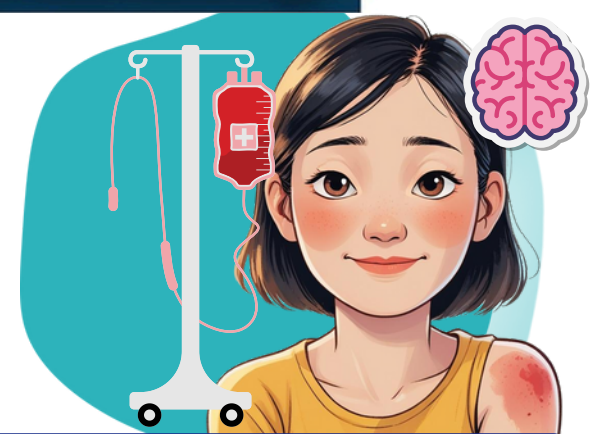


FROM SKIN TO BRAIN: THE STORM OF C-ANCA VASCULITIS WITH MULTISYSTEMIC INVOLVEMENT

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INTRODUCTION

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a rare autoimmune disorder characterized by inflammation of small blood vessels, often affecting multiple organs. Its clinical presentation varies, which can delay diagnosis and lead to significant morbidity and mortality if not treated promptly.

CASE REPORT

This case discusses a 42-year-old woman with HbH thalassemia, initially presenting with recurrent anemia requiring frequent blood transfusions. Her condition worsened with hemoptysis, shortness of breath, and a rash on her elbows.

Over two months, her kidney function deteriorated, and her eGFR dropped from 85 to 6 mL/min/m². Laboratory tests revealed a positive c-ANCA titer ($\geq 1:160$), and a renal biopsy confirmed pauci-immune necrotizing crescentic glomerulonephritis. A skin biopsy was consistent with leukocytoclastic vasculitis, and a chest CT scan showed granulomatous changes, diagnosing her with granulomatosis with polyangiitis (GPA).

She was treated with high-dose intravenous methylprednisolone and cyclophosphamide, but her kidney function continued to decline, requiring seven cycles of plasma exchange.

After completing plasma exchange, she was readmitted with seizures, and imaging bilateral parieto-occipital vasogenic edema (suggestive of posterior reversible encephalopathy syndrome), along with an acute intraparenchymal hemorrhage and subarachnoid hemorrhage. She was treated with dexamethasone, and infection was ruled out.


The patient is now on her 9th cycle of cyclophosphamide, with improving renal function and no need for dialysis.

CONCLUSION

The global incidence of AAV varies from 1.1 to 20.4 per 1 million person-years, with GPA being the most common subtype.¹ Cerebral small-vessel vasculitis occurs in about 4% of GPA patients, and PRES, though rare, has been linked to GPA.² Conditions like severe inflammation, immunosuppressive use, renal failure, or hypertension can cause endothelial dysfunction in the cerebral vasculature, contributing to PRES.³ Without treatment, the average life expectancy for a GPA patient is only 5 months, but with treatment, more than 80% survive for at least 8 to 9 years.⁴


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42 years
with HbH Thalassemia


HER CONDITION WORSENER WITH:



Hemoptysis




Shortness of breath




Rash on her elbow


Initial Presentation



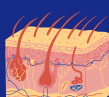
eGFR dropped
from 85 to 6 mL/min/m² (in just 2 month)



Lab test revealed
Positive c-ANCA titer ($\geq 1:160$)



Renal Biopsy
Pauci-immune necrotizing crescentic glomerulonephritis



Skin Biopsy
Leukocytoclastic vasculitis

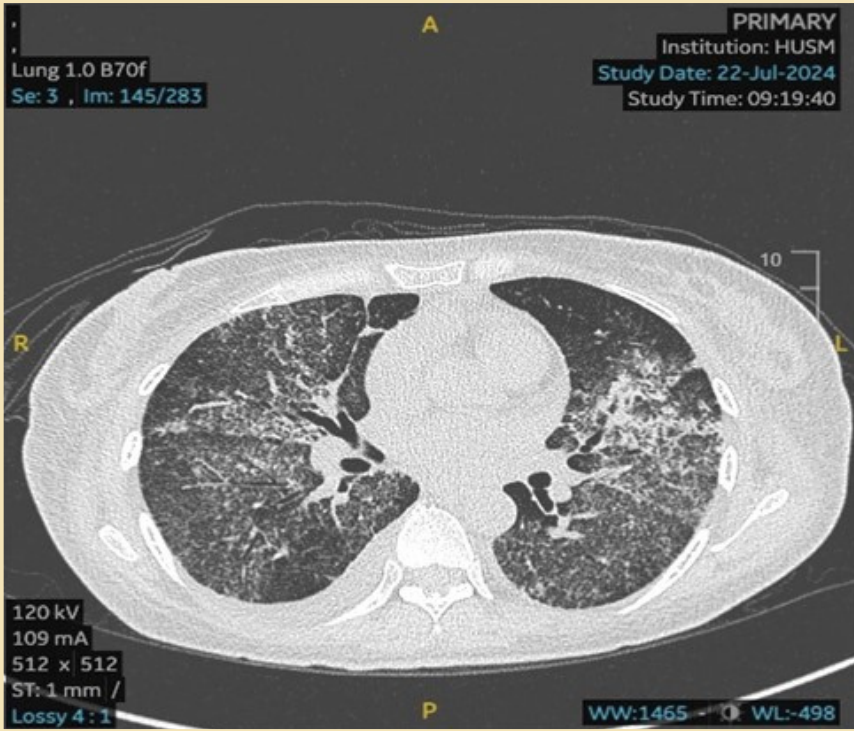




Figure 1 : Widespread ground-glass opacities are seen in both lungs, sparing the apices and extreme bases. Numerous perilymphatic micronodules are present within these areas, with some merging into consolidations, which suggestive of granulomatous disease.


Initial Treatment



Methylprednisolone & cyclophosphamide




Kidney function continued to decline




Seven cycles of plasma exchange

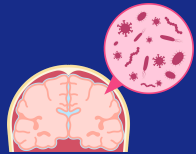
Neurological Complication



Readmit with seizures



Treated with dexamethasone



Infection was ruled out






Figure 2 : Bilateral parieto-occipital vasogenic edema, likely due to PRES, with acute left parietal brain bleed and subarachnoid hemorrhage.


Current Status



On 9th Cyclophosphamide cycle



Improving kidney function



No more dialysis needed