
Neuroimaging Highlight

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Primary Central Nervous System Angiitis in a 10-year-old Girl

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Primary angiitis of central nervous system (PACNS) is well reported, however its occurrence in the pediatric population is infrequent.^{1,2} We describe the clinical, neuroimaging and histopathological features of PACNS in a young girl.

A ten year-old, previously healthy girl presented with a three-week history of progressive left hemiparesis and facial weakness. Other findings included left hemineglect, impaired

concentration and memory. She had no evidence of systemic disease. Head computed tomography (CT), with and without contrast, revealed no abnormalities. Magnetic resonance imaging (MRI) demonstrated multifocal, bilateral signal abnormalities within basal ganglia, thalami, right frontal cortex and subcortical white matter on T2 and FLAIR images. However, these areas did not show restricted diffusion on diffusion weighted

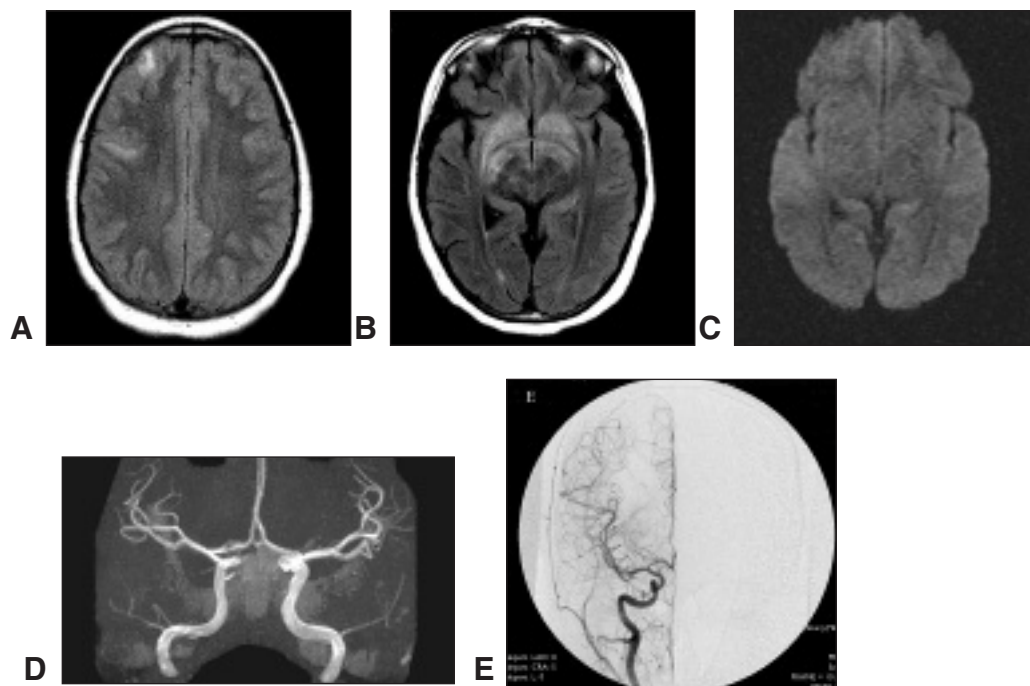


Figure 1: Magnetic Resonance Imaging of the ten year-old girl with PACNS. (A, B) FLAIR axial unenhanced images reveal asymmetric abnormal signal in the basal ganglia, right frontal cortex and subcortical white matter. (C) Diffusion Weighted Imaging shows normal diffusion in the areas of abnormal signal. (D, E) Magnetic resonance angiography and conventional catheter angiography with digital subtraction (only right selective angiogram shown as an example) did not reveal any vascular stenosis, dilatation or irregularity.

