ORIGINAL ARTICLE

Idiopathic Intracranial Hypertension (Prospective Study of 25 Patients)

Vishwanathan Ananth, Irshad Ahmad Sirwal, Tahira Banoo

Abstract

From January 1996 to January 2000, twenty-five patients were diagnosed to have idiopathic intracranial hypertension at our institution. Twenty-three were females and two were males with age range of 13-43 years. Main presenting features were headache (100%), vomiting (44%), blurred vision (36%), diplopia (28%), neck pain (12%), and transient visual obscuration (12%). Duration of symptoms before diagnosis ranged from one week to 8 months. Nine patients had cranial nerve involvement. Seventeen patients were obese, 3 were using oral contraceptives and one patient had taken tetracycline for 4 months for acne and other was taking Vit. A 50,000 units twice daily for 9 months. All patients had papilloedema. CT and MRI brain were done in all to exclude intracranial pathology. Patients were followed up every two to three months. At five years follow up, 19 were symptom-free with normal vision and 6 had residual visual deficit. Idiopathic intracranial hypertension once diagnosed necessitates regular follow-up to avoid permanent visual loss.

Key words

Idiopathic intracranial hypertension, Papilloedema.

Introduction

Idiopathic intracranial hypertension is a syndrome of obscure origin, also known as pseudotumor cerebri and benign intracranial hypertension, affecting women of child bearing age. Headache is the cardinal symptom and papilloedema, the single most important physical finding (1,2). A host of associations said to be causally related include endocrine disorders, collagen diseases, drugs, etc. Diagnosis requires documentation of elevated CSF pressure with normal CSF profile and normal cranial imaging. Therapy is directed at reduction of CSF pressure. Repeated lumbar puncture represents a practical treatment modality with high degree of success (3). Other modalities include weight reduction, short term corticosteroids, carbonic anhydrase inhibitors, oral hyperosmotic agents, and diuretics (4). Repeated examination of visual function is essential during follow up to detect early and potentially reversible visual loss. In unresponsive patients lumboperitoneal shunt or unilateral optic nerve sheath fenestration should be considered. We report here our experience with 25 patients of idiopathic intracranial hypertension and guidelines for managing such patients.

From the Department of Neurology, King Fahad Hospital, Medina Al Munawarah, Kingdon of Saudi Arabia.
Correspondence to : Dr. V. Ananth, Neurologist, Post Box 2874, King Fahad Hospital, Medina Al Munawarah, Kingdon of Saudi Arabia.

Material and Methods

Twenty-five patients who met Modified Dandy Criterias for diagnosis of idiopathic intracranial hypertension were included in the study. Patients with secondary intracranial hypertension were excluded. Detailed history for symptoms and associated risk factors, complete physical examination including ophthalmology, laboratory evaluation including work-up for vasculitis, complete CSF analysis and detailed radiological evaluation including cranial CT Scan, MRI and MRA were carried out in all patients. Repeat lumbar punctures were done on follow up in some of the patients. Medications incriminated as associated risk factors were stopped at first instance. Patients were regularly followed up in Outpatient Department every two to three months and frequent ophthalmological examination carried out as and when needed to evaluate the response to treatment and to detect visual complications.

Results

Of twenty-five patients, 23 were females and 2 were males. Age ranged between 13-43 years (mean 25.76). Patients' characteristics are shown in Table 1. The symptoms and signs at the initial presentation are shown in Table 11. The duration of symptoms and signs prior to diagnosis was from one week to 8 months. The most common presentation was headache and papilledema. CSF pressure ranged from 250 to 650 mm H_2O.

At 5 years, 19 patients (76%) had no visual deficit and 3 patients (12%) had mild bilateral optic atrophy, one (4%) had enlarged blind spot, one (4%) had caecocentral scotoma and one (4%) had bilateral severe optic atrophy. Eighteen patients did not require any further hospitalization whereas the disease recurred in 7 patients and 6 of these had persistent visual problem. These patients were irregular on follow-up and neglected their symptoms. Patients with history of drug intake had normal neuro-ophthalmological status at the first follow-up.

Table I

Patients' Characteristics

Total number	25
Female	23 (92%)
Male	2
Age range	13 - 43, (mean 25.76 years)
Associated risk factors	
Obesity	17 (68%)
Oral contraceptives	3 (12%)
Tetracycline	1 (4%)
Vitamin A	1 (4%)
Pregnancy	1 (4%)
Hypothyroidism	1 (4%)

Table II

Symptoms and signs at initial presentation

Signs & symptoms	No. (%)
1. Headache	25 (100%)
2. Papilloedema	25 (100%)
3. Vomiting	11 (44%)
4. Blurred vision	9 (36%)
5. Abducent nerve palsy	8 (32%)
6. Diplopia	7 (28%)
7. Neck pain	8 (32%)
8. Transient visual obscuration	3 (12%)
9. Numbness of face	3 (12%)
10. Diminished visual acuity	3 (12%)

Discussion

Idiopathic intracranial hypertension is common in obese females. In the present study of twenty-five patients, 17 were obese females. Obesity has been reported in 97% of such patients (5) and deterioration of visual field grade was significantly associated with weight gain during the year before diagnosis (2). In a

retrospective study of 15 patients, 6% weight loss was associated with marked resolution of papilledema (6). Noggle and Rodning described a morbidly obese patient with rapidly advancing pseudotumor cerebri in whom gastric exclusion surgery resulted in weight loss and subsequent stabilization of visual deficit (7). In the study reported by Sugerman *et. al.*, 24 morbidly obese women underwent bariatric surgery (23 gastric by-pass and one gastric banding) and concluded that this has higher rate of success than CSF-peritoneal shunting (8).

