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# A case of CPACNS (Childhood Primary Angiitis of the Central Nervous System) with poor correlation of severity between clinical picture and brain imaging

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## Introduction

Primary Angiitis of the Central Nervous System (PACNS) is a rare form of vasculitis of unknown cause and is less often in children than adults. The diagnosis of PACNS is often difficult. There are neither specific clinical features nor blood or imaging investigations that can confirm the diagnosis. Pathology reveals inflammation within the wall of Central Nervous System (CNS) blood vessels leading to occlusion and infarction [1]. No standardized treatment protocols exist, and evidence is limited to open-label cohort studies and case reports [2].

## Clinical image description

A 15-year-old female with unremarkable medical background presented with headaches and episodes of mild dizziness and weakness during the episodes, 2-3 times per week, starting six months ago. Her neurologic examination was normal. Brain MRI revealed multiple T2 and T2 - Fluid-Attenuated Inversion Recovery (FLAIR) hyperintensities in both cerebral hemispheres, located mainly in the corona radiata, centrum semioval, as well as in the internal capsule and the subcortical white matter. Lesions were also found in the basal ganglia, thalami, brainstem, and cerebellum (Figure 1A,1B). Findings were reminiscent of demyelinating lesions. MR-Angiopraphy and spine MRI were normal.

The patient underwent an extensive laboratory investigation for a wide panel of infectious as well as systemic immune diseases, metabolic and demyelination disorders (anti-AQP4, anti-MOG Abs) that came back negative. CSF examination revealed 25 cells/μl, 95% lymphocytes, high protein (prot: 113 mg/dl, glucose 50 mg/dl) and oligoclonal band type 4, consistent with systemic immune reaction. Due to the worsening of the symptoms, a second MRI was performed three weeks later including vessel wall imaging (Black Blood T1-weigthed sequences with contrast). In addition to the slight increase of the parenchymal T2 hyperintensities, multiple foci of parenchymal

and leptomeningeal enhancement were seen on post contrast images. Moreover, arterial wall enhancement was noticed in peripheral branches of the medial cerebral arteries, suggestive of vessel wall inflammation (Figures 1C-E). In order to exclude malignancy, a LP was repeated. Protein showed mild elevation (133 mg/dl) and cells were again 25 cells/µl, 95% lymphocytes. A flow cytometry and bone marrow aspiration and biopsy didn't reveal any pathological findings. An extended investigation for systemic immune diseases was again performed. Since primary systemic vasculitic syndrome and other alternative possible diagnoses were excluded, MRI evidence of vasculitis led to PACNS diagnosis. The teenager was treated with high doses of methylprednisolone (1 gr/day) for 5 days followed by a 12-week oral tapering, aspirin and vitamin D and showed immediate clinical improvement. At the 3-month follow-up she had no symptoms, while partial resolution of both T2 hyperintensities and abnormal contrast enhancement was seen on MRI (Figures 1G-H). At the time, her treatment included oral prednisone (5 mg/day) and aspirin and it was continued until her next follow-up MRI.

In the six-month follow-up, even though there was no clinical difference, brain MRI showed new areas of leptomeningeal enhancement, combined with few foci of parenchymal enhancement,, indicating possible remission (Figure 1H). A broad workup was repeated and enriched with genetic testing for CNS HLH (NGS for specific gene panel) in order to exclude all other possible diagnoses. Again, all results came back negative. Consequently, the patient was treated with methylprednisolone pulses for 3 days (and then oral tapering) followed by Rituximab, (375 mg/m$^2$), a monoclonal antibody which targets CD20 B cells, as an adjunctive therapy. The follow-up MRI after six months revealed lesion resolution and no abnormal contrast enhancement. Until today the patient has not shown any clinical symptoms. (CD20 B cells count is zero). Our patient now follows a 2-year plan with Rituximab infusions every six months while she continues her antithrombotic treatment with aspirin.

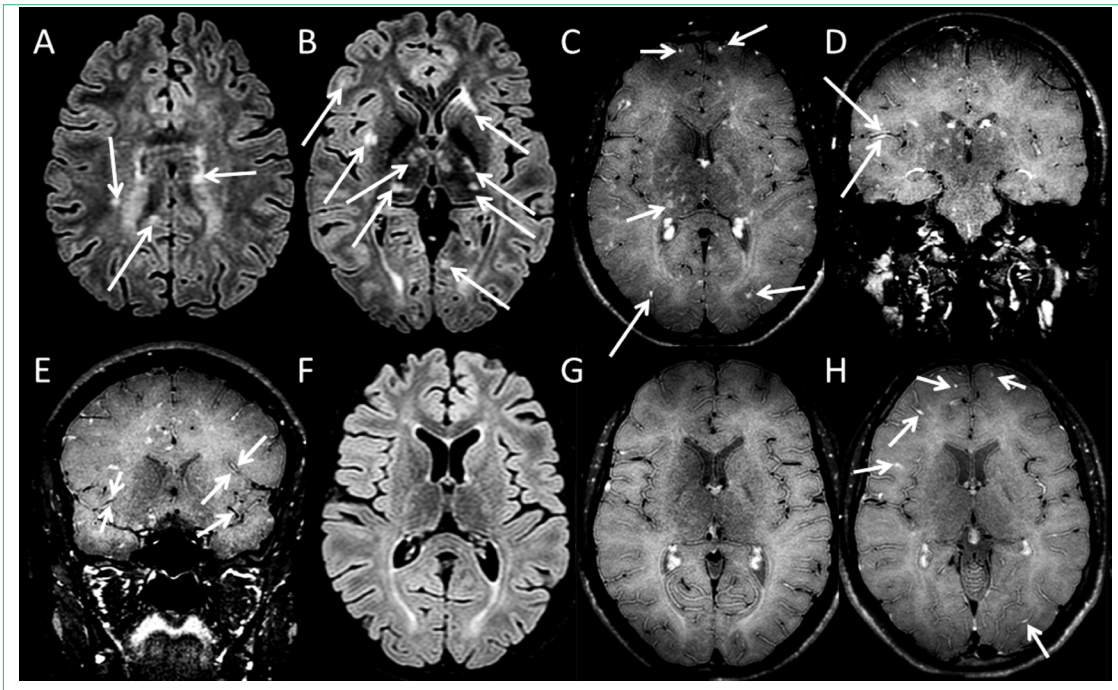


**Figure 1:** Serial MRI findings. Axial FLAIR images on initial MRI **(A,B)** reveal multiple T2-FLAIR hyperintensities (arrows A,B) in the white matter, the basal ganglia and the thalami. T1 black-blood post contrast administration images after 3 weeks on axial **(C)** and coronal **(D,E)** planes show multiple foci of leptomeningeal and parenchymal enhancement (arrows C), as well as vessel wall enhancement of peripheral branches of the medial cerebral arteries (arrows D, E). Follow up MRI 3 months later show partial lesion resolution both on axial FLAIR **(F)** and axial T1 black blood post contrast **(G)** sequences. T1 black blood with contrast image (H) from the MRI performed six months later reveals new leptomeningeal and parenchymal enhancing foci (arrows H).

## Discussion

Primary CNS vasculitis remains a difficult-to-reach diagnosis. Laboratory testing is not specific and brain biopsy is not a risk-free procedure. According to the latest proposed criteria [1], it is important to exclude other possible diagnoses and to have histopathologic evidence of vasculitis. The degree of severity might not be indicated by the patient's symptoms alone and the imaging follow-up is crucial for the early detection of possible remission. This case, as well as others described [4], shows that anti-inflammatory immunomodulatory maintenance therapy is needed in order to optimize survival and prevent permanent brain injury.

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