

INTRODUCTION:

Isolated vasculitis of the central nervous system (CNS) or primary angiitis of the CNS (PACNS) is an extremely rare condition, with incidence of 2.4 per million, in the 4th to 5th decade of life, predominantly male, difficult to diagnose, severe outcome and potentially fatal. Manifestations are non-specific, such as headache, cognitive dysfunction and only 25-30% of patients have neurologic deficit.

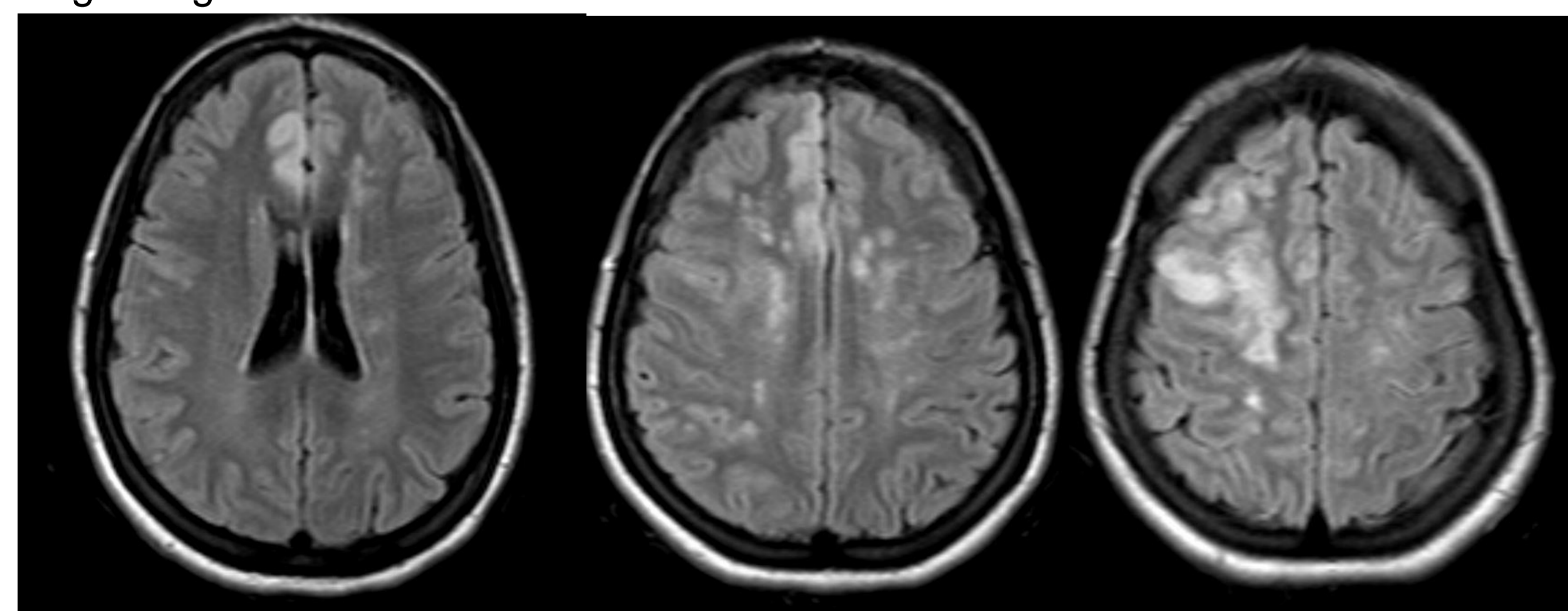
CASE REPORT:

CAB, female, 28 years old, hypertensive, presented acute disproportionate left hemiparesis, crural prevalence and severe global aphasia associated with headache. Onset alert, but confused, double hemiparesis, worse on the left, bone-tendon reflexes globally live without sensory or eye movement changes. MRI showed sign of pearl necklace (alternating areas of stenosis and dilation) in the segments M1, P1, P2, P3 and A1 right. In laboratory research, infectious, rheumatologic, hematologic and endocrine causes was excluded.

CSF analysis without abnormalities. Undergone pulse therapy with methylprednisolone for 3 days with partial improvement of strength and aphasia. Maintenance treatment with azathioprine.

PCR: 0,49	c-ANCA: Neg	Anticardiolipina: Neg	HBV, HCV: Neg
VHS: 24	TSH: 3,31	Anticoagulante lupico: Neg	CMV, toxo: Neg
FAN: Neg	T4L: 1,47	Homocisteina: 11,8	HSV IgM: Neg
FR: Neg	Anti-TPO: neg	HIV, HTLV: Neg	HSV IgG: reag
p-ANCA: Neg	TRAB: neg	VDRL: Neg	

