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Benign intracranial hypertension

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Benign intracranial hypertension (BIH) is a syndrome characterized by papilledema and elevated intracranial pressure in the absence of hydrocephalus or intracranial mass. The condition is found most often in obese females in the fourth decade of life. Etiology remains unclear but a wide variety of medications, disease states and altered physiology have been associated with its onset. The complaints of headache and disturbed visual acuity are those directly related to increased intracranial pressure. The most serious sequelae of untreated BIH is permanent, partial visual deficit. Early diagnosis and referral is important if visual loss is to be minimized or prevented. The case of a 33-year-old female with BIH presenting to a chiropractic office is described. The limited role of the chiropractor in diagnosis and monitoring of the condition is reviewed.
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KEY WORDS: chiropractic, headache, benign intracranial hypertension.

L'hypertension intracrânienne bénigne est un syndrome qui se caractérise par un oedème papillaire et une tension intracrânienne élevée en l'absence de masse hydrocéphale ou intracrânienne. Les cas se présente le plus fréquemment chez les femmes obèses dans la quarantaine. L'étiologie demeure obscure mais une grande variété de médicaments, d'états maladifs et de caractères physiologiques altérés ont été associés avec son apparition. Les plaintes relatives à des céphalées et à des troubles au niveau de l'acuité visuelle font partie des symptômes directement reliés à l'augmentation de la tension intracrânienne. La séquelle la plus grave de cas d'hypertension intracrânienne bénigne non traitée est la pert partielle et permanente de la vue. Un diagnostic précoce et l'orientation vers un spécialiste permettent de réduire ou prévenir la perte de la vue. Le cas d'une femme de 33 ans souffrant d'hypertension intracrânienne bénigne et se présentant dans le bureau d'un chiropraticien est décrit, et le rôle limité du chiropraticien dans le diagnostic et la surveillance de la condition de la patiente est passé en revue.
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MOTS CLÉ : chiropratique, céphalée, hypertension intracrânienne bénigne.

Introduction

Benign intracranial hypertension (BIH), also known as pseudotumor cerebri, is a disorder of intracranial pressure regulation characterized by papilledema and elevated intracranial pressure in the absence of hydrocephalus or intracranial mass. While the defect in the regulation of intracranial pressure is not clearly understood, the onset of the disorder has been associated with a wide variety of medications, disease states and altered physiology. Presenting complaints of headache, blurred vision and nausea are most often associated with an increase in intracranial pressure. The syndrome has a peak incidence in the fourth decade and affects women two to five times more often than men.[1] The typical patient presenting with BIH is usually female, obese and in her thirties.[2]

While the syndrome is considered to be benign, it is estimated that 17 percent of patients who develop BIH will experience partial, permanent visual deficits secondary to optic nerve atrophy.[3] Since six percent of patients who present to chiropractors do so for the examination and treatment of headache,[4] it is important to recognize those patients who may be experiencing headache associated with BIH. Early recognition of the disorder and referral for appropriate treatment is imperative if permanent visual loss is to be prevented.

The case of a patient with BIH presenting to a chiropractic office for treatment of headache is described. The following discussion highlights the possible mechanisms of BIH and associated factors thought to play a role in its onset. The most common presenting signs and symptoms, various treatments and prognosis are described. The role of the chiropractor in diagnosis and monitoring is emphasized.

Case report

A 33-year-old female customs clerk presented with headaches of one year duration. Initially the headaches were mild and transient, but in the past three months had increased in intensity and duration and were currently present on a daily basis. She described her headaches as diffuse, originating in the suboccipital region and radiating bilaterally to the frontal area. They were usually associated with increased levels of cervical and upper

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thoracic muscular stiffness and discomfort. Occupational and postural stresses aggravated her condition while rest decreased her symptoms. The patient had noticed that she had begun to clench her teeth when the headaches were particularly intense resulting in some right temporomandibular joint pain. Imipramine, an anti-depressant, had been prescribed by her physician but this had been ineffective in controlling her symptoms. She had been treated by a chiropractor nine months prior for the headaches and stiffness and this had afforded some relief. Her past health history indicated a prior use of oral contraceptives, mild alcohol intake and 15 pack/year cigarette use. She related a history of chronic ear infections, the most recent being three months prior. The patient also suffered from Wolff-Parkinson-White syndrome, a ventricular pre-excitation tachycardia, but this had not required treatment in several years.

