Chronic Lymphocytic Infiltration With Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS): A Case Report in a Teenager

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Atypical Knee Hyperextension Injury
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Mentor: Jason Meredith
Program: Family Medicine
Type: Case Report
Background: Subchondral knee fractures are typically non-traumatic, over-use injuries that occur in the middle aged or elderly. Adolescents rarely suffer subchondral fractures of the knee. These fractures are usually managed conservatively, and typically require several months of recovery before returning to baseline.

Case: A 14-year-old female volleyball player presented for evaluation of left knee pain. She injured her knee during agility drills the day prior, feeling a pop in her knee and subsequent instability. She initially tolerated weight bearing but was unable the following day. On exam, she had a moderate effusion, limited range of motion (ROM) secondary to pain, and tenderness to palpation over lateral proximal tibia and lateral femoral condyle. Plain films were obtained and unremarkable; however, MRI demonstrated subchondral impaction fractures of the anterolateral tibia and lateral femoral condyle.

She was made non-weightbearing, placed in a compression sleeve, and advised to transition slowly to weight bearing as tolerated. At 6-week follow up, she remained NWB secondary to pain. Her ROM remained significantly restricted. She was made toe-touch weight bearing with goal of transitioning to weight bearing as tolerated in 7-10 days. She was referred to physical therapy and made slow improvements in strength and ROM over 2.5 months. She returned to volleyball 5 months post injury. Consent was obtained to use this case for educational purposes.

Conclusion: This case represents a rare presentation of a non-contact, knee hyperextension injury causing subchondral compression fracture without other associated knee injury in an adolescent. While rare in the young, management and expectant recovery was like that seen in older adults.

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Chronic Lymphocytic Infiltration With Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS): A Case Report in a Teenager
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Mentor: Rana K. Zabad
Program: Multiple Sclerosis/Neuroimmunology
Type: Case Report
Background: Chronic lymphocytic infiltration with pontine perivascular enhancement responsive to steroids (CLIPPERS) is a rare central nervous system inflammatory disorder first described in 2010. The mean age of onset is 50-years. Very few cases were reported in youth. Its cardinal features include punctate gadolinium enhancement 'peppering' the pons, adjacent hindbrain, and spine on magnetic resonance imaging (MRI). Brain biopsy demonstrates T-cell predominant infiltrate. CLIPPERS is a steroid-dependent condition requiring long-term immunosuppression with methotrexate, hydroxychloroquine, or cyclophosphamide. Recently, treatment with rituximab resulted in disease remission. We describe a pediatric case of CLIPPERS with planned rituximab treatment.
Case: A 17-year-old adolescent male presented with subacute diplopia and gait ataxia. He has a history of prematurity, attention-deficit, and hyperactive disorder (ADHD) and several congenital anomalies, and a family history of autoimmune diseases. Cerebrospinal fluid analysis showed a protein of 62 (range 15-45 mg/dl) and two oligoclonal bands. The brain and spine MRI findings and the steroid-dependent course suggested CLIPPERS with disease exacerbation upon corticosteroid withdrawal. After extensive workup, the diagnosis was made, excluding other etiologies such as infectious, neoplastic, and autoimmune conditions. We plan to treat him with rituximab and follow him up longitudinally. We obtained patient consent to use this case for educational purposes.

Conclusion: CLIPPERS presents distinctive clinical and MRI findings. Induction of remission by steroids should be followed by immunosuppression. B-cell depletion might effectively treat this condition by altering T cells' subset composition, activation, and function. Longitudinal follow-up is recommended for the possibility of an alternative diagnosis. ■

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Negative Is Not Always Negative: Improving Outcomes in Scalp Negative Seizures Using Intracranial EEG
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Mentor: Corey Georgesen
Program: Dermatology & Neurology
Type: Case Report

Background: Refractory seizures sometimes arise from deeper foci within the brain. When difficult to detect on scalp EEG, chances of successful epilepsy surgery are reduced. Two patients had scalp EEG negative seizures, got intracranial EEG and did well with responsive neurostimulation. Patient consent was obtained to utilize these cases for educational purposes.

Case: Patient I is a 29 yo F, with prior right temporal lobectomy, s/p VNS and 5-year seizure freedom before recurrence. MRI and PET showed signs of prior surgery. Scalp EEG and MEG were unremarkable. Neuropsychological testing showed diminished core verbal function and memory. Wada testing revealed left dominance for language and memory. Stereotactic EEG captured focal impaired awareness seizures and focal aware seizures with early involvement of the right posterior cingulate (RPC) and right posterior insular (RPI) regions. Brain mapping/cortical stimulation revealed motor function in RPC and sensory in RPI regions precluding resection/ laser ablation. RNS implantation in the RPC and RPI regions achieved seizure freedom 4 months after implantation.

Patient II is a 33 yo F, who would wake up, laugh/curse, vocalize and show left (focal) predominant hyper motor movements progressing to tonic-clonic convulsion. MRI brain showed a venous angioma. PET revealed decreased uptake in right posterior parasagittal frontal and right inferior parasagittal frontal regions. MEG was unremarkable. Neuropsychological testing showed weak bi-frontotemporal systems. IAP/Wada showed left dominance for language and memory. SEEG captured hyper motor seizures with early involvement of right orbito frontal (ROF) with spread to right hippocampal (RH) regions. RNS implantation in the ROF and RH regions achieved 30% seizure reduction 3 months post implantation.

Conclusion: Scalp EEG negative seizures remain challenging to treat. SEEG delineates seizure network and localizes the onset zone. This understanding will better help treat patients using neuromodulation or targeted therapies like ablation. ■

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Use of Carotid Embolic Protection Devices for Left Atrial Myxoma Resection in a Pediatric Patient
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Mentor: Jeffrey Delaney
Program: Pediatric Cardiology
Type: Case Report

Background: Carotid embolic protection devices are used in adults for manipulation of atherosclerotic lesions with concern for embolization. This is not a routine practice done in pediatrics as thromboembolic sequela is not a commonly encountered entity.

Case: A 14-year-old girl presented with left foot numbness and pallor with CT angiography showing a left femoral and popliteal arterial thrombus. She underwent thrombectomy of the left popliteal artery with restoration of flow. Echocardiogram revealed a large heterogeneous mobile mass in the left atrial appendage measuring 2 x 2 cm consistent with a myxoma requiring resection.

Prior to sternotomy, a 6Fr sheath was placed in the left femoral artery and a catheter was advanced to the right carotid artery. Angiography showed a vessel measuring 5-6 mm so a 6mm SpiderFX embolic protection device was deployed. The same process was completed in the left carotid artery. The sternotomy and operative resection of the left atrial mass was performed uneventfully on cardiopulmonary bypass after which the retrieval catheter was advanced and both carotid sleeves were removed. Direct inspection of the baskets showed no significant embolic material present. Consent was obtained to use this case for educational purposes.

Conclusion: This case represents a unique presentation of a left atrial myxoma in a teenager with a limb threatening embolic event. Carotid embolic protection devices are not commonly utilized in pediatrics but we report this case, which utilized these devices in a hybrid approach. We also believe that there are other indications in which utilizing these devices would be beneficial in pediatrics. ■

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