Resident & Fellow Rounds

March 2018 **Neurology.org/rf**

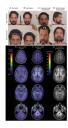
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A summary of recently published articles in the Neurology® Resident & Fellow Section

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March 6, 2018 issue

This issue includes 4 instructive clinical cases. The Pearls & Oy-sters provides an important lesson regarding the oculomotor examination in adults. The Clinical Reasoning covers the basics of the peripheral nervous system examination and highlights the value of genetic testing. In addition, there are 2 Teaching Neuro-Images cases. The first shows images of a rare metastatic lesion and the second a neuroinfectious disease case.

Pearls & Oy-sters: Eyes up: Ocular motor apraxia as essential differential diagnosis to supranuclear gaze palsy

We describe a patient with an unusual corticobasal syndrome who presented with oculomotor apraxia as the leading clinical sign. Loss of voluntary saccade control with preservation of the vestibulo-ocular reflex could be mistaken as supranuclear gaze palsy. This case illustrates oculomotor apraxia as an important clinical clue for differentiation of tauopathies.

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Clinical Reasoning: A teenager with left arm weakness

A 16-year-old boy presented with 2 painless episodes of left arm weakness. EMG showed left upper trunk brachial plexopathy as well as entrapment neuropathies at multiple sites. Genetic testing confirmed hereditary neuropathy with liability to pressure palsies (HNPP). HNPP should be considered in patients with painless brachial plexopathy.

Page e907

Teaching NeuroImages: Intradural, intramedullary spinal cord metastasis from primary renal cell carcinoma

Intradural intramedullary metastatic lesion from primary renal cell carcinoma is rare. A patient presented with lower extremity weakness and numbness and MRI of the thoracic spine showed metastatic lesions. Biopsy revealed positive stained cells with PAX-8 and Vimentin and negative for glial fibrillary acid protein, which was consistent with renal cell carcinoma.

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NPub.org/rf

Teaching NeuroImages: Gummatous neurosyphilis: An atypical presentation in a patient with HIV

A 29-year-old HIV-infected man presented with right eyelid ptosis and diplopia. On examination, a third nerve palsy was identified. Serum and CSF analysis revealed positive venereal disease research laboratory and *Treponema pallidum* hemagglutination titers. MRI showed right oculomotor nerve enhancement. A course of penicillin was administered with partial clinical recovery and complete radiographic resolution.

Page e913

March 13, 2018 issue

This issue begins with a discussion of the clinical presentation of poisoning in a pediatric patient. The next Clinical Reasoning case involves symptoms of infection in an immunocompromised adult. The 2 Teaching Neuro-Images cases highlight a unique cause of stroke and the use of ictal fundus photography in migraine.

Page 524

Clinical Reasoning: A 12-year-old girl with headache and change in mental status

A 12-year-old girl initially presented with headache and subsequently became obtunded with bulbar dysfunction and flaccid quadriparesis. MRI brain showed diffuse white matter injury involving periventricular regions, brainstem, and cerebellum. She was diagnosed with paradichlorobenzene (PDCB) (a substance in mothballs) toxicity, confirmed by elevated serum PDCB and urine dichlorobenzene levels.

Page 524

Clinical Reasoning: A 52-year-old woman with 3 weeks of progressive gait ataxia and dysarthria

A 52-year-old woman with a history of AIDS presented with subacute progressive ataxia and dysarthria. On examination, she also had frontal release signs and left hemisensory loss, suggesting a multifocal process. An MRI brain showed multiple areas of nonenhancing T2 hyperintensity, and the CSF JC virus PCR was positive.

Page e985

Teaching NeuroImages: Vertebrobasilar dolichoectasia with dissection manifested as infarct and subarachnoid hemorrhage

A 60-year-old man presented with vertigo, dysarthria, and left weakness for cerebral infarction with vertebrobasilar dolichoectasia (VBD) and dissection. He died of subarachnoid hemorrhage (SAH) several days later. VBD with dissection can manifest as sequential cerebral infarction and SAH, which predicts poor prognosis. Antithrombotic therapy and surgical and endovascular treatment are treatment options.