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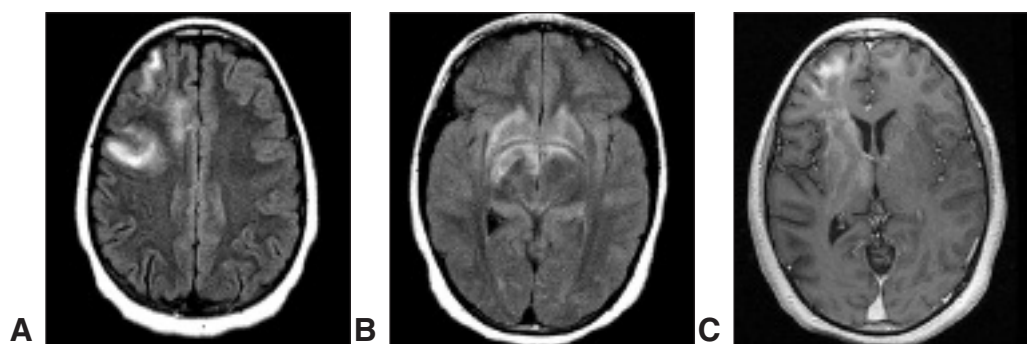


Figure 2: Follow-up MRI three weeks later. (A, B) FLAIR axial images show worsening of signal abnormalities noted in Figure 1. (C) Gadolinium enhanced T1 images shows heterogeneous enhancement in the basal ganglia and right frontal lobe. Rounded punctate areas of enhancement suggest perivascular inflammation.

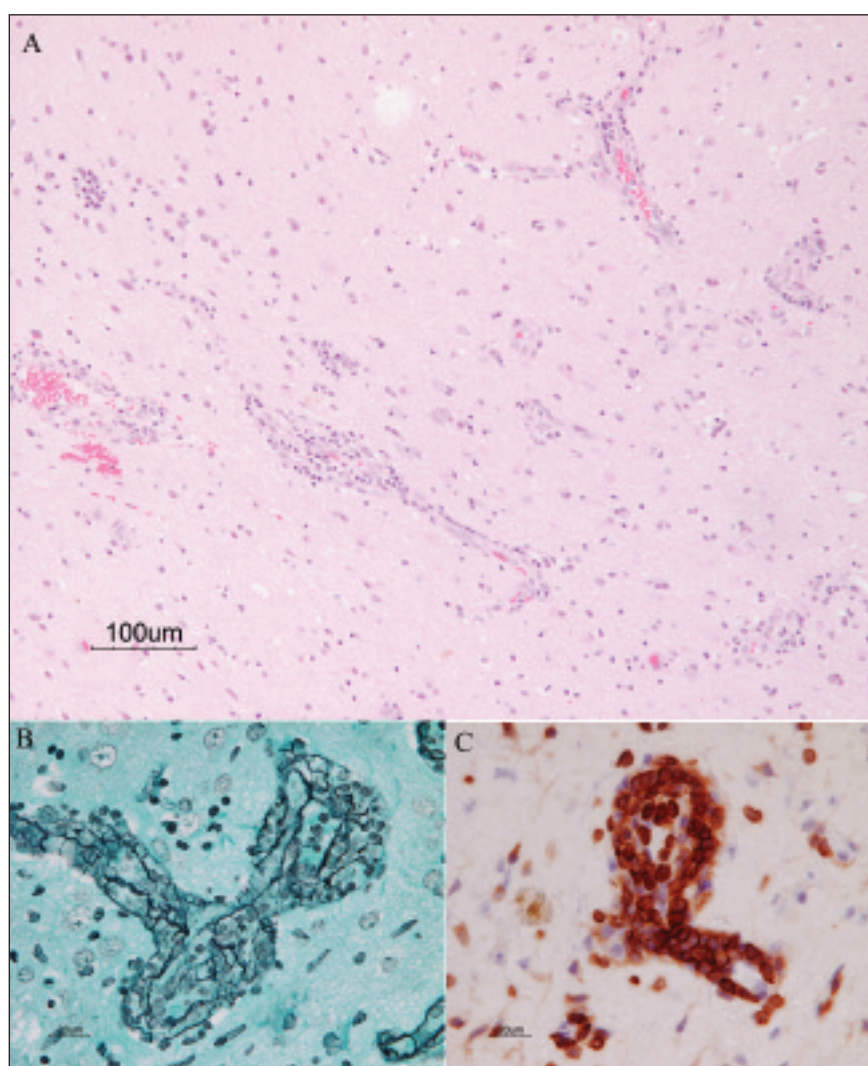


Figure 3: Histopathology of the right frontal lobe brain biopsy specimen. (A) Mural mononuclear infiltrates are seen in multiple small and medium-sized blood vessels at low power (H&E X100). (B) Reticulin staining demonstrating expansion of the vessel wall by lymphocytes (X400). (C) CD45 immunohistochemical staining demonstrating intramural lymphocytes (X400).