On examination the patient appeared to be mildly overweight but healthy. Postural assessment was unremarkable. The range of motion of the cervical spine was limited on forward flexion, rotation and lateral flexion. Digital palpation of the associated cervical and thoracic musculature revealed hypertonic and tender levator scapulae, trapezius, rhomboid and suboccipital muscles bilaterally. Motion palpation demonstrated painful restrictions at the occiput – C1, C1–C2 and C2–C3 levels of the cervical spine. Deep tendon reflexes were normal bilaterally and muscle strength testing in both upper and lower limbs was 5/5. Pathological reflexes were absent and cranial nerves were intact. Cardinal fields of gaze, visual fields and fundoscopic examination were normal. Orthopedic examination of the cervical and thoracic spine was negative. Hypertonic and tender right masseter and temporalis muscles were noted but the temporomandibular joints were otherwise unremarkable.

A diagnosis of cervicogenic cephalgia was made. The patient was treated with spinal manipulative therapy to reduce the painful restrictions in the upper cervical spine, while soft tissue therapy was directed towards the associated muscular hypertonicities. Range of motion and resistive exercises were prescribed to enhance flexibility and strength of the cervical spine musculature. Initial progress was encouraging with a decrease in muscular hypertonicities and improved range of motion, but no improvement in frequency, intensity or duration of the headaches was noted. Three weeks following the initiation of treatment the patient began to experience blurred vision and paraesthesias in the ophthalmic, maxillary and mandibular distributions of the right trigeminal nerve. Because of the poor treatment progress and onset of altered cranial nerve function the patient was referred to her physician for further neurologic examination.

Over the next several months the patient underwent further extensive evaluation, including Doppler testing of the carotid and vertebral arteries, evoked potentials, CT and MRI scans of the brain; all tests were normal. Panorex views and tomograms of the temporomandibular joints failed to reveal any abnormalities within the articulations. On a subsequent physical examination papilledema became evident, suggesting increased intracranial pressure. A lumbar puncture was performed and an increased opening pressure of 310 mm H20 was noted. Analysis of the spinal fluid failed to indicate any abnormality. The patient reported immediate reduction in the intensity of the headaches and facial numbness following the lumbar puncture. Within four days her symptoms returned to the pre-lumbar puncture levels.

Based on the presence of papilledema, increased lumbar puncture opening pressure with normal cerebral spinal fluid analysis and negative intracranial investigations, a diagnosis of benign intracranial hypertension (BIH) was made. The patient was placed on Diamox to decrease aqueous humor secretion and to reduce overall fluid retention, and was instructed to undergo weight loss. Follow-up three months later showed excellent resolution of the headaches, but mild residual facial numbness was present. Subsequent ophthalmic examination demonstrated permanent partial vision loss of the right eye secondary to optic nerve atrophy.

Discussion

The cause of BIH is not fully understood. Under normal conditions, 450–500 ml of cerebral spinal fluid is produced daily in the choroid plexus within the third and fourth ventricles. Absorption of equal volumes of cerebral spinal fluid by the small vessels of the pia, ventricular walls and the arachnoid villi maintain a normal intracranial pressure of between 75 and 150 mm H20.[5] BIH is thought to occur from an imbalance in the production and absorption of cerebral spinal fluid. Possible mechanisms for this imbalance include increased blood volume (secondary to autoregulatory dysfunction), hypersecretion of cerebral spinal fluid and/or reduced cerebral spinal fluid absorption.[1] More specifically, some investigators feel that alterations in steroid hormone metabolism leads to a disturbance in the neuro-endocrine control of blood-brain water permeability, resulting in increased brain water and intracranial pressure.[6,7]

While the physiologic mechanism of BIH remains unclear certain types of medications, altered physiology and disease states have been associated with the onset of the syndrome. Long-term tetracycline therapy,[8] corticosteroid use,[9] lithium administration (for bipolar disorders)[10] and excessive Vitamin A intake have been implicated.[11] Reports linking the onset of BIH with hypoparathyroidism,[12] renal insufficiency,[13] systemic lupus erythematosus,[14] middle ear infection,[2,3] pregnancy,[15,16] and the resolution of chronic depression have also been documented.[17] The most frequently correlated physical finding in patients experiencing BIH is obesity.[1,2]

Clinical examination

The most common presenting complaints of patients with BIH are those directly related to increased intracranial pressure. Headache secondary to pressure on the pain sensitive structures of the brain has been shown to be present in 99 percent of all cases.[3] Other physical findings include unilateral or bilateral

