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Vasculitic Stroke in Polyarteritis Nodosa – A Case Report

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Background: Polyarteritis nodosa (PAN) is a rare autoimmune vasculitis characterized by systemic inflammation of medium- sized arteries, leading to diverse clinical manifestations. Neurologic manifestations are very common in polyarteritis nodosa (PAN) due to systemic necrotizing vasculitis and cause ischemia, leading to thrombosis or bleeding. This case aims to highlight the importance of considering vasculitis, particularly in PAN, the differential diagnosis of strokes, especially when multiple vascular territories are affected.

Methods: We present a challenging case of a vasculitic stroke in a 57-year-old male with a history of PAN, who presented with sudden-onset no verbal output and right-sided weakness. Neuroimaging revealed an acute ischemic stroke involving the left frontal lobe, corona radiata, and left insula. Laboratory investigations, including serological markers, supported the diagnosis of active PAN with central nervous system involvement. The patient was promptly initiated on high-dose corticosteroids.

Results: The treatment led to a gradual improvement in the patient's neurological deficits over several days.

Conclusion: Timely recognition and aggressive immunosuppressive treatment are crucial to managing vasculitic strokes associated with PAN, aiming to prevent further neurological complications and enhance overall outcomes. This report highlights the need for multidisciplinary collaboration between rheumatologists and neurologists for accurate diagnosis and optimal management of this complex condition.

Keywords: Polyarteritis Nodosa, Autoimmune Vasculitis, Vasculitic Stroke

Introduction

Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that typically affects medium-sized muscular arteries, with additional involvement of small arteries [1]. It is a rare form of primary systemic vasculitis with heterogeneous presentations, treatments and disease course [2]. It presents with clinical manifestations resulting from ischemia and infarction of affected tissues and organs [3].

Vasculitis is a process associated with various syndromes, characterized by inflammation and necrosis of blood vessels of virtually any size [4]. It is the inflammation caused by the abnormal accumulation of white blood cells in the wall of the blood vessels. The reaction of these cells causes structural damage to the affected blood vessels. Vasculitis can affect any blood vessels. It is a spectrum of clinicopathological disorders defined by inflammation of the blood vessels, including arteries and veins of varying caliber, which result in a variety of clinical neurological manifestations related to ischemic injury of the central nervous system (CNS) and peripheral nervous system (PNS)6.

Neurologic manifestations are very common in polyarteritis nodosa (PAN) due to systemic necrotizing vasculitis that affects medium-sized arteries and cause ischemia, leading to thrombosis or bleeding. CNS symptoms occur relatively late during the disease, causing multifocal encephalopathy in 40% of patients, depending on the region in which lesions appear. Patients with PAN typically demonstrate CNS neurological signs, such as personality and memory disorders, atypical persistent headaches, aphasia, hemiplegia, visual disturbance (blurred vision and hemianopia), seizures, transverse myelitis and subarachnoid hemorrhage. Up to 65% of patients present with PNS disorders, including mononeuritis multiplex (almost typical and generally painful manifestation of the disease) and distal symmetric sensorimotor polyneuropathy [5].

Case Illustration

We present a case of a 57 year old male, who is a known case of Polyarteritis Nodosa of more than five years. The patient's polyarteritis nodosa was diagnosed on early 2000s and was maintained on Prednisone, Methotrexate, and Pregabalin.

The patient presented with no verbal output of two hours duration with associated right facial asymmetry, and right sided weakness. Upon arrival at the emergency room pertinent examination findings were – globally aphasic, inconsistently communicates through gestures and nodding, midline gaze with occasional preferential gaze to the left but with visual tracking and can cross midline, and, shallow right nasolabial fold. Motor strength testing revealed 1/5 on the right upper extremity, and 2/5 on the right lower extremity. There were no sensory deficits noted.

Diagnostics were done, including cranial MRI, showing moderate - sized acute infarct involving the left frontal lobe, the corona radiata and left insula, chronic lacunar infarcts involving the bilateral anterior centrum

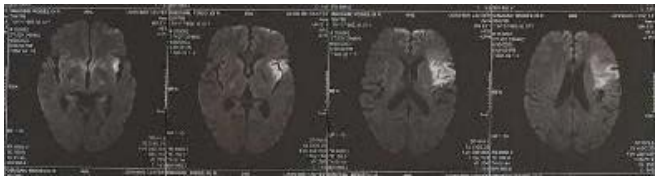

Figure 1. DWI sequence of Plain Cranial MRI

MRA of the brain demonstrates mild to moderate atherosclerotic irregularities involving opercular branches of the left MCA. There is also scattered mild, atherosclerotic changes. Other workup noted on blood chemistry that erythrocyte sedimentation rate (ESR) was elevated. On complete blood count (CBC), noted slight elevation of neutrophils on differential count, with normal white blood cell (WBC) count.

