

A CASE OF POSSIBLE PRIMARY CENTRAL NERVOUS SYSTEM VASCULITIS

Abstract

Background

Primary angiitis of the Central Nervous System is a distinct and rare inflammatory disorder involving the vasculature of the brain and spinal cord. It predominantly affects males. The usual age of presentation is 50s. Clinical presentation is varied and nonspecific and usually starts with headache. When the disease progresses there will be a multitude of neurologic phenomena and stroke like phenomena. Diagnosis is by the characteristic radiologic features in the Magnetic Resonance Image and Magnetic Resonance Angiograms and brain biopsy.

Clinical presentation

We report a case of 43 years old Sri Lankan female with a back ground history of diabetes. She presented with one-month history of episodes of diffuse headache, malaise and vertigo. The lady got admitted to the hospital with mouth deviation to right side and slurring of speech over five days' duration. The condition did show an accelerated progression where she had reduced level of consciousness and multiple cranial nerve palsies. Magnetic Resonance Image(MRI) brain and Angiogram revealed irregular beaded appearance of the cerebral vasculature with multiple infarcts. She did not respond to the prompt treatment with high dose steroids and cyclophosphamide.

Conclusion

After complete evaluation of an acquired neurologic deficit, if there are no other available differentials to consider for the etiology and there are no associated systemic features and if the angiographic features are characteristic with associated vessel irregularity, luminal narrowing and the beaded appearance, it is always prudent to consider the diagnosis of Central Nervous System(CNS)

vasculitis and start urgent therapy because there are subtypes with favorable outcome.

Key words: CNS Vasculitis, Angiogram, Brain biopsy