manifestations of Granulomatosis with Polyangiitis. *Czech and Slovak Ophthalmology*, **74**, 167-174.

[10] Cao, Y., Zhang, W., Wu, J., Zhang, H. and Zhou, H. (2017) Peripheral Ulcerative Keratitis Associated with Autoimmune Disease: Pathogenesis and Treatment. *Journal of Ophthalmology*, **2017**, Article ID: 7298026. https://doi.org/10.1155/2017/7298026

[11] Sfiniadaki, E., Tsiara, I., Theodossiadis, P. and Chatziralli, I. (2019) Ocular Manifestations of Granulomatosis with Polyangiitis: A Review of the Literature. *Ophthalmology and Therapy*, **8**, 227-234. https://doi.org/10.1007/s40123-019-0176-8

[12] Reynolds, I., Tullo, A.B., John, S.L., Holt, P.J.L. and Hillarby, M.C. (1999) Corneal Epithelial-Specific Cytokeratin 3 Is an Autoantigen in Wegener's Granulomatosis-Associated Peripheral Ulcerative Keratitis. *Investigative Ophthalmology & Visual Science*, **40**, 2147-2151.

[13] Hellmich, B., Sanchez-Alamo, B., Schirmer, J.H., Berti, A., Blockmans, D., Cid, M.C., *et al*. (2024) EULAR Recommendations for the Management of ANCA-Associated Vasculitis: 2022 Update. *Annals of the Rheumatic Diseases*, **83**, 30-47. https://doi.org/10.1136/ard-2022-223764

[14] PNDS Vascularites Nécrosantes Systémiques. HAS 2021. https://www.fai2r.org/pnds/vascularites-necrosantes-systemiques/