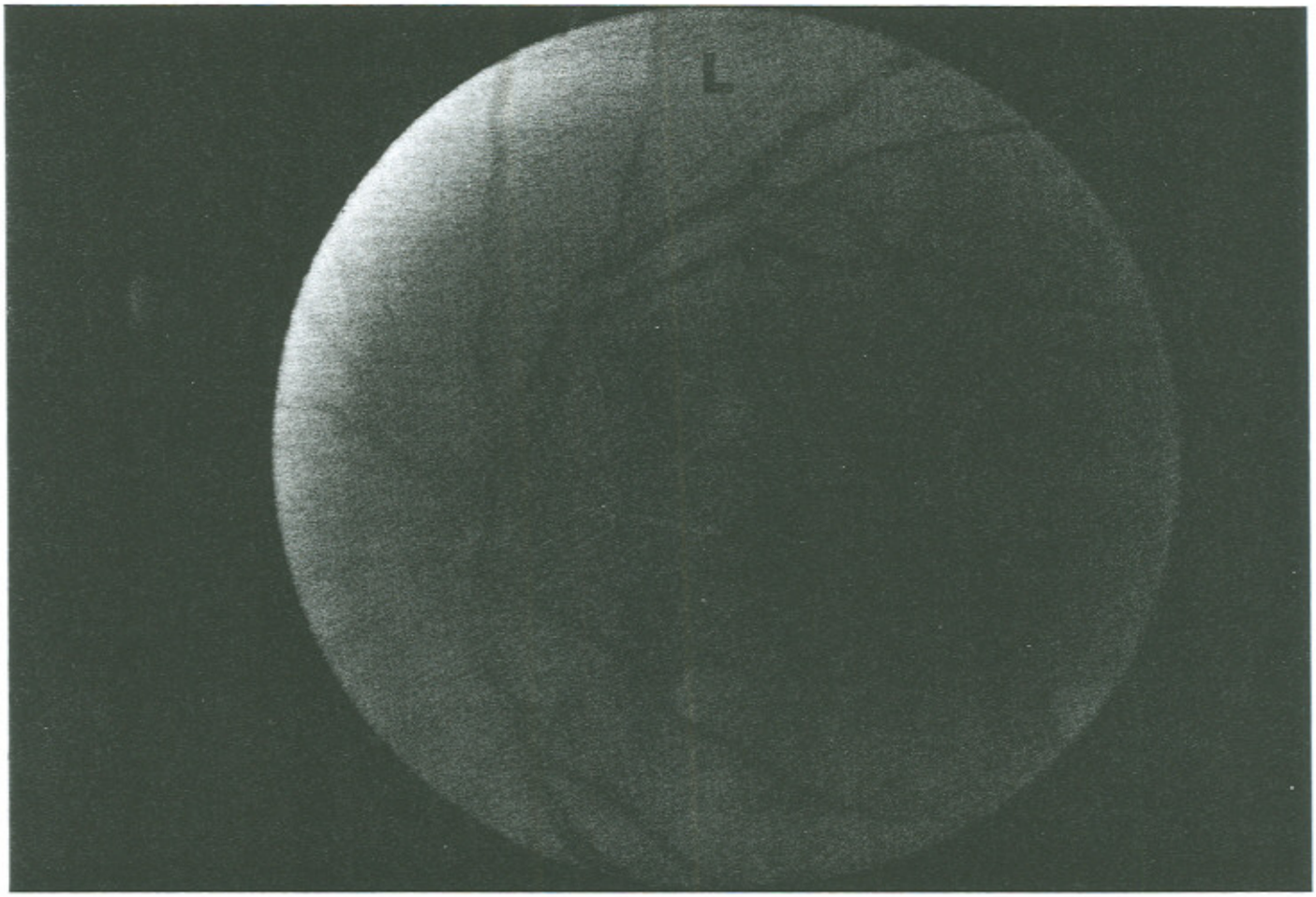


Figure 1. Fundoscopic examination of a patient exhibiting unilateral papilledema associated with BIH. Note the elevated and blurred margins of the

papilledema (Figure 1), disturbances of visual acuity, enlarged blind spots, visual field defects and nystagmus.[3] Subjective complaints include nausea, diplopia, alterations of consciousness, tinnitus and parasthesias.[3] Round and Keane, in a study of 102 patients, noted arthralgia as well as back and leg pain in 13 percent and 5 percent of cases, respectively.[18] Incidence of objective findings and subjective complaints are summarized in Table I.