Neg = negative

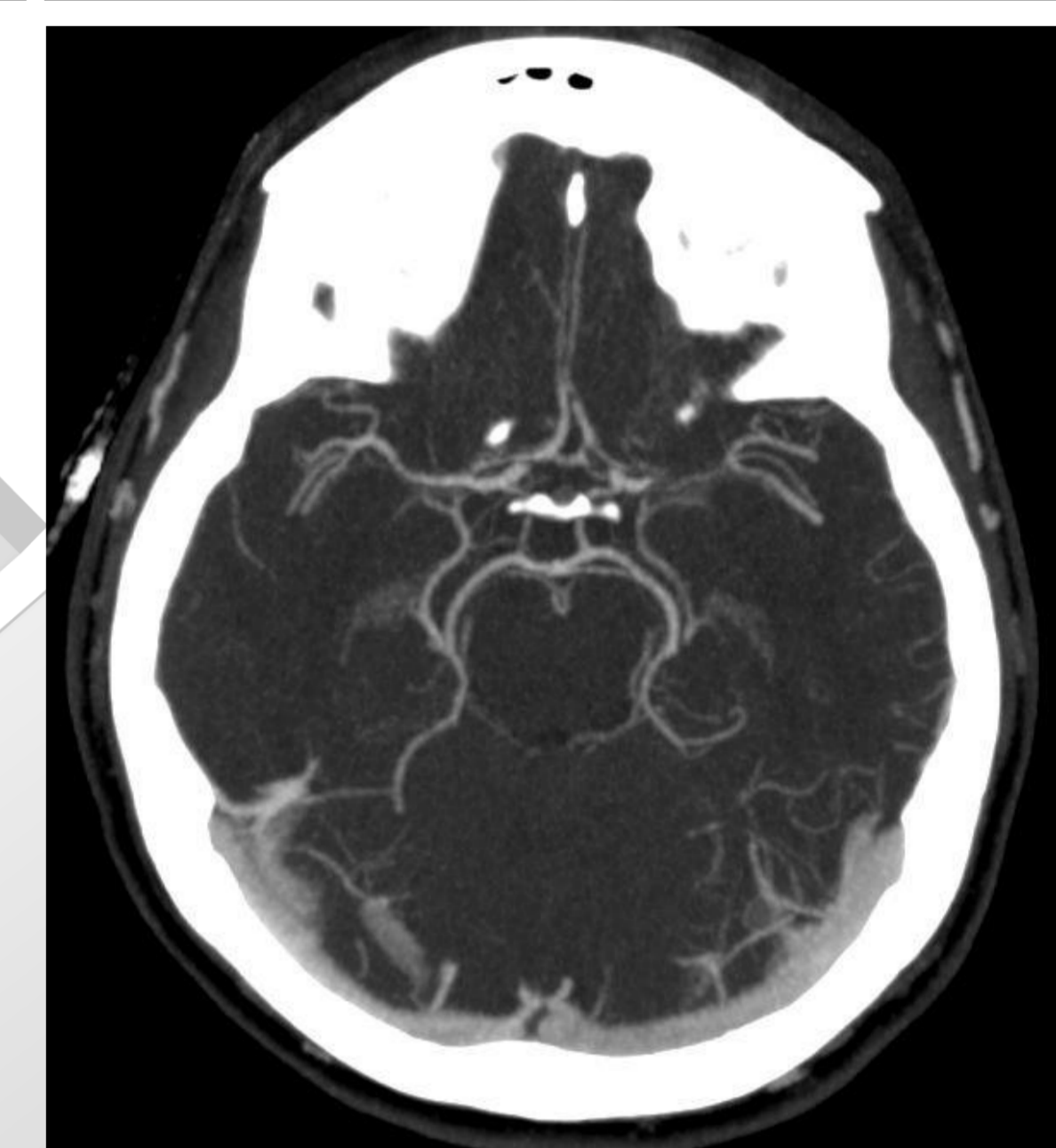


Brain MRI: FLAIR shows cortical and subcortical areas of ischemic injury bilateral but mainly in the right hemisphere



Above: MRI shows alternating areas of arterial stenosis and segmentar dilatations

Right: TC angiography shows the same aspect of stenosis and dilatation of brain arterial vessels



DISCUSSION:

This case differs from the literature, because reports a patient in the 3rd decade. As there is no typical clinical presentation, it is interesting to note that constitutional symptoms can be found, such as headache described by the patient. Other symptoms may also be present such as fever and focal deficits, topographical related injuries. In the research, 90% of patients present abnormal CSF with increased protein or lymphocytic pleocytosis, but differential diagnosis as Syphilis, Hepatitis, Varicella Zoster, systemic vasculitis such as polyarteritis nodosa and granulomatous diseases may have similar findings. Inflammatory activity markers help identify the silent systemic involvement. The MRI has a sensitivity 90-100% and may show changes both white or gray matter as well as infarcts in different territories. The classic "string of beads" has low sensitivity and specificity, and can appear in vasospasm, reversible cerebral vasoconstriction syndrome (RCVS), intravascular lymphoma and CNS infection. A biopsy is still the gold standard, but 35% are inconclusive.

CONCLUSION:

PACNS is a rare condition of unknown etiology, variable clinical manifestation and non-specific neuroimaging. The difficult diagnosis depends on high clinical suspicion, because treatment is important in the prognosis.

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