BJMHR
British Journal of Medical and Health Research
Journal home page: www.bjmhr.com

Granulomatosis with polyangiitis (Wegener's disease) in a black patient revealed by neuro-ophthalmological involvement

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ABSTRACT

Granulomatosis with polyangiitis (GPA) is a systemic vasculitis. The authors report a case of Wegener's disease in a black patient revealed by neuro-ophthalmological involvement. A 38-year-old patient was seen in consultation for dizziness and headaches. In 2007, he presented with uveitis and retinal detachment, resulting in blindness in his right eye. Eleven years later, the condition recurred in the contralateral eye, although less severely. The neurological examination in December 2024 was normal. The ophthalmological examination noted sequelae lesions. Angio-MRI noted non-specific hypersignals in the white matter. The anti-polynuclear cytoplasm antibody test was positive for c-ANCA with anti-proteinase 3 specificity. Wegener's granulomatosis is a rare condition in our work context. It is a chronic, recurrent condition that is severe in both vital and functional terms.

Keywords: Wegener, granulomatosis with polyangiitis, black subject

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Received 02 August 2025, Accepted 28 August 2025

Please cite this article as: Thibault KY *et al*., Granulomatosis with polyangiitis (Wegener's disease) in a black patient revealed by neuro-ophthalmological involvement. British Journal of Medical and Health Research 2025.

INTRODUCTION

Wegener's granulomatosis is a necrotizing vasculitis involving inflammation of the vascular wall and peri- and extravascular granulomatosis, as well as the presence of autoantibodies directed against the cytoplasm of neutrophils (proteinase 3-type c-ANCA) [1]. Granulomatosis with polyangiitis (GPA) is one of the systemic vasculitides affecting small-caliber vessels, according to the classification adopted at the Chapel Hill consensus conference [2]. The authors report a case of Wegener's disease in a black patient with cytoplasmic anti-neutrophil cytoplasmic antibodies with anti-protease specificity revealed by neuro-ophthalmological involvement.

CASE REPORT

Mr. SD, born in 1987, was seen in neurology consultation in December 2024 for episodes of vertigo associated with transient headaches. The history of the symptoms reveals that they began in 2007 with a rapidly progressive decline in visual acuity in the right eye, leading to monocular blindness in the right eye. The patient then consulted an ophthalmology department, where initial examinations concluded that he had uveitis, which was accompanied by retinal detachment. The patient was then treated with oral corticosteroids without any impact on the progression of symptoms. Eleven years later (2018) by the onset of a rapidly progressive decline in vision in the left eye, for which the patient again consulted an ophthalmology department. A diagnosis of retinal detachment was made and the patient was treated with oral corticosteroids combined with eye surgery involving silicone injection (eight surgeries), without success. These ocular symptoms were associated with intermittent vertigo, slight tremors in the extremities, and mild to moderate headaches. These symptoms prompted a consultation with a neurologist, who noted instability when standing and performed an MRI angiogram, which suggested cerebral vasculitis. Autoimmune testing was negative, and the patient was again treated with oral corticosteroids. The patient then noted a regression of neurological disorders and stabilization of visual disturbances.

The patient was seen again in neurology consultation in December 2024 for a recurrence of mild to moderate headaches. The neurological examination was normal. The ophthalmological examination revealed reduced visual acuity in the right eye to negative light perception and pupillary seclusion with a central slit on slit lamp examination. The fundus examination was inaccessible on the right side. In the left eye, visual acuity was reduced to positive light perception, and pseudophakia was observed on slit lamp examination with a flat, grayish retina at the fundus. In addition, small, mobile, painless submandibular lymphadenopathy was noted. Para clinically, angio-MRI revealed non-specific hypersignals in the white matter, with no evidence of vasculitis (Figure 1). The chest CT scan was normal.

