A Case of Refractory Primary Angiitis of the Central Nervous System.

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INTRODUCTION

- Primary angiitis of the CNS (PACNS) is a rare vasculitis that is particularly challenging to diagnose.
- It should be considered in the setting of recurrent strokes, continued cognitive decline, and recurrent or progressive focal neurological deficits.
- It is essentially a diagnosis of exclusion after ruling out systemic involvement with confirmatory cerebrovascular imaging or histopathological evidence of vasculitis.[1]
- Current treatment recommendations include high dose corticosteroids and cyclophosphamide. Limited data supports the use of rituximab in refractory cases of PACNS.[2,3]

CASE

- Our patient is a 56-year-old male with recurrent multifocal cerebral infarcts, encephalopathy and seizures secondary to PACNS diagnosed a year prior with cerebral arteriogram.
- He presented with worsening right sided weakness, expressive aphasia, and urinary incontinence.
- MRI upon admission revealed new areas of acute/subacute infarctions in the basal ganglia and corona radiata.
- Lumbar puncture was unremarkable. Other labs confirmed no underlying systemic vasculitis with negative ANA and ANCA profiles. Renal biopsy showed diabetic nephropathy without signs of connective tissue disease.
- Cerebral arteriogram was repeated, showing multiple small peripheral arterial narrowing and beading consistent with a diagnosis of PACNS.
- Despite increasing IV corticosteroids and treatment with Cytoxan for over 6 months, our patient continued to demonstrate decline.
- During the hospital course, Rituxan infusions were initiated and patient began to show clinical improvement over the following 6 weeks. He unfortunately regressed after a few months of treatment.

DISCUSSION

- Most patients with PACNS demonstrate improvement in symptoms and on imaging when trialed on steroids with or without Cytoxan.
- No studies suggest an optimal treatment regimen or duration.
- Three case reports in the current literature describe clinical and radiographic improvement in adults with PACNS after treatment with rituximab.[2,3]
- In one case where the diagnosis was made by angiographic findings and an abnormal CSF, rituximab was given as first-line therapy.[2]
- In the other two cases, the diagnosis of PACNS was histopathological with evidence of a lymphocytic vasculitic process involving the brain and spinal cord. Rituximab was used as first-line in one and after failing Cytoxan in the other.[2]
- Our patient was deemed to have failed Cytoxan and unfortunately Rituxan.
- This case shows the difficulty of diagnosing PACNS. The goal standard for the diagnosis of PACNS is a brain biopsy.
- Only a positive biopsy can ascertain the diagnosis which we did not have. Staining, immunohistochemistry studies, and immunophenotyping are needed to rule out other mimickers.
- Refractory cases are rare and require reconsidering the diagnosis.

References:

1. Birnbaum J, Hellmann DB. Primary angiitis of the central nervous system. *Arch Neurol.* 2009;66(6):704-9.
2. Salvarani C, Brown RD, Huston J, Morris JM, Hunder GG. Treatment of primary CNS vasculitis with rituximab: case report. *Neurology.* 2014;82(14):1287-8.
3. De boysson H, Arquizan C, Guillevin L, Pagnoux C. Rituximab for primary angiitis of the central nervous system: report of 2 patients from the French COVAC cohort and review of the literature. *J Rheumatol.* 2013;40(12):2102-3.