Intracranial Hemorrhage Due to ANCA-Associated Isolated Cerebral Vasculitis

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Because of its rarity, cerebral vasculitis may seem like a medical zebra. Early diagnosis of this rare, but treatable, cause of neurologic symptoms improves chances of survival. For that reason, vasculitis should be included in the differential diagnosis for stroke patients presenting with evidence of systemic inflammation such as myalgias, joint pains or constitutional symptoms.1

Case
A 64-year-old healthy female presented with a three-hour history of right-sided hemiparesis. Three weeks prior to presentation, the patient had developed myalgias and difficulty controlling her left ankle. One week prior to admission, she had noted painless vision loss in her right eye.

Pertinent physical exam findings included a right eye without pupillary reflex, left foot drop, right upper and lower extremity weakness, and right up-going Babinski reflex. The patient was noted to be lethargic and was becoming progressively more confused.

MRI showed a large left parietal parenchyma hemorrhage, a small hemorrhage in the left temporal subcortical area and a small basal ganglia infarct. Additionally, there was mild reactive meningeal enhancement. On presentation, she had leukocytosis at 31.7, and her c-reactive protein was 185.8 mg/L. Infectious work-up was negative. Rheumatologic work-up resulted in a very high titer for cANCA/PR3 with final diagnosis being granulomatosis with polyangiitis (formally known as Wegener’s granulomatosis) with isolated cerebral vasculitis.

Once inflammatory markers and MRI findings were apparent, systemic corticosteroids were initiated. At discharge, the patient had recovered almost full function of her right arm and leg. Arrangements were made for outpatient rituximab infusions.

Discussion
The major symptoms of cerebral vasculitis are stroke, headache and encephalopathy. When evaluating a patient with a new neurologic deficit, the diagnosis is broad and includes embolic disease, coagulation disorders and degenerative vasopathies. Vasculitis should be included in the differential diagnosis for stroke patients presenting with evidence of systemic inflammation such as myalgias, joint pain or constitutional symptoms.2

About 25% of patients with ANCA-associated vasculitis will develop nervous system involvement.3 Their symptoms generally present in the form of peripheral or cranial mononeuropathies. However, 2% to 8% of patients will have involvement of the brain or meninges, presenting with infarctions or seizures.4

For the past 40 years, cyclophosphamide has been used to successfully treat ANCA-positive vasculitis. However, recent work suggests rituximab may be superior in preventing disease relapse. In a recent randomized control trial, induction therapy with one month of weekly rituximab was not inferior to six months of daily cyclophosphamide. Furthermore, rituximab is better tolerated.2,4

This case demonstrates the importance of fully reviewing systems in patients presenting with common disorders such as a stroke. Early diagnosis is vital in these cases because ANCA-associated vasculitis is highly treatable. Remission can occur with current treatment strategies, improving both survival and neurologic outcomes. MM

REFERENCES