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Case Report **OPEN ACCESS**

Recurrent anemia to status epilepticus: A complex case of granulomatosis with polyangiitis with concurrent posterior reversible encephalopathy syndrome and intracranial hemorrhage

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ABSTRACT

Granulomatosis with polyangiitis (GPA) is a type of antineutrophil cytoplasmic antibody-associated vasculitis, a rare disorder affecting small vessels, primarily the respiratory tracts and the kidneys, with less common involvement of the central nervous system. Although posterior reversible encephalopathy syndrome (PRES) and intracranial hemorrhage (ICH) are documented complications, their concomitant occurrence in adults is extremely rare in this condition. We report a case of a 42-year-old Malay woman with heterozygous hemoglobin constant spring who presented with prolonged fever and recurrent severe anemia, posing a diagnostic dilemma. The diagnosis of GPA was confirmed through renal biopsy, and treatment was initiated. After a brief improvement, she developed status epilepticus secondary to concurrent PRES and ICH. This case highlights the diagnostic challenges of GPA and the rare, simultaneous occurrence of PRES and ICH, warranting careful therapeutic consideration due to both the disease and the treatment complications.

Keywords: granulomatosis with polyangiitis, hemoglobin constant spring, intracranial hemorrhage, posterior reversible encephalopathy syndrome

INTRODUCTION

Antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides (AAV) are a group of small vessel vasculitis which primarily include granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) apart from eosinophilic granulomatosis with polyangiitis (EGPA or Churg-Strauss syndrome) [1]. The staining patterns produced by c-ANCA or proteinase-3 antibodies in GPA are cytoplasmic, while p-ANCA or myeloperoxidase antibodies in MPA exhibit a perinuclear pattern. Histologically, GPA is characterized by the presence of granulomatous inflammation and multinucleated giant cells, which are absent in MPA. While distinguishing between these two conditions based on symptoms is challenging due to their overlapping manifestations, this differentiation may be of limited clinical significance as they share similar treatment approaches. The EGPA, however, is commonly discussed separately due to its differences in pathophysiology, symptoms and treatment [2].

The GPA and MPA can affect any small blood vessels, but the upper and lower respiratory tracts, kidneys, and skin are most commonly affected. Their initial manifestations are extremely heterogeneous. Nonspecific symptoms include fever, malaise, anorexia, weight loss, myalgias, and arthralgias [2]. Anemia is common, though severe anemia necessitating blood transfusions typically occurs later in the disease, where a significant degree of renal impairment is observed [3]. Organ-specific symptoms that are not severe include nasal and paranasal disease, skin involvement, myositis, non-cavitating pulmonary nodules and episcleritis. Potentially life-threatening manifestations encompass glomerulonephritis, pulmonary hemorrhage, neurological involvement, cardiac complications and retro-orbital disease [4].

The involvement of the central nervous system (CNS) is a rare but significant complication of AAV, due to its potentially devastating consequences. In GPA, neurological symptoms largely comprise the peripheral nervous system. CNS involvement only represents 10%, commonly presenting as pachymeningitis, and CNS vasculitis [5, 6]. Posterior reversible encephalopathy syndrome (PRES) is an uncommon complication of GPA, especially in normotensive adult patients with GPA. Concurrent PRES with intracranial hemorrhage (ICH), to our best knowledge, has not been documented so far in adults with GPA.

