

PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 18, No. 3

March 2004

HEADACHE DISORDERS

PSEUDOTUMOR CEREBRI AND SICKLE CELL DISEASE

Three children, all girls, ages 9, 8, and 16 years, with sickle cell disease (SCD) presented with headache and were diagnosed with pseudotumor cerebri (PC) at the Children's National Medical Center, Washington, DC. Patient 1 had SCD-Hgb SC and patients 2 and 3 had SCD-Hgb SS. Hemoglobin levels in patients 1, 2 and 3 were 9.3, 8.4, and 6.6 g/dL, respectively. Ophthalmologic examination revealed bilateral papilledema and normal visual acuity, except for an enlarged blind-spot in patient 1. Brain magnetic resonance (MR) imaging, including MR angiography and MR venography, were normal, with no signs of hydrocephalus, stroke, or venous thrombosis. Lumbar puncture showed elevated opening pressures of 44.5, 29, and 36 cm H₂O, respectively (normal <20 cm H₂O). The CSF protein, glucose, and cell counts were normal. Oral acetazolamide treatment was started at 8–15 mg/kg/day and was titrated up to 20 mg/kg/d. Treatment was discontinued after 3 months in patient 2 without relapse, patient 1 continues on a maintenance dose of 8 mg/kg/d at 10 months follow-up, and patient 3 is maintained on 15 mg/kg/d at 9 months follow-up. Patient 1 is obese, with a body mass index of 26.6 kg/m² (95th percentile for age), and she required prolonged treatment with acetazolamide and 2 therapeutic lumbar punctures for relief of symptoms of PC. Hydroxyurea, given to patients with SCD to diminish erythrocyte sickling, was discontinued in patient 1 because of continued severe headache. (Hydroxyurea also increases red blood cell mass and blood viscosity, perpetuating mechanisms responsible for PC.) These cases are the first reported of PC with pediatric SCD, and symptoms of PC improved or resolved despite chronic anemia. (Henry M, Driscoll MC, Miller M et al. Pseudotumor cerebri in children with sickle cell disease: a case series. *Pediatrics* March 2004;113:e265-e269). (Respond: Caterina P Minniti MD, Department of Hematology/Oncology, Children's National Medical Center, 111 Michigan Ave, NW, Washington, DC 20010).

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The editor is Pediatric Neurologist at Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois. PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals. Fax: 312-943-0123.

COMMENT. The diagnostic criteria for pseudotumor cerebri, also termed "Idiopathic Intracranial Hypertension" (IIH), include the following: headache, papilledema, and other symptoms/signs of increased intracranial pressure (ICP), CSF pressures of >250 mm H₂O, normal CSF findings, no or only false neurologic localizing signs (eg VIth nerve palsy), normal brain imaging, an awake and alert patient, and no identifiable cause of increased ICP (Dandy criteria reviewed by Binder DK et al. **Neurosurgery** March 2004;54:538-552). Increased ICP caused by venous sinus thrombosis, sleep apnea, or choroid plexus papilloma is termed a "secondary pseudotumor syndrome." Theories advanced in pathogenesis of IIH include: 1) excess CSF production; 2) increase in cerebral blood volume or brain water content; 3) disturbed CSF absorption secondary to increased sagittal sinus pressure; and 4) endocrinological dysfunction related to obesity and female preponderance. Among secondary causes, the association of increased ICP with dural venous sinus thrombosis is long recognized as "otitic hydrocephalus" secondary to otitis and mastoiditis (Foley J. **Brain** 1955;78:1-41). Magnetic resonance venography should exclude sinus thrombosis in atypical patients with suspected IIH. Other secondary causes for pseudotumor are those associated with medications and with systemic disease. Medications include vitamin A, antibiotics (eg tetracycline), sulfa drugs, growth hormone, oral contraceptives, corticosteroid withdrawal, and lithium. Systemic diseases associated with IIH include systemic lupus erythematosus, malignancies, Addison's disease, thyroid disease, uremia, and various anemias, including iron deficiency in childhood (Yager JY, Hartfield DS. **Pediatr Neurol** 2002;27:85-92) and SCD SC in a pregnant adult (Thomas E. **Obstet Gynecol** 1986;67(3 suppl):75-95). The above report is the first case of PC with SCD-SS. Symptoms of PC abate with the correction of iron-deficiency anemia, and in the above 3 cases of SCD, symptoms resolved regardless of a persistent chronic anemia.

TRIPTANS DO NOT INCREASE RISK OF STROKE IN MIGRAINE

The incidence of stroke, cardiovascular events, and death among 63,575 patients with migraine was compared in those prescribed a triptan (13,664, 21.5%), non-triptan-treated, and matched nonmigraine control subjects (77,239) identified from the General Practice Research Database by researchers at the Institute of Neurology, University College, London, UK, and Pfizer Inc, New York, NY. The mean observation periods were 35.6 months for migraine patients and 33.3 months for controls. Mean number of triptan prescriptions was 8.6 (range 1-316). Ergotamine treatment was also used in 582 (4.3%) of triptan-treated and 1,598 (3.2%) of non-triptan-treated group. Triptans (at least 3 prescriptions) were not associated with an increased risk of stroke, serious cardiovascular events, or death in this cohort of men and women patients between 15 and 60+ years. A small increased risk of stroke occurred in migraineurs not treated with triptan. The association between migraine and stroke decreased with increasing age at stroke. An association with migraine and stroke was observed across all age and sex groups, with the strongest associations in men aged 45 to 59 years and women of child-bearing age. Triptans were prescribed to those less at risk of stroke and myocardial infarction. (Hall GC, Brown MM, Mo J, MacRae KD. Triptans in migraine. The risks of stroke, cardiovascular disease, and death in practice. **Neurology** February (2 of 2) 2004;62:563-568). (Reprints: Dr G Hall, Grimsdyke House, Ravenscroft Park, Barnet, EN5 4ND, UK).