COMMENTARY. A commentary from the Department of Neurology, University of Szeged, Hungary, discusses the trigemino-vascular theory of migraine and the pain-transmission link between the vascular and neuronal regions [1]. PACAP and other neuropeptides have essential roles in activation of the trigemino-vascular system. PACAP38 is present in the trigeminal ganglion and caudal trigeminal nucleus. The effects of PACAP38 are mediated through G-protein-linked receptors, including PAC1. The clinical study by Amin et al [2] is consistent with previous laboratory animal studies comparing the effects of nitroglycerol and PACAP on the trigemino-vascular system [3].

References.
1. Vecsei L, et al. Brain. 2014 Mar;137(Pt 3):650-1.
2. Amin F, et al. Brain. 2014 Mar;137(Pt 3):779-94.
3. Markovics A, et al. Neurobiol Dis. 2012 Jan;45(1):633-44.

NEUROMUSCULAR DISORDERS

PHARYNGEAL-CERVICAL-BRACHIAL VARIANT OF GUILLAIN-BARRE SYNDROME

Investigators from National University Hospital, Singapore, review the clinical features of 13 cases of pharyngeal-cervical-brachial (PCB) variant of Guillain-Barre syndrome (GBS) and outline new diagnostic criteria. In a series of 100 patients with PCB reported previously from Japan [1] the age of onset ranged from 5-83 years (median age 43), antecedent upper respiratory tract infections and diarrhea occurred in 71% and 30%, respectively (similar to GBS), and 31% had serological evidence of Campylobacter jejuni infection. Cytomegalovirus, Epstein-Barr virus, Mycoplasma pneumoniae and Hemophilus influenzae were associated infrequently (6-1%). PCB is defined as 'pure' in patients presenting with rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia/hyporeflexia but without ophthalmoplegia or leg weakness [2]. In contrast, some recent series describe sensory disturbance in the upper limbs, and normal or exaggerated reflexes. GBS forms a continuum of overlapping syndromes, those cases with ophthalmoplegia and ataxia overlapping with Fisher syndrome. One half of patients with PCB carry IgG anti-GT1a antibodies that cross-react with GQ1b, whereas most patients with Miller Fisher syndrome carry IgG and GQ1b antibodies that always cross-react with GT1a. Significant overlap between the clinical and serological profiles of these syndromes suggests a PCB/Fisher syndrome continuous spectrum. The neurophysiological findings in PCB are axonal rather than demyelinating. Myasthenia gravis, botulism and other myopathic disorders are differentiated from PCB by the absence of sensory deficits or areflexia. (Wakerly BR, Yuki N. Pharyngeal-cervical-brachial variant of Guillain-Barre syndrome. **J Neurol Neurosurg Psychiatry** 2014 Mar;85(3):339-44).

COMMENTARY. The diagnostic criteria for PCB variant of GBS include: 1) symmetric oropharyngeal weakness, arm weakness and areflexia/hyporeflexia; 2) absence of leg weakness, ataxia and disturbed consciousness; and 3) 12hr-28day interval

between onset and weakness. Supportive findings include: antecedent infection, CSF albumino-cytological dissociation, neurophysiological evidence of neuropathy, and presence of IgG anti-GT1a or anti-GQ1b antibodies. Brain MRI may be indicated to exclude brainstem ischemia, inflammation or brain tumor.

References.
1. Nagashima T, et al. Arch Neurol. 2007 Oct;64(10):1519-23.
2. Ropper AH. Arch Neurol. 1986 Nov;43(11):1150-2.

RECURRENT MILLER FISHER SYNDROME

Investigators from University of Siena, Italy, describe 2 children with recurrent Miller Fisher syndrome. Episodes occurred at age 11.5 and 13 years in Patient 1 and at age 8 and 13 years in Patient 2. Both patients responded to treatment with steroids. Patient 1 presented with diplopia, unsteady gait and clumsiness. Neurologic examination showed ataxia, hyporeflexia, and ophthalmoplegia. Treatment with iv immunoglobulin was effective initially, but failed to prevent progressive weakness during the second attack that subsequently responded to steroid therapy. Patient 2 presented with paresthesia of hands and diplopia, ataxia, paresis of 6^{th} and 7^{th} cranial nerves, muscle weakness, and hyporeflexia. Recovery from both the initial and second attack followed steroid therapy. (Grosso S, Verrotti A, Tei M, Cornacchione S, Giannini F, Balestri P. Recurrent Miller Fisher syndrome in children. **Pediatr Neurol** 2014 Mar;50(3):269-71).

COMMENTARY. Recurrent Miller Fisher syndrome [1] is rare in childhood, and the second attack may be more aggressive and resistant to therapy. Steroids may be indicated if iv immunoglobulin is ineffective [2].

References.
1. Fisher M. N Engl J Med. 1956 Jul 12;255(2):57-65.
2. Toru S, et al. Muscle Nerve. 2012 May;45(5):763-4.

PARANEOPLASTIC DISORDERS

PARANEOPLASTIC DISORDERS AND OVARIAN TUMORS

Researchers in Chang Gung University, Kaohsiung, and National Cheng Kung University, Tainan, Taiwan, assessed the prevalence and spectrum of paraneoplastic neurological disorders (PND) in children with benign ovarian tumor and the long-term outcome. The charts of 133 female patients below 18 years of age diagnosed with a pathologically proven benign ovarian tumor, Jan 1993 – Dec 2010, were reviewed, mostly mature teratoma. Six patients (4.5%) had neuropsychiatric manifestations, the majority (5) with onset after age 10 years. Depression or low mood, headache, mutism, hypoventilation, seizures, hallucination, vomiting and hypersalivation were the most common symptoms. NMDAR encephalitis in 2 patients and acute disseminated encephalomyelitis in 1 partially resolved after tumor removal and immunotherapy. One patient not receiving immunotherapy had neurological sequelae and long ICU stay. (Hsu