Here we report the unusual case of a woman with heterozygous hemoglobin constant spring (Hb CS) who presented with severe anemia at an early stage of the disease,

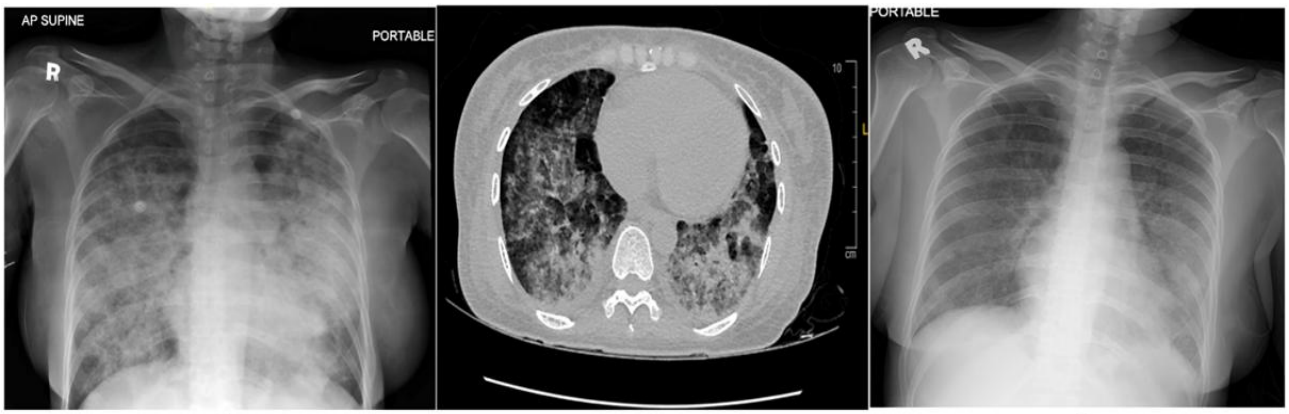

Figure 1. (a) Antero-posterior view of the chest X-ray showing diffuse nodular opacities suggestive of pulmonary hemorrhage; (b) Axial computed tomography of the chest showing "crazy paving" of bilateral lower zones, in line with pulmonary hemorrhage; & (c) Antero-posterior view of the chest x-ray showing resolution of pulmonary hemorrhage after completion of the first cycle of induction (Reprinted with permission of the patient)

warranting multiple blood transfusions, alongside persistent fever. GPA was diagnosed after she developed pulmonary-renal syndrome, and induction therapy was initiated. During treatment, she developed status epilepticus secondary to concurrent PRES and ICH. This case highlights the diagnostic challenges and rare complications involved, necessitating the need for meticulous management of both the disease and its treatment-related issues.

CASE PRESENTATION

This is a case of a 42-year-old Malay woman who presented with a two-month history of prolonged fever and anemia. Her medical history includes heterozygous Hb CS with a stable hemoglobin baseline of 10 to 11 g/dL. She was previously well with no history of hospitalization. She works as a dental assistant, where she prepares dental fillings with amalgam and glass ionomer cement containing mercury and silica.

Prior to the diagnosis, she had recurrent hospitalizations and was treated for pneumonia with symptomatic anemia. However, she reported no symptoms related to her lungs, ears, nose, throat, skin, and connective tissues. Initial blood results showed severe anemia with hemoglobin of 5.2 g/dl while the total white cell count was 12 × 109/L, platelet count 513 × 109/L, C-reactive protein 60 mg/L. Renal function was normal. The peripheral blood smear showed erythrocyte changes consistent with hemoglobinopathy with concurrent iron deficiency anemia. In these hospitalizations, she received intravenous antibiotics and blood transfusions, which temporarily improved her condition, permitting her to be discharged.

She was later readmitted due to recurrent anemia and rapidly progressive glomerulonephritis. Her hemoglobin level was 4.6 g/dL, and her renal function deteriorated significantly, with creatinine rising from 65 µmol/L to 298 µmol/L. Although she was febrile, blood cultures were negative. Both echocardiogram and abdominal ultrasound showed no source of infection. Additionally, a renal Doppler ultrasound indicated no evidence of obstruction with normal renal blood flow. A gastrointestinal endoscopy was planned to exclude occult bleeding, and bone marrow aspiration was considered due to concerns of hematological abnormalities; however, the patient declined both procedures.

