Resident & Fellow Rounds

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

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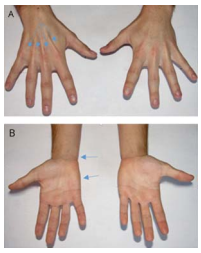

January 1, 2019 issue

Monomelic amyotrophy (Hirayama disease) is a rare lower motor neuron disease and the subject of this month's Clinical Reasoning case. Next, an Education Research study evaluates a neurology resident clinic. The Teaching NeuroImages describes a rare neurocutaneous syndrome involving swelling of the face. The Teaching Video NeuroImages depicts a rare dystonic movement disorder with bilateral upward eye deviation.

Clinical Reasoning: A 17-year-old baseball player with right hand weakness

This article presents the case of a 17-year-old with Hirayama disease, details the typical presentation of a rare entity, and outlines the characteristic electromyographic and imaging findings.
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Education Research: Electronic patient portal enrollment and no-show rates within a neurology resident clinic

These findings suggest targets to focus on to improve patient adherence to their clinic appointments, thereby increasing educational value for residents via increased encounters. In addition, practices may utilize their resources more efficiently to increase patient volumes, via reduction in no-shows, leading to potential increased revenue.
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Teaching NeuroImages: Melkersson-Rosenthal syndrome with permanent bilateral facial weakness

A 56-year-old woman presented with multiple, alternating attacks of Bell palsy, associated with lower labial edema and dysgeusia. The facial paresis and edema resolved only partially, and over the years she lost the ability to smile and kiss, and developed lacrimation while eating.
Page e81

Teaching Video NeuroImages: Oculogyric crisis in a 10-year-old girl

This case highlights a typical oculogyric crisis in a 10-year-old girl with a family history of a similarly affected sibling. The significance of the report lies in the recognition of oculogyric crisis in children that can lead to an accurate diagnosis of pediatric neurotransmitter disorders.
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January 8, 2019 issue

This issue contains multiple case reports that emphasize the importance of genetic testing in clinical neurologic care. First, the Clinical Reasoning case describes a genetic predisposition to cerebral thrombosis. Next, the Pearls & Oy-sters case describes a rare form of transmissible spongiform encephalopathy. Finally, one of the Teaching NeuroImage cases has imaging features of a pediatric mitochondrial disease. In contrast, the other Teaching NeuroImage case shows the neurologic sequelae of an acquired condition instead.

Clinical Reasoning: A 14-year-old girl with headache, seizures, and confusion

This article presents a case of cerebral venous sinus thrombosis due to homocystinuria with 2 novel compound heterozygous variants of the cystathionine β-synthase gene.
Page e161

Pearls & Oy-sters: Challenging diagnosis of Gerstmann-Sträussler-Scheinker disease: Clinical and imaging findings

This case report presents a patient with Gerstmann-Sträussler-Scheinker disease with de novo P102L pathogenic variant in *PRNP* and atypical clinical symptoms.
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Teaching Video NeuroImages: A 20-year-old man with distal paresthesia

A 20-year-old man presented with a 3-week history of progressive distal paresthesia in his lower limbs. His gait became clumsy, which led to loss of mobility.
Page e170

Teaching NeuroImages: Leigh-like features expand the picture of PMPCA-related disorders

The clinical and imaging features, resembling Leigh-like picture, in a child with biallelic pathogenic variants in PMPCA are described. Pathogenic variants in the *PMPCA* gene (the mitochondrial processing peptidase responsible for the cleavage of nuclear-encoded mitochondrial precursor proteins after import in the mitochondria) have been associated with cerebellar atrophy on MRI and an autosomal recessive spinocerebellar disorder (SCAR2).
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January 15, 2019 issue

The Pearls & Oy-sters case report from this issue emphasizes the importance of atypical presenting symptoms of a common disease. Next, new-onset seizures is the presenting symptom of the Clinical Reasoning case. The Right Brain is a poem highlighting the value of understanding the patient experience. Finally, the Teaching NeuroImage shows brain ischemia following the presenting symptom of monocular vision loss.