Ocular ultrasound showed an appearance consistent with total retinal detachment in the right eye and an appearance consistent with surgically repaired retinal detachment in the left eye. Optical coherence tomography of the macula was difficult to analyze in both the right and left eyes. Pathological examination of the accessory salivary gland biopsy showed normal findings. Analysis of the cerebrospinal fluid revealed lymphocytic meningitis (Table 1). Biological analysis to detect systemic disease revealed negative results for antinuclear antibodies, angiotensin-converting enzyme, antiphospholipid antibodies, and rheumatoid factor. The anti-neutrophil cytoplasmic antibody (ANCA) test was positive for c-ANCA with anti-proteinase 3 (anti-PR3) specificity. We therefore concluded that this patient had GPA, for which we reintroduced treatment with oral corticosteroids at 1 mg/kg/day for one month, followed by tapering doses combined with cyclophosphamide, for which we recommended six monthly courses of 1 g. The patient's condition remained stable after the first course of cyclophosphamide.

Table 1 Results of cerebrospinal fluid (CSF) and serum analyses

Parameters	Patient	Reference values
LCS analysis		
White blood cell	**19**/mm^3	<5/ mm^3
Red blood cell	**100**/ mm^3	**0**/ mm^3
Atypical cells	**0**/ mm^3	**0**/ mm^3
White blood cell subtypes		
• Lymphocytes	90%	Lymphocytes: 100%
• Neutrophils	10%	Neutrophils: 0%
• Lymphoblasts	0%	Lymphoblasts: 0%
Protein	0,35	< 0.50 g/l
Glucose	0,68	0.45 à 0.80 g/l
protein electrophoresis		
BOC	absent	absent
Ig Index	0,641	< 0,650
bacteriology	Negative	Negative
TPHA/VDRL	Negative	Negative
PCR BK	Negative	Negative
Serum analysis		
AAN	Negative	Negative (<10)
Facteur rhumatoide	Negative	Negative
ANCA	Positive (c-ANCA)	Negative
Anti-PR3 ANCA	11	>3 UI/ml (Positive)
Anti-MPO	0,4	<3,5UI/mL (Negative)
ECA	Negative	Negative
APL	Negative	Negative
Electrophorèse des protéines sériques	Normal	Normal
Urine analysis		
24-hour proteinuria	0,51	< 0,15g/24H

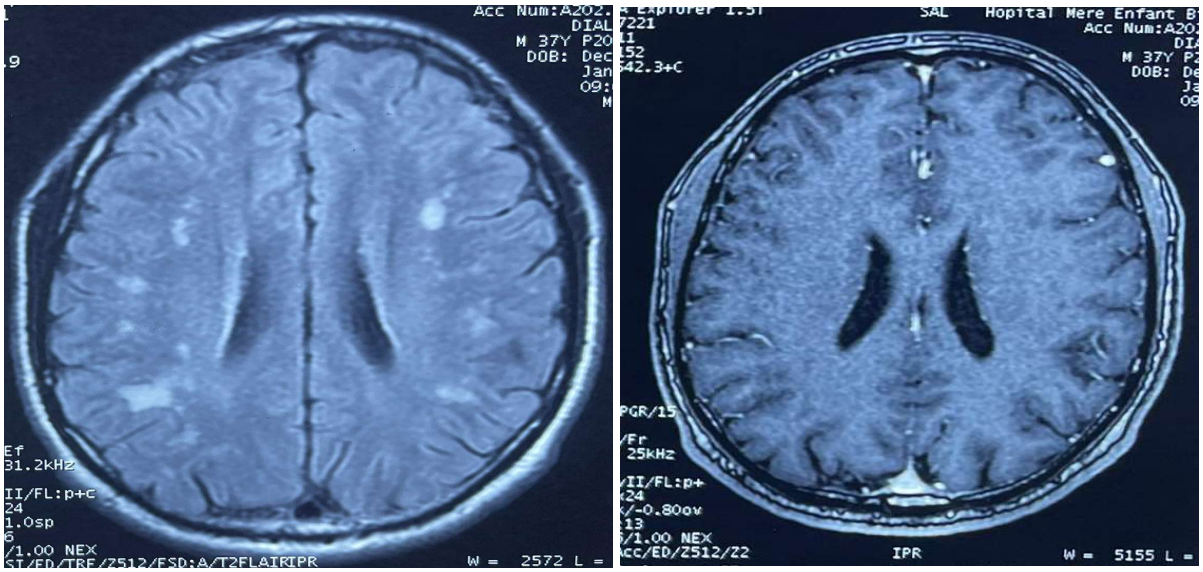

Figure 1: **Brain MRI using FLAIR and T1 sequences with contrast injection showing non-specific bihemispheric FLAIR hypersignals in the white matter with no contrast uptake after gadolinium injection**

