



## Cerebral Vasculitis:

Fariborz Khorvash, MD,.

Associate Professor of Neurology, Isfahan University of Medical Sciences, Iran.

\*Corresponding Author: Email: [fkhorvash@gmail.com](mailto:fkhorvash@gmail.com)

### Introduction:

Vasculitis is an inflammation systems may be involved of blood vessels due to various origins. Vessels of the peripheral and/or central nervous. Vasculitis of the CNS is rare and occurs in the context of systemic diseases or as primary angiitis of the CNS.

### Epidemiology:

The overall incidence of primary vasculitis is about 40/1,000,000 persons [excluding giant cell (temporal) arteritis, GCA]. Its incidence increases with age. The incidence of GCA is much higher (around 200/1,000,000 persons in the age group [50 years]).

### Clinical Presentation:

Clinical and pathological presentation in CNS vasculitis represents a wide spectrum. Among others, headache, cranial nerve affections, encephalopathy, seizures, psychosis, myelitis, stroke, intracranial haemorrhage and aseptic meningoencephalitis are described. Primary and secondary vasculitides leading more frequently to CNS manifestations are discussed.

### Primary and secondary Vasculitides:

Including Giant Cell (Temporal) Arteritis, Takayasu arteritis, Polyarteritis nodosa, Primary angiitis of the CNS, Wegener's granulomatosis, and Connective tissue diseases, such as systemic lupus erythematosus (SLE), scleroderma, rheumatoid arthritis, mixed connective disease and Sjögren syndrome, are systemic immune-mediated diseases that lead to multiple organ affections.

### Cerebral Vasculitis: Imaging and Differential Diagnosis:

Vasculitides represent a heterogeneous group of inflammatory diseases that affect blood vessel walls of varying calibers (inflammatory vasculopathy). Since the devastating symptoms of CNS vasculitis are at least partially reversible, early diagnosis and appropriate treatment are important. In order to establish a differential diagnosis clinical features, disease progression, age of onset, blood results, as well as CSF examinations have to be taken into consideration. Neuroimaging techniques, such as MRI and DSA, play a central role in the diagnosis and disease monitoring. The diagnostic protocol for cerebral vasculitis should include initial MRI to assess the degree of parenchymal damage and to detect vessel wall changes, in particular in large-vessel vasculitis. If the results are ambiguous or medium-sized arteries are affected (beyond the spatial resolution of MRI) DSA should follow. Small-vessel vasculitides entirely evade detection by vascular imaging and consequently require brain or leptomeningeal biopsy. Exclusion of differential diagnoses: Important differential diagnoses include RCVS, intracranial atherosclerosis Moyamoy disease, autoimmune encephalopathies and infectious disorder such as varicella zoster virus (VZV) vasculopathies or endocarditis.

### Therapy:

Vasculitis is a serious disease that is potentially fatal or leads to permanent disability and requires rapid institution of immunosuppressive treatment. Possible therapeutic options include glucocorticoids, cyclophosphamide, azathioprine, intravenous immunoglobulins and mycophenolate mofetil.

Key words: stroke, primary and secondary vasculitis, Differential diagnosis



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