Acute Psychosis Associated with Granulomatosis with Polyangiitis: A Case Discussion and Literature Review

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BACKGROUND

Granulomatosis with polyangiitis (GPA), also known as Wegener's granulomatosis, is a rare disorder causing vasculitis that predominantly impacts the kidneys, lungs, and upper respiratory tract. We present a case of recurrent psychosis in a patient with GPA that resolved with rituximab, and discuss the differential diagnosis and clinical course.

CASE

A 44 year-old woman with a history of posttraumatic stress disorder (PTSD), anti-neutrophil cytoplasmic antibody (ANCA)-negative GPA, and trigeminal neuralgia was admitted to the intensive care unit following a suicide attempt by ingestion of Excedrin in the context of persecutory delusions. In the months preceding her hospitalization, she was seen by outpatient otolaryngology and rheumatology for necrotic nasal lesions secondary to GPA, confirmed by biopsy. This included treatment with oral prednisone, which the patient self-discontinued a week prior to hospitalization. She also engaged in occasional cocaine use, including several days before her presentation. She had one prior episode of psychosis several months before hospital admission which rapidly resolved after overnight observation and was attributed to high-dose steroids and cocaine use. The duration of the patient's psychiatric symptoms remained uncertain based on available history.

While hospitalized, the rheumatology team diagnosed active vasculitis and started rituximab treatment. Brain magnetic resonance imaging (MRI) showed mild atypical white matter hyperintensities on T2 sequences. Although she initially refused antipsychotic medication, her persecutory delusions improved over the following three days, with some residual paranoia. She eventually agreed to treatment with aripiprazole, and was discharged two days later as her psychotic symptoms resolved.

CASE TIMELINE

1 week prior to presentation
Patient self-discontinued PO steroid

Several days prior
Patient reported most recent cocaine use

Hospital presentation
The patient was admitted to the hospital following a suicide attempt by intentional medication overdose

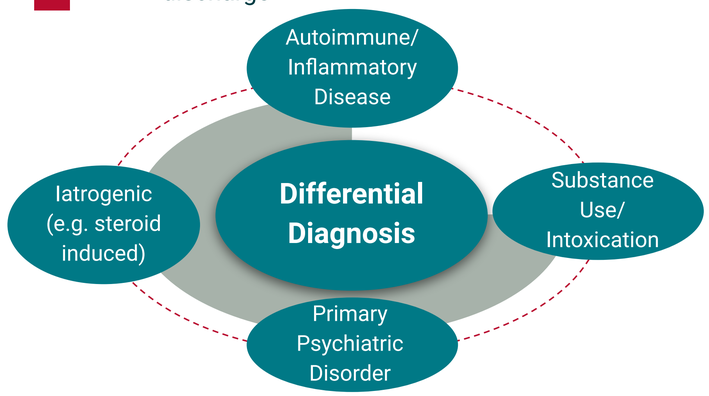
Hospital day 2
Rheumatology team established diagnosis of active ANCA-negative granulomatosis with polyangiitis

Hospital day 3
Rituximab induction completed

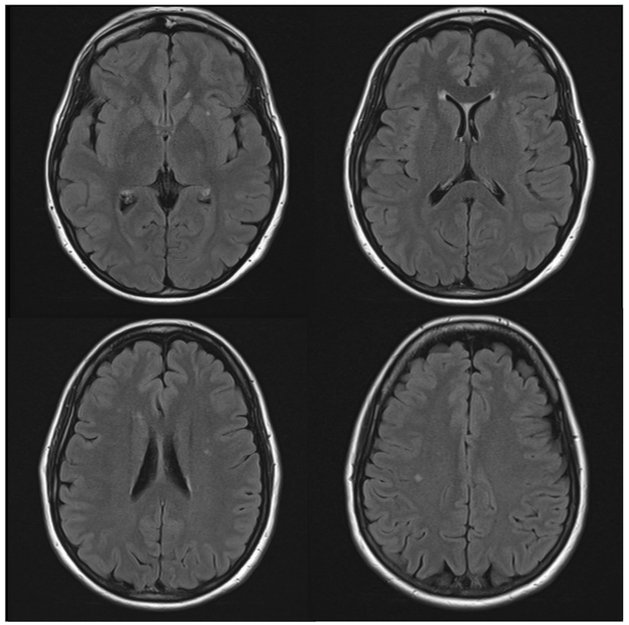
Hospital day 6
Improvement in psychotic symptoms noted by psychiatry team

Hospital day 7
Aripiprazole initiated

Hospital day 9
Resolution of psychotic symptoms and hospital discharge



RESULTS



Lab	Result
ESR	2 mm/hr
CRP	<3 mg/dL
ANCA Antibodies	Negative
Anti-MPO Antibodies	Negative
Anti-PR3 Antibodies	Positive

(Left) Selected images from brain MRI (T2 FLAIR) completed during admission showing mild white matter hyperintensities, atypical for age.
(Above) Selected laboratory results from inpatient admission.

DISCUSSION

Identification of neuropsychiatric symptoms due to rheumatologic disorders such as GPA has important ramifications for treatment selection, yet is challenging due to their insidious onset, comorbid conditions, and known behavioral effects of corticosteroids. Case reports have documented ANCA-negative GPA causing psychosis[1] and delusional parasitosis[2]. Definitive central nervous system involvement has also been described. Prompt identification and treatment is imperative; if left untreated, the mortality rate of ANCA-associated vasculitides is over 80%[3].

In assessing this patient's psychotic symptoms, the presence of active vasculitis, persistence of psychosis after cocaine metabolization, and improvement following immunomodulatory treatment and prior to initiating antipsychotic medication elevated suspicion for an autoimmune or steroid-induced etiology. Accurate history-taking, physical examination for inflammatory stigmata, biomarker and imaging evaluation, and longitudinal assessment of symptoms are necessary components of an approach to psychosis due to GPA.

References

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