Legend:

ECA: angiotensin-converting enzyme, **AAN**: antinuclear antibodies, **ANCA**: anti-neutrophil cytoplasmic antibodies, **Anti-MPO ANCA** : anti-myeloperoxidase ANCA, **Anti-PR3 ANCA** : the anti-proteinase 3 ANCA, **APL**: Anticorps anti-Phospholipid (the antibodies tested are anti-cardiolipin, anti-phosphatidylserine, anti-phosphatidylinositol, anti-phosphatidylethanolamine, anti-phosphatidic acid, and anti-beta 2-glycoprotein), **IgG**: Immunoglobulin G, **BOC**: Oligoclonal band , **PCR BK:** polymerase chain reaction au Bacille de Koch, **TPHA/VDRL**: Treponema Pallidum Hemagglutinations Assay/ Venereal Disease Research Laboratory. **WBC:** White Blood Cells Count

DISCUSSION

The incidence of GPA is estimated at between 2 and 10 new cases per year per million inhabitants, and its prevalence at between 24 and 150 cases per million inhabitants [2]. Caucasian ethnicity is the most frequently affected, accounting for more than 90% of patients in most series published to date [2]. It is rarer in black subjects, who account for only 2% of reported cases [3]. GPA affects both sexes with a mean age of onset of 45 years [1].

The disease usually manifests itself through varying degrees of damage to the ENT, respiratory, and renal systems [4]. Neurological damage in GPA is reported in 22 to 54% of patients. It is mainly peripheral neurogenic damage [4]. Central neurological involvement in SLE is rarer, affecting 4 to 8% of patients depending on the series [5]. Central nervous system involvement can manifest as headaches, cranial nerve involvement, meningeal syndrome, or even central motor or sensory deficits [2]. Brain MRI remains the gold standard for examining patients with SLE and central neurological manifestations. Examination

methods such as electroencephalogram and CSF examination do not provide evidence for a diagnosis of central involvement in GPA [4]. Brain MRI shows diffuse non-specific hypersignals in the white matter in 50% of cases [6].

Ocular involvement is present in 28 to 87% of patients and may be the initial symptom in 10 to 23% of cases. It can largely be explained by the proximity between the eye and the upper airways (sinuses and nasal cavities), which are very often affected during GPA [7]. It most often takes the form of scleritis, with uveitis being rarer in GPA [6].

GPA is accompanied by an essential element in the diagnosis and monitoring of the disease: the presence of ANCA, diffuse cytoplasmic fluorescence, directed against proteinase 3 in 75% of cases and much more rarely against myeloperoxidase. They are present in approximately 90% of diffuse forms and 50% of localized forms of the disease. They are highly specific and therefore have diagnostic value [1].

Treatment for GPA consists of induction therapy and maintenance therapy, based on the use of corticosteroids and immunosuppressants, which have radically transformed the prognosis for this condition [2]. The goal of induction therapy is to achieve remission in the patient, which is achieved within 3 to 6 months in the vast majority of cases. Once remission is achieved, a less toxic maintenance treatment is then started with the aim of maintaining this remission [2]. Cyclophosphamide, in combination with corticosteroid therapy, allows for prolonged remissions [1].

CONCLUSION

GPA is a rare condition in our work context. It is a chronic, recurrent condition that is severe in both vital and functional terms. Lack of knowledge about this condition has a negative impact on patient prognosis. However, patient prognosis can be improved with appropriate treatment

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