

Primary Angiitis of CNS: A Diagnostic Dilemma

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Introduction

PACNS is not a rare but uncommon case that we encounter. Early differentiation from Reversible Cerebral Vasoconstriction Syndrome (RCVS) is important and has profound impact on clinical outcome.

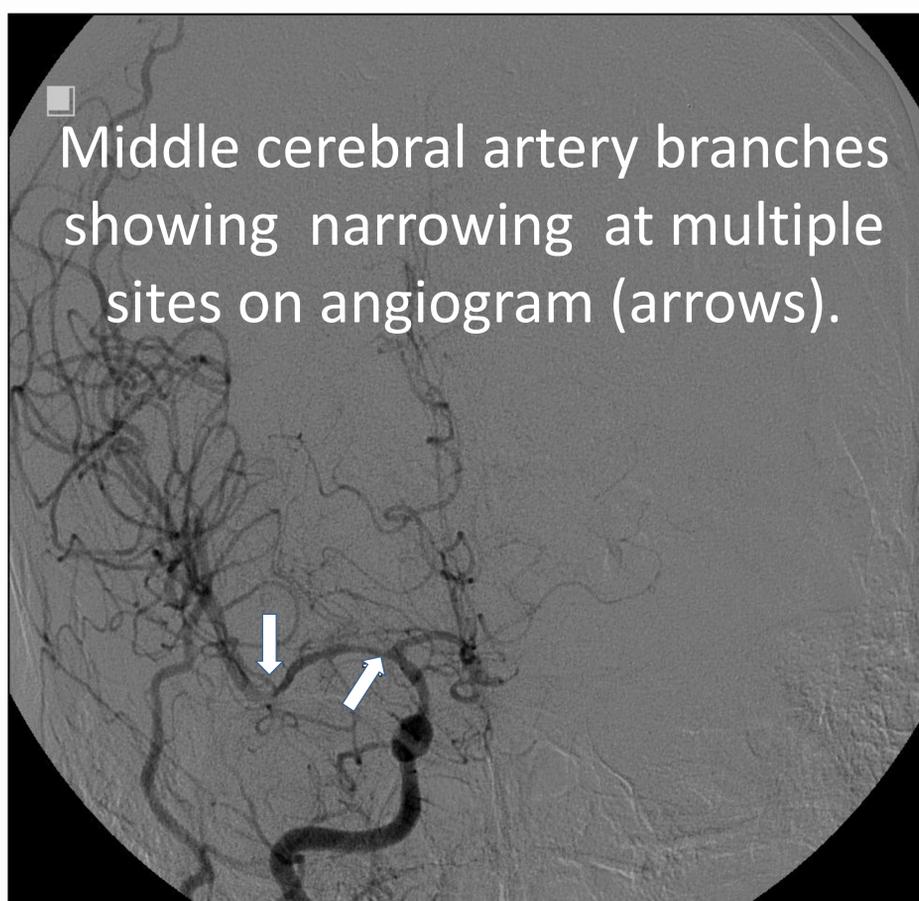
Case

A 42-year-old female, known to have migraine headaches, presented with occipital headaches, neck stiffness, and lower extremity weakness of few hours duration. Physical examination showed motor strength 4/5 in all 4 extremities.

CSF analysis showed: protein 51mg/dL, glucose 55mg/dL, WBC 9/mm³, lymphocytes 42%, neutrophils 47%. HIV, Lyme's disease, tularemia, syphilis, *Bartonella henselae*, mycoplasma, *Coxiella burnetii*, West Nile virus serologies, and ANA, C-ANCA, P-ANCA were negative.

MRI showed ischemia in the right paracentral lobule. MRA revealed multiple short segment areas of stenosis with normal intervening segments throughout bilateral middle, anterior, and posterior cerebral artery, suggestive of vasculopathy. Findings were verified by arteriogram and biopsy demonstrated mild gliosis. Impression of PACNS was made and the condition improved with IV methylprednisolone.

Variables	RCVS	PACNS
Sex	Female predominance	No sex predilection
Onset	Seconds to minutes	Sub-acute
Headache	Acute throbbing	Dull, insidious
CSF	Normal-near normal	Abnormal > 95%
CT/MRI	Normal in majority or small watershed infarcts.	Abnormal > 90%, Small infarctions
Neuro-vascular imaging	Diffuse stenosis and dilatation or arteries	Frequently normal or indistinguishable from RCVS



Discussion

RCVS is the most important clinical mimic of PACNS.

PACNS is fatal as compared to the more benign and reversible course of RCVS. Misdiagnosing PACNS patients with RCVS can prove fatal.

Early administration of immunosuppressive agents has significant impact on the prognosis of PACNS

References

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