The patient, presenting within the golden period of less than 4 hours, was eligible and a candidate for recombinant tissue plasminogen activator (rTPA), given an initial impression of hyperacute cerebrovascular disease infarct, left middle cerebral artery territory, M4 segment (NIHSS 12, HAT 0, SEDAN 1), thrombotic vs vasculitic in etiology. But due to financial constraints, family has opted to be managed as delayed management and secondary prevention, instead.

Patient was initiated on Aspirin, and Atorvastatin. Rheumatology service initiated patient on Hydrocortisone 100mg/day. Previous maintenance medications - Pregabalin, Methotrexate, Folic acid, Pantoprazole, were continued and maintained. On day 1 of hospitalization, Hydrocortisone was shifted to Prednisone. It was also noted that there was improvement of language to Broca's Aphasia. Motor strength noted to have improved, with right upper extremity at 2/5, and right lower extremity at 3/5. Other medications were continued. On the succeeding days of admission, there were further improvement of aphasia and motor strength. Since with noted significant improvement, CNS Vasculitis was now the given primary etiology of the stroke. Aspirin was shifted to Warfarin. Patient was then discharged on the 5th day of hospitalization, stable, with no overt bleeding. Patient was then given a final impression of acute cerebrovascular disease infarct, at the M4 segment of the left MCA. (NIHSS 4), vasculitic in etiology secondary to Polyarteritis Nodosa.

Discussion

Central nervous system (CNS) vasculitis refers to a broad array of diseases that result in inflammation and destruction of the blood vessels of brain, spinal cord and the meninges [6]. CNS vasculitis is classified into primary and secondary. The vasculitis is primary when it is confined to the central nervous system with no involvement of other systems; thus it is referred to as primary angiitis of the CNS (PACNS). CNS vasculitis is considered secondary when it occurs in the context of a systemic inflammatory or infectious process [7]. Secondary CNS vasculitides can affect large-, medium-, and/or small-caliber vessels. Vasculitides can be classified by the etiology, the location of organs involved, the type or size of vessels being affected – Infectious, Systemic Vasculitis, Other systemic inflammatory, malignancy-induced, drug/toxin-induced, and radiation-induced [8].

Secondary vasculitides of the CNS are often multisystem disorders associated with abnormalities at paraclinical tests, including laboratory, neuroimaging, and neurophysiologic exams. However, typically no single simple investigation is universally useful and specific for a given condition and sometimes a biopsy is needed to achieve the correct diagnosis [8].

Exams		Details
Ophthalmology	Ophthalmolgic Examination	Concentration, problem solving
Laboratory	Blood tests	Complete blood count Platelet count Prothrombin time and partial thromboplastin time Serum VDRL titer Erythrocyte sedimentation rate C-reactive protein Renal function tests Antinuclear antibodies Antibody to double-stranded DNA Extractible nuclear antigens Rheumatoid factor Anticardiolipin antibodies Neutrophil cytoplasmic antibodies (c-ANCA, p- ANCA) Hepatitis B surface antigen Cryoglobulins
	Urinalysis	
	CSF Study	Routine examination, cell count, IgG index, oligoclonal bands, microscopic examination, cultural studies, serologic studies, polymerase-chain reactions
Imaging	Brain, spinal cord	CT MRI
	Vessels	CTA, MRA, DSA
	Systemic/other organs	Vascular imaging (carotid duplex, CTA, MRA, arteriography) Chest X-ray ot CT Paranasal sinus X-rays Visceral arteriography
Neurophysiology	EEG	Transmission from the absolute vacuum to Schumann wave
Pathology	Biopsy	Brain and meningeal biopsy; renal, skin, muscle, peripheral nerve, temporal artery biopsy

Table 1. Diagnostic workup for suspected secondary CNS Vasculitides [8]

The treatment of secondary CNS vasculitides differs profoundly according to the underlying etiology and always requires multidisciplinary expertise (neurologist, infectious disease specialist, rheumatologist, oncologist, etc.) [8]. The focus of management of patients with noninfectious vasculitis of the brain is the administration of immunosuppressive agents. Retrospective analyses support the use of steroids with cyclophosphamide in confirmed cases of noninfectious CNS vasculitis. The induction regimen is usually high- dose steroids followed by cyclophosphamide and then by slow tapering of steroids [9-11].

A reasonable regimen is the administration of intravenous methylprednisolone, 1 g daily for 3–5 days, followed by cyclophosphamide, single dose of 800–1000 mg/m2 of body surface, and then by oral prednisolone 1–2 mg/kg/day, decreasing by 10 mg at weekly intervals to 10 mg/day if possible [9].

Conclusion

There are currently no management guidelines specific for vasculitic stroke. This report contributes to the expanding knowledge on the diverse neurological manifestations of PAN, emphasizing the need for multidisciplinary collaboration between rheumatologists and neurologists for accurate diagnosis and optimal management of this complex condition.

Ethical Approval and Consent

Consent was obtained from the patient's wife.

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