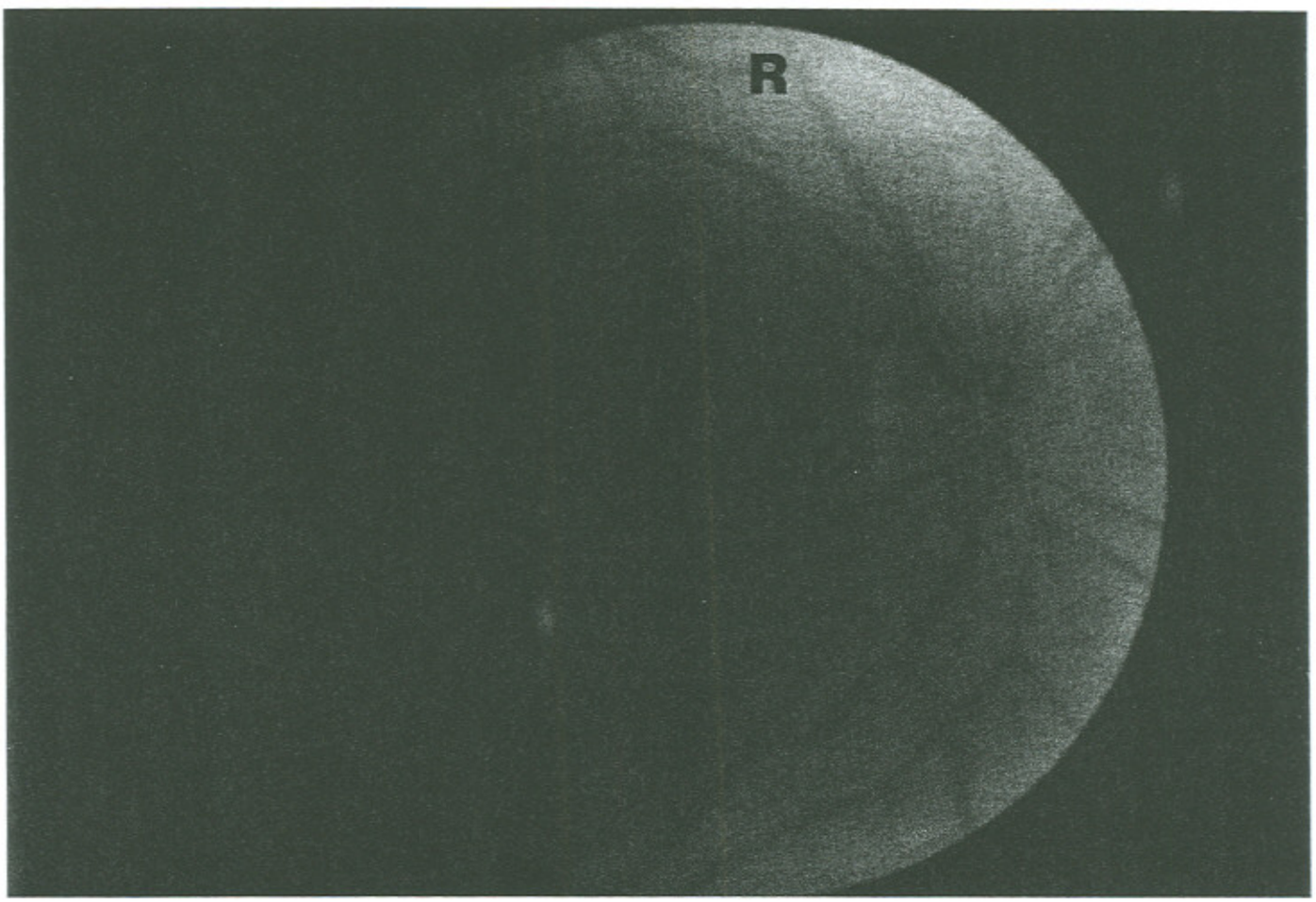
In most cases the onset of signs and symptoms is insidious, developing over a three to nine month period. Patients will typically avoid examination and treatment until the onset of visual disturbances. In cases where BIH occurs as a result of medication, the onset of symptoms can be within days. A particularly aggressive but rare form of "malignant pseudotumor cerebri" has been described in which signs and symptoms develop within hours resulting in rapid permanent blindness.[19]

A variety of treatments for BIH have been employed with varying degrees of success. In those patients where the onset is medication-induced, removal of the causative agent usually results in remission. Spontaneous remission often occurs fol-