During this admission, she developed hemoptysis and respiratory distress requiring high-flow nasal cannula and transfer to the intensive care unit. A plain chest radiograph revealed diffuse lung opacities (part a in **Figure 1**), and computed tomography of the chest showed diffuse consolidation of bilateral lungs with "crazy paving", suggesting pulmonary hemorrhage (part b in **Figure 1**). Antinuclear antibodies, immunoglobulins, complements, tumor markers, and anti-glomerular basement membrane (anti-GBM) study were negative. However, an indirect immunofluorescence ANCA test was positive, with a cytoplasmic pattern at a titer exceeding 1:160. This led to a diagnosis of pulmonary renal syndrome secondary to ANCA-associated vasculitis, with GPA being the likely subtype. Subsequent renal biopsy revealed pauci-immune necrotizing crescentic glomerulonephritis, with disruption of Bowman's capsule and aggregation of histiocytes forming granuloma-like structures around the glomeruli. Tubulointerstitial inflammation was also present (**Figure 2**). These findings confirmed the diagnosis of GPA. Pulse intravenous cyclophosphamide (12.5 mg/kg) was then initiated with intravenous methylprednisolone 500 mg daily for five days. After completing the first cycle of induction therapy, hemoptysis and respiratory distress resolved, with significant chest radiograph improvement (part c in **Figure 1**). Her renal function also showed marked improvement, and her hemoglobin level stabilized, allowing her to be discharged. She was scheduled for a total of ten cycles of pulse intravenous cyclophosphamide, three weeks apart and a tapering dose of oral prednisolone.

However, she returned two weeks after discharge for recurrent anemia and new skin lesions. The skin examination showed multiple purpuric and necrotic papules and plaques over several digits of her bilateral hands, ecchymoses over her bilateral cubital fossae, and an irregularly shaped necrotic plaque with multiple bullae over her left-hand dorsum (**Figure 3**). Biopsy of these lesions revealed changes consistent with leukocytoclastic vasculitis. Immunofluorescent staining was negative. Her serum creatinine surged from 120 µmol/L to 648 µmol/L, and her hemoglobin dropped from 9.5 g/dL to 5.6 g/dL, necessitating hemodialysis and blood transfusions.

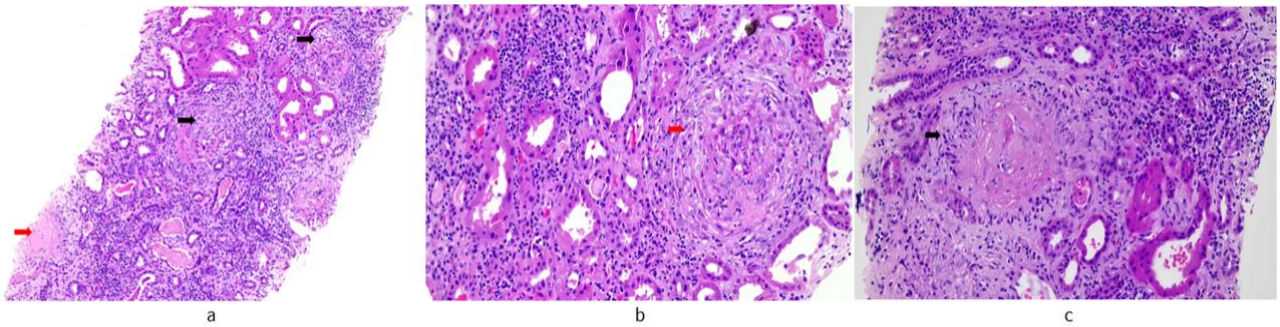

Figure 2. (a) The section shows a globally sclerotic glomeruli (red arrow) and a cellular crescent (black arrow). Moderate infiltration of lymphocytes is observed in the interstitium (hematoxylin and eosin [H&E] stain, 100× magnification); (b) The cellular crescent (red arrow) is noted with infiltration by neutrophils, along with mononuclear cells and karyorrhectic debris in the glomeruli with a small focus of fibrinoid necrosis (H&E stain, 200× magnification); & (c) Disruption of the Bowman's capsule is observed (black arrow), accompanied by an aggregation of histiocytes forming a periglomerular like epithelioid granuloma formation (H&E stain, 200× magnification) (Reprinted with permission of the Department of Pathology of authors' institution)