Page e990

Teaching NeuroImages: Retinal migraine in action

A 56-year-old man with migraine headache reported a 2-day history of 5 episodes of left eye visual loss, each lasting 5 minutes. Ictal fundus photography showed multiple vasoconstrictions of the central retinal artery/vein and their branches with resolution 10 minutes later. Vasospasm remains a possible contributor/epiphenomenon in retinal migraine.

Page e992

March 20, 2018 issue

This issue includes 2 Clinical Reasoning and 2 Teaching NeuroImages cases. The first case describes the approach to lower motor neuron syndromes. The second case reports the presentation of paraneoplastic limbic encephalitis. Next, striking magnetic resonance neuroimages are helpful in the diagnosis of leukoencephalopathy. Finally, there is a Teaching NeuroImages case of isolated mammillary body infarction presenting with memory impairment.

Page e1085

Clinical Reasoning: A 42-year-old man with unilateral leg weakness

This case describes a man with a family history of muscle weakness who developed proximal leg weakness. The authors review the differential diagnosis of lower motor neuron (LMN) syndromes, focusing on conditions that can affect the motor nerve up to the anterior horn and muscle diseases that can mimic LMN syndromes.

Page e1085

Clinical Reasoning: A 75-year-old man with parkinsonism, mood depression, and weight loss

A 75-year-old man with progressive parkinsonism and a history of weight loss and depression underwent several diagnostic examinations to investigate possible etiologies. LGI1 antibodies were found in serum. Neuroimaging was consistent with LGI1 encephalitis. The patient was treated with corticosteroids with initial improvement, but he eventually died of underlying leukemia.

Page 572

Teaching NeuroImages: Adult-onset vanishing white matter disease

Vanishing white matter disease is an inherited leukoencephalopathy with extremely wide phenotypic variation typically associated with episodes of sudden major neurologic degeneration triggered by febrile infections, minor head trauma, or stressful events. We report an 18-year-old patient who presented with headache and epileptic seizures with diagnostic MRI findings along with a p.Arg113His missense mutation in the *EIF2B5* gene.

Page e1091

Teaching NeuroImages: Isolated mammillary body infarction presenting with memory impairment

A right-handed man presented with acute-onset memory impairment. His verbal memory functions exhibited significant impairments. A brain MRI revealed an acute infarction in the left mammillary body.

Page e1093

March 27, 2018 issue

Congratulations to our readers who chose the correct diagnosis for the Mystery Case of an infant with pili torti and intractable epilepsy. Next, an adult with a rare cause of pediatric-onset progressive myoclonic epilepsy is discussed. The first Teaching NeuroImage describes a variation of a classic spinal cord syndrome. Finally, there is a video depiction of a rare cause of severe throat pain associated with syncope.

Page e1174

Mystery Case: Tortuous hairs and tortuous blood vessels

A 5-month-old boy, born to a consanguineous couple, presented with history of global developmental delay and drug-refractory epilepsy. On examination, he had microcephaly, scalp seborrhea, short twisted silvery wiry scalp hairs, loose skin folds, and central hypotonia. Hair microscopy showed typical features and bone radiographs showed metaphyseal spur and wormian bones.

Page e1174

Child Neurology: Type 1 sialidosis due to a novel mutation in *NEU1* gene

We describe type 1 sialidosis due to a novel missense mutation P210L in the *NEU1* gene in a 39-year-old Ecuadorian man with progressive myoclonic epilepsy, ataxia, and cherry-red spots. Differential diagnoses of this clinical presentation, including Unverricht-Lundborg disease, myoclonic epilepsy associated with ragged-red fibers, Lafora disease, neuronal ceroid lipofuscinosis, and Gaucher disease, are discussed.

Page 622

Teaching NeuroImages: Sulcal artery syndrome: A hemicord infarct presenting with incomplete Brown-Sequard syndrome

A 59-year-old man presented with acute right hemiparesis, pain, and temperature loss on the left below T4. Proprioception and vibration were normal. MRI showed a short segment of T2 hyperintensity, with restricted diffusion, at the right half of C5 spinal cord. He had sulcal artery occlusion, presenting with incomplete Brown-Sequard syndrome.

Page e1177

Teaching Video NeuroImages: Vagoglossopharyngeal neuralgia mimicking as a seizure

This is a case of vagoglossopharyngeal neuralgia, a very rare neurologic condition characterized by bradycardia and asystole in a patient with severe glossopharyngeal neuralgia.

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