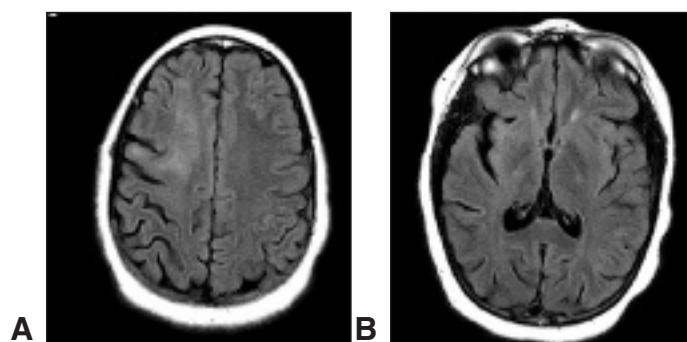


Figure 4: Post treatment follow-up MRI. (A, B) FLAIR axial unenhanced images, three months later, shows improvement of signal abnormalities noted on Figures 1 and 2 and parenchymal volume loss with increased extra-axial Cerebrospinal Fluid spaces and sulcal prominence.

imaging (DWI). Cerebral angiography revealed normal intracranial vessels (Figure 1). Neuroimaging, three weeks later, showed progression of these abnormalities. The areas of abnormal signal showed heterogeneous and nodular enhancement on gadolinium enhanced T1 weighted images (Figure 2). Rheumatologic, metabolic, prothrombotic and cerebral spinal fluid investigations were normal.

Right frontal lobe biopsy demonstrated mixed inflammatory mural infiltrates including lymphocytes, plasma cells and occasional histiocytes in small cortical and white matter blood vessels and leptomeninges. Vessel wall fibrosis and necrosis with brain tissue gliosis and infarction were noted. Macrophages were observed perivascularly and in infarcted areas. There was no evidence of giant cells or granulomas (Figure 3). Based on histopathology and lack of systemic findings, a diagnosis of PACNS was made. Follow-up brain MRI, three months after treatment initiation, showed significant improvement of previously noted increased signal intensities (Figure 4). At six months, physical examination was significantly improved, but concentration and memory problems persisted.

Diagnosis of PACNS is based on a combination of clinical and characteristic imaging features, however, brain biopsy is required for confirmation.³ Abnormal blood and serology tests seen in connective tissue disorders are not detected in PACNS.⁴ Cerebrospinal fluid abnormalities, including elevated protein and lymphocytic pleocytosis, are observed in 80–90%.⁵

Brain MRI appears to be more sensitive than CT and angiography to demonstrate abnormalities in PACNS.^{1,5-8} However, MRI findings are nonspecific and may simply show changes suggestive of ischemic and inflammatory processes, including multifocal, bilateral, frequently supratentorial signal abnormalities with and without leptomeningeal enhancement. Abnormalities are best appreciated on T2 and FLAIR images.^{4,7} Previous reports on DWI in PACNS are heterogeneous suggesting co-existence of cytotoxic and vasogenic edema.⁹ Normal diffusion in our case suggested that the abnormalities

were more than seven to ten days old. The enhancing nodular pattern suggested perivascular inflammation. This along with extension of the abnormality in multiple vascular territories and progression with lack of diffusion restriction helped to differentiate this process from usual arterial ischemic infarction. There are no pathognomonic findings for PACNS with cerebral angiography and angiogram abnormalities may not be seen if disease is isolated to small vessels.¹⁰

In conclusion, progressive multi-focal brain lesions on neuroimaging in children presenting with acquired focal neurological deficits, should alert the clinician to the possibility of PACNS. Prompt diagnosis is important for early intervention in such children.

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