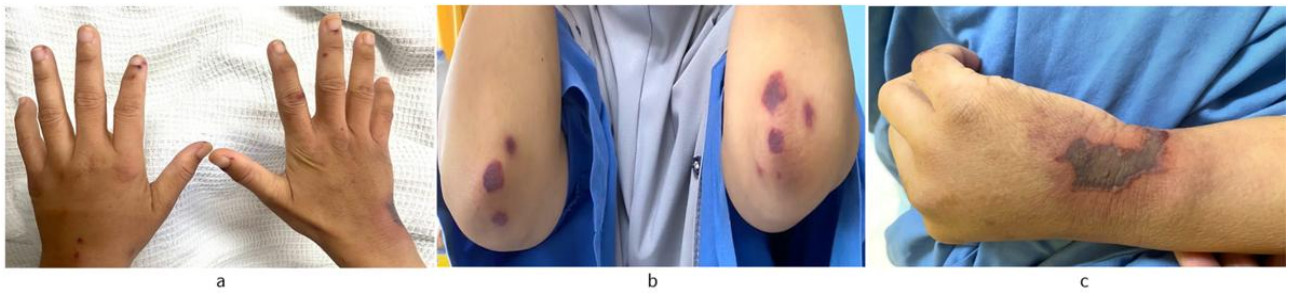

Figure 3. (a) Multiple purpuric and necrotic papules and plaques over several digits of bilateral hands; (b) Ecchymoses on bilateral cubital fossae; & (c) An irregularly shaped bullous necrotic plaque on left-hand dorsum (Reprinted with permission of the patient)

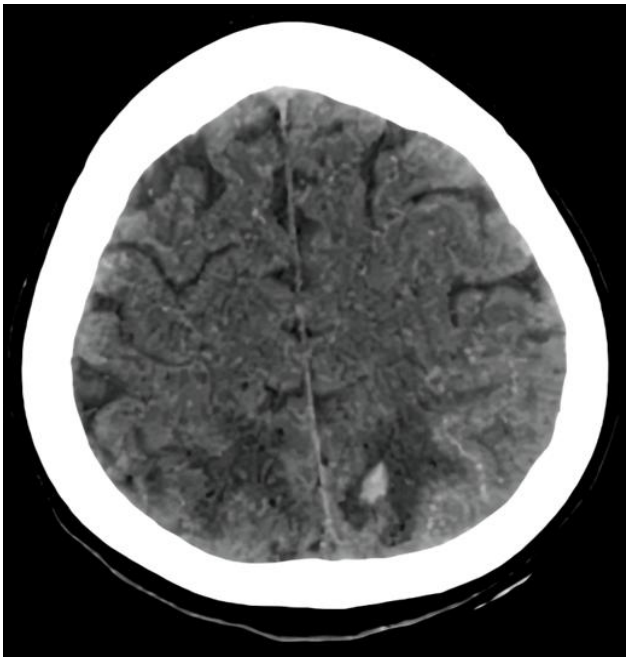

Figure 4. Axial computed tomography of the brain showing symmetrical hypodensities of the white matter over bilateral parieto-occipital areas, consistent with vasogenic edema (hyperdensity over left parieto-occipital region, indicating intraparenchymal and subarachnoid hemorrhage) (Reprinted with permission of the patient)

Plasma exchange was initiated, and a second cycle of intravenous cyclophosphamide commenced. The patient was discharged after completing seven cycles of plasma exchange and was scheduled to continue induction in three weeks.

