P4 6-039

Unilateral Primary CNS Vasculitis in a Child Associated with Increased ICP and Treated with Maximal Medical Therapy and Decompressive Hemicraniectomy

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Objective

- We present a case of a 6-year-old boy with new-onset seizures and altered mental status associated with multifocal right hemispheric lesions resulting from primary CNS vasculitis.
- Unique features of the case including unilateral involvement and aggressive medical and surgical management are discussed.

Background

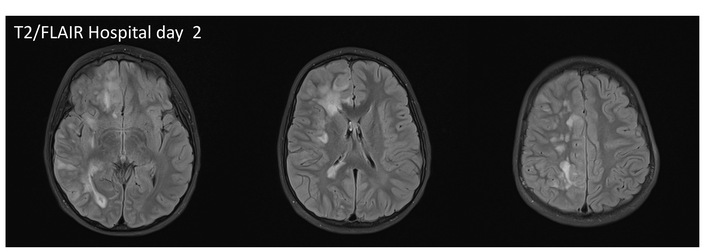
- Primary CNS vasculitis or primary angiitis of the CNS (PACNS) is a rare vascular inflammatory brain disease, often associated with variable neurological manifestations and a lengthy diagnostic evaluation.
- Secondary causes of CNS vasculitis and other inflammatory brain disorders must be excluded and a definite diagnosis relies on tissue biopsy.
- Some forms of PACNS can be rapidly progressive and life-threatening, further complicating the diagnostic and treatment process.

Methods

- Case report featuring clinical presentation, laboratory, pathology, and neuroimaging results, and discussion of diagnostic/treatment decision-making.

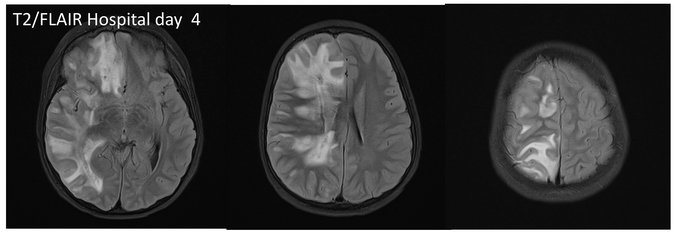
Results

- A 6-year-old previously healthy boy with intermittent headaches developed focal seizures and altered mentation.
- CSF showed marked pleocytosis (WBC 139 cells/mm3) and elevated protein (299 mg/dL). IgG index 0.93, 0 oligoclonal bands, AQP4, MOG negative.
- MRI brain demonstrated multifocal areas of T2 hyperintense signal, restricted diffusion, and microhemorrhage involving the deep and superficial white matter of the right hemisphere.



Results

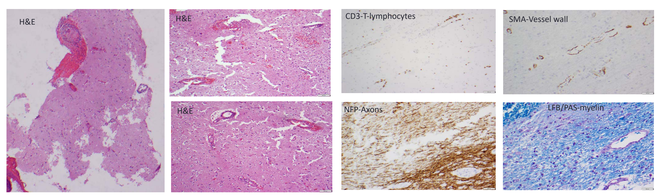
- Despite maximal osmotic, sedative, and immune-directed therapies, he exhibited worsening cerebral edema and midline shift with persistently elevated intracranial pressures.



Hospital course

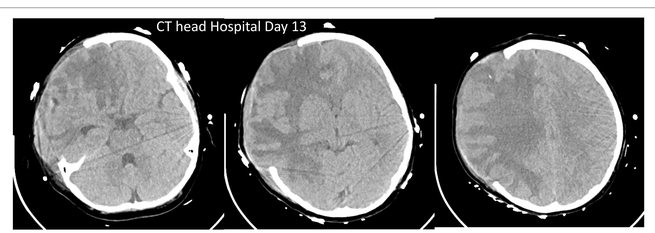
Day 2 – IV methylprednisolone started
Day 4 – Codman ICP monitor placed
Day 5– Pentobarbital infusion , hypertonic saline with goal Na 160s, PLEX started
Day 10 – Had ICP spikes – went for decompressive right frontotemporoparietal hemicraniectomy with brain biopsy
Day 12– IV cyclophosphamide
Day 37 – Cranioplasty
Day 44 – Transfer to inpatient rehab

- He underwent decompressive hemicraniectomy and open brain biopsy showing perivascular lymphocytic cuffing and lymphocytes in the vessel walls, interpreted as vasculitis with white matter infarction and hemorrhage, consistent with PACNS.



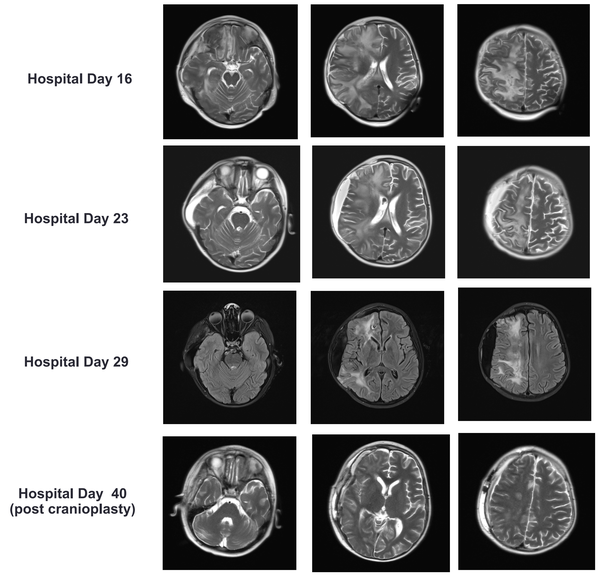
Biopsy showed parenchymal loss/pallor associated with numerous macrophages and hemorrhage, perivascular lymphocytic cuffing with lymphocytes in the vessel wall and fibrinoid necrosis of rare vessels, in the cortical parenchyma and leptomeninges

The predominant lymphocytes are CD 3 positive T-lymphocytes with a few CD 20 positive B-lymphocytes; Neurofilament (NFP) and LFB/PAS highlight the axonal and myelin breakdown in the areas of parenchymal loss/infarction and SMA highlights the disruption in the smooth muscle wall of many of the small vessels



Results

- He was treated with cyclophosphamide with gradual improvement in cerebral edema.
- He underwent cranioplasty four weeks after hemicraniectomy (Hospital Day 37).



- His left hemiparesis improved significantly over one month in inpatient rehabilitation and he regained the ability to ambulate independently.

Conclusions / Discussion

- Our case demonstrates an unusual presentation of PACNS in a child given unilateral involvement and fulminant course.
- Hemicraniectomy should be considered in patients with medically refractory increased intracranial pressure, as it can cause irreversible morbidity and mortality.
- Management of this child involved a multidisciplinary team of providers and complex diagnostic and therapeutic decision-making, ultimately resulting in a favorable outcome.

Disclosures

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Drs. Miles, Rajaram, and Whittemore report no relevant disclosures.