# Primary angiitis of the central nervous system

Childhood primary angiitis of the central nervous system (PACNS) is a rare condition that most often affects small vessels (SV), is nearly exclusively lymphocytic and is devoid of a vessel necrosis.

## Diagnosis

Diagnosis of Childhood Primary angiitis of the central nervous system is challenging. Gilani and Kleinschmidt-DeMasters noted a possible histological overlap with Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) on biopsy, prompting a 10-year retrospective review of the experience.

Database search for brain biopsy cases, age <18 years, performed for an acquired neurological deficit with suspicion of vasculitis, with histological evidence of lymphocytic small-vessel inflammation.

They identified 7 patients; 2/7 were serum-positive for anti-MOG antibodies and 1/7 for anti-NMDA antibodies. The remaining 4/7 proved to be idiopathic lymphocytic vasculitis/cSV-PACNS. All 7 showed overlapping features of lymphocytes permeating parenchymal SV walls, vessel wall distortion without fibrinoid necrosis, and absence of microglial clusters or intravascular thrombi. Tissue infarction was confined to a single case of idiopathic lymphocytic vasculitis. Although demyelination was diligently sought, only subtle demyelination was identified in the 2 MOGAD cases and absent in the remainder.

There is considerable histological overlap between cSV-PACNS and at least some cases of MOGAD or anti-NMDA-encephalitis; at diagnosis, the differential should include cSV-PACNS but correct classification requires post-biopsy serological testing [1)].

1)
Gilani A, Kleinschmidt-DeMasters BK. Childhood Small-Vessel Primary Angiitis of the Central Nervous System: Overlap With MOG-Associated Disease. Pediatr Dev Pathol. 2022 Nov 15:10935266221121445. doi: 10.1177/10935266221121445. Epub ahead of print. PMID: 36377607.