She was readmitted five days later due to complaints of blurred vision and headaches, which subsequently progressed to status epilepticus. She was hemodynamically stable upon arrival with a blood pressure of 109/54 mmHg. The seizure was aborted with intravenous diazepam and phenytoin, and she was intubated. Computed tomography of the brain with venogram revealed symmetrical white matter hypodensities at the bilateral parietooccipital regions, implying vasogenic edema consistent with PRES. Left parietal intraparenchymal hemorrhage and left parieto-occipital subarachnoid hemorrhage were also seen (**Figure 4**). Intravenous dexamethasone 40 mg three times daily was started, and no seizure was then observed. A lumbar puncture revealed normal cerebrospinal fluid biochemistry and no bacterial growth. She was extubated after two days of admission, and a third dose of pulse intravenous cyclophosphamide was administered during her stay. She remained normal neurologically and had stable blood parameters in the ward. She was discharged, with further continuation of her induction treatment.

DISCUSSION

GPA is a type of AAV characterized by granulomatosis and systemic necrotizing vasculitis. As with most autoimmune diseases, their exact cause is poorly understood, but genetic predisposition, environmental exposures, and infections are thought to be contributing factors. An interesting point is that the exposure to silica and mercury, both components of dental

filling, as observed in our patient, has been linked to an increased risk of ANCA positivity [7].

The pathogenesis of AAV involves the production of pro-inflammatory cytokines, which prime neutrophils to express target antigens, either proteinase-3 or myeloperoxidase. The subsequent binding of circulating ANCAs to these antigens and the Fcγ receptors of neutrophils leads to a cascade of inflammation-induced endothelial cell injury, ultimately resulting in vascular injury and ischemia, manifesting as symptoms depending on the organs affected [8]. While the symptoms can be broad, the classic triad typically involves the ear, nose, and throat, lungs and kidneys. Nonspecific, less life-threatening symptoms are, however, the common initial presentations, making the diagnosis challenging and delaying the institution of the much-needed immunosuppressants, as more indolent symptoms will usually lead to less rigorous investigation and intervention.

We aim to highlight two key points in this case report: First, the diagnostic challenge posed by the unusual initial presentation of intractable anemia early in the course of the disease, without the more classical symptoms, which obscured the underlying vasculitis. Second, the rare occurrence of simultaneous PRES and ICH underscores treatment complexity, as both the active disease and its therapy predispose the patient to these life-threatening complications.

Anemia in AAV is primarily contributed to by renal anemia, anemia of inflammation and iron deficiency anemia [3]. Concurrent sepsis, pulmonary hemorrhage and malnutrition can exaggerate the degree of anemia in hospitalized AAV patients. Renal vasculitis in AAV causes peritubular capillaritis and Bowman's capsule rupture, impairing erythropoietin production and leading to anemia [9]. The degree of tubulointerstitial and glomerular damage correlates with the severity of anemia, which is associated with a poor prognosis [3]. Due to our patient's underlying heterozygous Hb CS and an ongoing systemic inflammation, her anemia is significantly more pronounced at an early stage of AAV, when her renal impairment was relatively mild. The severity of her anemia and the absence of more classical manifestations of AAV, such as those related to ear, nose, and throat, created a diagnostic dilemma. The initial workup was intended to investigate possible causes of blood loss and hematological abnormalities. It was when the patient developed the pulmonary-renal syndrome, which is a more typical complication of AAV, that ANCA and anti-GBM serologies were conducted, leading to a possible diagnosis of AAV and initiation of treatment.

Neurological involvement is a significant, life-threatening manifestation of AAV [4]. CNS manifestation is rare in GPA, reported in 10% of patients [5]. The main CNS manifestations include pachymeningitis, cranial neuropathies, vasculitis of the cerebral arteries leading to ischemic infarction, intracerebral or subarachnoid hemorrhage, and arterial and venous thrombosis [6]. The PRES is a recognized CNS complication of AAV. To date, only nine cases of PRES in GPA have been reported, with only one presenting without hypertension [10-18]. The coexistence of PRES and ICB in GPA is exceedingly rare in adults.