Pearls & Oy-sters: The dangers of PRES: An atypical case with life-threatening presentation

A case of posterior reversible encephalopathy syndrome in a transplant patient on immunosuppression is described. The presentation with acute hydrocephalus that required urgent external ventricular drain placement is unusual.
Page e282

Clinical Reasoning: A middle-aged man with new-onset seizures and myoclonic jerks

A 53-year-old man with a medical history of hypertension presented with new-onset seizures. On the day of admission, he was witnessed by his son to have right-sided jerking that lasted for 1 minute, after which he fell forward and lost consciousness.
Page e274

Right Brain: Little did he know

This poem was written after personal interactions with a patient who was recently seen by a medical practitioner for a seemingly benign disease with moderate functional impairment. The impact of a disease on a person is unique and variable based on the person's background, functional baseline, and beliefs.
Page 148

Teaching NeuroImages: CRAO and silent brain infarcts caused by cardiac myxomas in Carney complex

A 14-year-old girl presented with sudden painless left eye vision loss. The fundus photography showed left central retinal artery occlusion. Brain MRI revealed subclinical embolic infarcts. Chest CT confirmed multicentric cardiac myxomas. Gene testing revealed a pathogenic variant in the *PRKAR1A* gene.
Page e286

January 22, 2019 issue

This issue starts off with an Education Research study that evaluated neuroimaging training during residency. The first case is a Clinical Reasoning describing an adult who presents with mild symptoms of stroke. The Teaching Video NeuroImages case shows the specific eye examination findings in one type of mitochondrial disease. Finally, there is a pediatric Teaching NeuroImages case with an unusual-appearing lesion.

Education Research: Neuroradiology curriculum in neurology residency training programs: How we teach neuroimaging

Opportunities exist to improve neuroimaging education in neurology resident education. This can be done by closer adherence to the American Academy of Neurology neuroimaging curriculum guidelines, especially by expanding access to online resources and additional emphasis on imaging review with neurology subspecialists.
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Clinical Reasoning: A 61-year-old woman with acute-onset dysgraphia

A 61-year-old woman presented to the emergency department following acute-onset dysgraphia. Her presenting NIH Stroke Scale score was only 2 and for mild, nondebilitating symptoms (minimal right nasolabial fold flattening and mild, lingual dysarthria).

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Teaching Video NeuroImages: MT-TL1 mutation presenting as chronic progressive external ophthalmoplegia

This video case report demonstrates the typical eye movement findings in chronic progressive external ophthalmoplegia. The case was unique due to the presence of a MT-TL1 pathogenic variant, a gene typically associated with mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS).

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Teaching NeuroImages: Cerebral inception: A "brain" within a brain

An infant was found to have an unusually large area of dysplastic brain, with imaging characteristics of glioneuronal hamartoma. The mass has a brain-like appearance and is isointense to cortex.

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January 29, 2019 issue

Patients often present with alterations of gait for various reasons. This issue contains a fascinating Clinical Reasoning case that presents with lower extremity weakness. Congratulations to our readers who correctly identified the diagnosis in the latest Mystery Case. The first Teaching NeuroImages case describes the imaging findings in an atypical presentation of stroke in a dialysis patient. Finally, the last Teaching NeuroImages describes the constellation of symptoms and imaging findings in a pediatric-onset mitochondrial disease.

Clinical Reasoning: A 40-year-old woman presenting with distal leg weakness

A 40-year-old woman reported 15 years of progressive lower leg and hand weakness. She had trouble standing on her toes and developed progressive bilateral foot drop, worse on the right.

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Mystery Case: Eccentric target sign in CNS toxoplasmosis: A rare presentation of Good syndrome

A 61-year-old man with myasthenia gravis, who previously underwent post-thymectomy radiotherapy for a metastatic thymoma, presented with altered mental status. Examination revealed papilledema and fronto-executive dysfunction.

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Teaching NeuroImages: Facial swelling and intracerebral hemorrhage from venous hypertension in a dialysis patient

A case of intracerebral hemorrhage in a dialysis patient due to central venous hypertension from arteriovenous fistula obstruction, who initially developed facial swelling, is presented.

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Teaching NeuroImages: Kearns-Sayre syndrome

A healthy 19-year-old man presented with a complete heart block, subsequent ptosis and symmetric ophthalmoplegia, and pigmentary retinopathy. He was eventually diagnosed with Kearns-Sayre syndrome, a mitochondrial disease. While once considered extremely rare, mitochondrial diseases have an estimated prevalence of 1 in 5,000.

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