TABLE I
Incidence of presenting signs and symptoms in BIH*

Symptoms		Signs	
Headache	99%	Papilledema	100%
Disturbances of visual activity	57	Enlarged blind spots	32
Diplopia	36	CN VI Palsy	24
Nausea	32	Decreased visual acuity	17
Dizziness	13	Visual field defects	15
Alteration of consciousness	10	Horizontal nystagmus	6
Tinnitus	8		
Parasthesias	2		

* Johnston and Patterson[3]



optic disc (large arrow) and increased tortuosity of the retinal vessels (small arrow) of the left eye. The optic disc of the right eye is normal.

lowing the initial diagnostic lumbar puncture. Certain medications such as carbonic anhydrase inhibitors have been shown to be effective in inhibiting the production of cerebral spinal fluid and reducing intracranial pressure. Prednisone, which decreases intracranial pressure, has been an effective agent in controlling BIH. Weight loss in obese patients has also been shown to be an effective method of treatment.[20] In extreme cases where patients fail to respond to the removal of known causative agents, or undergo spontaneous remission, serial lumbar punctures are performed to maintain normal intracranial pressure until lumbo-peritoneal shunting can be employed.

Patient prognosis is usually good. A retrospective analysis of 92 patients diagnosed with BIH found only three percent failed to experience remission after 11 years.[3] Twelve percent of patients had developed recurrences of the condition within five years of initial remission. The most significant sequelae of BIH is mild to severe partial permanent visual deficit secondary to optic nerve atrophy. Of the 92 patients studied 17 percent retained some degree of partial permanent visual loss. More importantly, there appears to be little correlation between duration of symptoms, degree of papilledema and visual deficit with the degree of permanent visual loss.[21] Furthermore, permanent visual loss can occur early or late in the condition.[22] Therefore, a patient presenting with minor levels of headache and little evidence of papilledema, may be experiencing early permanent visual loss. Identifying the patient with BIH early in its course is the major challenge to the primary contact practitioner.

While the treatment of BIH is primarily medical, chiropractors can play a role in diagnosis and management. It is estimated that six percent of all patients presenting to chiropractors do so for the examination and treatment of headaches.[4] In a few of these cases BIH may be an appropriate differential diagnosis. A careful, thorough history may indicate recent exposure to the medications and illnesses which play a role in the onset of BIH. The physical examination should include a thorough neurologic workup. Fundoscopic examination for papilledema, visual field testing for visual deficit and visual acuity is mandatory. If history and examination findings indicate the possibility of BIH, the patient should be referred for further diagnostic procedures. While history and examination findings may raise the

index of suspicion of BIH the diagnosis is not confirmed until the diagnostic triad of papilledema, elevated lumbar puncture opening pressure with normal cerebral spinal fluid and absence of intracranial mass is satisified.

In all cases of headache treated by chiropractors frequent re-examination is advised. In the case presented, the patient's headaches were initially diagnosed as cervicogenic. Failure to respond to manipulative therapy and the onset of further neurologic signs prompted a referral and the condition was subsequently diagnosed. If the treatment had continued and the referral delayed until the onset of papilledema became evident, further delay in diagnosis and treatment could have resulted, increasing the risk of permanent visual deficit.

Once the diagnosis of BIH has been made chiropractic treatment should be directed at the secondary manifestation of the headaches. Soft tissue therapy should be applied at the associated muscular hypertonicity and any spinal fixations adjusted. In cases where obesity is a factor, the chiropractor should reinforce the importance of weight loss and proper nutrition while establishing an appropriate exercise program.

In those patients with a known history of BIH, frequent neurologic assessment, including fundoscopic and visual field examination, should be performed. It is estimated that 12 percent of patients will experience a recurrence of the syndrome and further permanent visual deficit can develop during these exacerbations.[22] As in all headache patients, failure to respond to spinal manipulation or, when symptomatology and examination findings suggest progressive deficits, the patient should be referred for further investigative procedures.

Summary

The case of a patient presenting with headache associated with benign intracranial hypertension has been described. A thorough case history and physical examination can help idientify those patients at risk for this syndrome. Since visual loss can occur early or late in the syndrome, it is important that patients presenting with headache associated with BIH be referred for further diagnostic procedure and treatment. Once a diagnosis has been established, chiropractic management of the secondary manifestations of the syndrome can be instituted. Since recurrence of the syndrome develops in 12 percent of cases, with the possibility of further permanent visual deficit, frequent re-examination should be performed. Chiropractors should also realize that a thorough and complete fundoscopic examination is sometimes difficult without mydriatic solutions to dilate the pupils. In all cases of headache that fail to respond early to spinal manipulative therapy, and when fundoscopic examination has been inconclusive, referral for further examination is prudent.

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