PRES, first described by Hinchey in 1996, is a clinical-radiological diagnosis. It typically presents with acute seizures, visual disturbances and headaches. The hallmark neuroimaging finding is vasogenic edema, which predominantly affects the parieto-occipital regions in a bilateral, subcortical and symmetrical fashion. Concurrent intracerebral hemorrhage is an atypical finding [19]. Triggering factors of PRES include hypertension, pre-eclampsia/eclampsia, renal failure, cytotoxic agents and autoimmune conditions [20].

The pathophysiology of PRES is still poorly understood, but several mechanisms have been proposed. The main theory, the vasogenic theory, suggests that rapid hypertension causes the cerebral autoregulatory system to fail, leading to a breakdown of the blood-brain barrier and vasogenic edema. However, this theory does not fully explain PRES in patients with borderline or normal blood pressure, such as in our case. Alternative explanations are more in line with our patient's condition. One such theory, the neuropeptide theory, posits that vasoconstrictors such as endothelin-1 and thromboxane A2 induce endothelial injury. Conversely, the immunogenic theory postulates that cytokine release and T-cell activation lead to endothelial dysfunction.

Additionally, the cytotoxic theory suggests that both endogenous stimulants and exogenous toxins, including those from immunosuppressants, directly damage endothelial cells. Despite the different mechanisms, all these theories ultimately converge on endothelial injury, blood-brain barrier disruption, and the resulting vasogenic edema that characterizes PRES [19]. This endothelial dysfunction also predisposes vessels to rupture, resulting in ICH. The treatment of PRES and ICH in GPA is mainly supportive. Key components include blood pressure stabilization in hypertensive patients, discontinuation of offending agents such as immunosuppressants or chemotherapy, treatment and prevention of seizures, hydration and correction of electrolytes imbalance and maintenance of good oxygenation. In pregnant women, prompt delivery should be considered. Neurosurgical intervention is warranted if ICH is large or associated with a significant mass effect [20].

Immunosuppressants are the cornerstone therapy in patients with AAV and should be started once the diagnosis is made. Given the fatal consequences of untreated AAV and the risks related to therapy, overarching principles of AAV treatment should be applied to ensure patient's understanding of their disease, enabling them to make an informed decision and adhere to their treatment plans [4]. In patients with life-threatening organ involvement, remission induction with glucocorticoids and either cyclophosphamide or rituximab, along with plasma exchange in rapidly progressive glomerulonephritis, should be started [4]. In our case, while the severity of AAV was initially managed, the patient subsequently developed a life-threatening CNS complication of PRES and ICH, which is attributable to both the disease itself and the effects of immunosuppression. In regard to prognosis, a study concluded that a serum creatinine of ≥ 124 μmol/L and age of ≥ 65 years at diagnosis are the most important predictors of reduced patient survival [21]. As our patient faces complications involving the skin, kidneys, lungs and brain, her prognosis is unfortunately unpromising. Remission will be challenging; close monitoring in a rheumatology center is essential.

CONCLUSION

GPA poses substantial diagnostic challenges owing to its protean clinical manifestations, which can overlap with existing patient conditions, making timely diagnosis and

treatment difficult. The concurrent occurrence of PRES and ICH, both rare manifestations in their own right, is even more unusual in GPA. These complications, combined with multisystem involvement, reflect a poor prognosis and a challenge in treatment as both the disease and its therapies carry significant risks, underscoring the need for the patients' understanding of this debilitating disease.

Author contributions: WMNWN: wrote the clinical presentation, findings, and discussion; **MNN & WSWG:** were responsible for the concept of the manuscript, clinical case, and discussion; **AI:** edited and reviewed the manuscript substantively; **HS:** performed the skin biopsy and wrote the clinical findings section; & **NACJ:** was responsible for the histopathological examination and images. All authors have agreed with the results and conclusions.

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Declaration of interest: No conflict of interest is declared by the authors.

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