We encountered associated drug intake in 5 patients. Various drugs associated with development of idiopathic intracranial hypertension are corticosteroids, hypo or hypervitaminosis A, tetracycline, nalidixic acid, minocycline, nitrofurantoin, amiodarone, indomethacin, anabolic steroids, lithium, etc. (3).

The commonest cranial nerve palsy is the sixth, which was encountered in 8 of our patients as reported by others. Chutorian AM (9) reported facial palsy and Speer C (10) reported fourth nerve palsy in patients with idiopathic intracranial hypertension.

Rowe FJ (11), in a prospective study of 35 patients found that visual outcome was excellent in 83% of cases. In the present study, 19 (76%) had normal vision and 6 (24%) patients had visual deficit at 5 years follow-up. Absence of symptoms after initial treatment does not always signal the end of elevated intracranial pressure. Hence, periodic visual assessment is necessary to detect insidious visual loss. Visual field assessment is the most sensitive indicator of visual loss than visual acuity and contrast sensitivity testing.

All our patients underwent cranial CT and MRI. Though MRI is predominantly done to exclude secondary causes of intracranial hypertension, a constellation of MRI imaging signs can assist in establishing the diagnosis and predicting increased intracranial pressure (12). However, MR venography might not add significantly to the evaluation of typical idiopathic intracranial hypertension but may be indicated in male, thin or elderly patients (13).

Although conventional methods of treatment still hold true, surgical intervention should be considered in the event of failure of medical treatment at the onset, severe visual loss, pregnancy or recurrence (14) . The two basic surgical interventions are optic nerve fenestration and CSF shunting, and both appear to be effective surgical means to reduce the pressure on the optic disc (15,16).

To avoid permanent visual deficit, patients with idiopathic intracranial hypertension should be evaluated by perimetry and fundoscopy periodically. Surgical intervention should be offered at the earliest if medical therapy fails. Stress should be laid on avoiding offending drugs.

References

1. Corbett JJ, Mehta MP. CSF pressure in normal obese subjects and patients with pseudotumor cerebri. *Neurology* 1983 ; 33 : 1386-88.
2. Wall M. Idiopathic intracranial hypertension. *Neurol Clinics* 1991 ; 9 : 73-95.
3. Steven L Wald. Disorders of cerebrospinal fluid circulation and brain edema. In: Walter G, Bradley Robert B, Doroff eds. Neurology in clinical practice, 2nd edition, Boston Butterworth Heivemann. 1996 ; 1431-58.
4. Disturbances of cerebrospinal fluid and its circulation including hydrocephalus and meningeal reactions In: Raymond D. Admas, Maurice Victor, Alla H Ropper. eds. Principals of Neurology 6th edition. New York, Mac Graw Hill 1997 ; 623-41.
5. Wall M, George D, Idiopathic intracranial hypertension: A prospective study of 50 patients. *Brain* 1991 ; 114 (Pt IA) : 155-80.
6. Jhonson LN, Krohel GB, Marsden RW, *et. al.* Role of weight loss and acetozolamide in the treatment of idiopathic

intracranial hypertension. *Ophthalmology* 1998 ; 105 (12) : 2313-17.

7. Noggle JD, Rodning CB. Rapidly advancing pseudotumor cerebri associated with morbid obesity: an indication for gastric exclusion. *South Med J* 1986 ; 79(6) : 761-63.
8. Sugerman HJ, Felton WL 3rd; Sismanis A, *et. al.* Gastric surgery for pseudotumor cerebri associated with severe obesity. *Ann Surg* 1999 ; 229(5) : 634-40 ; discussion 640-42.
9. Chutorian AM, Gold AP, Brawn CW *et. al.* Benign intracranial hypertension in Bell's Palsy. *N Eng J Med* 1997 ; 296 : 1214-15.
10. Speer C, Pearlman J, Phillips PH *et. al.* Fourth cranial nerve palsy in paediatric patients with pseudotumor cerebri. *Am J Ophthalmol* 1999 ; 127(2) : 236-7.
11. Rowe FJ, Sarkies NJ. Assessment of visual function in idiopathic intracranial hypertension: A prospective study *Eye* 1998 ; 12 (Pt I): 111-18.
12. Broadsky MC, Vaphides M. Magnetic resonance imaging in pseudotumor cerebri. *Ophthalmol* 1998 ; 105 (9) : 1686-93.
13. Lee AG; Brazis PW. Magnetic resonance venography in idiopathic pseudotumor cerebri. *J Neuro Ophthalmol* 2000 ; 20(I) : 12-13.
14. Smadjad MH, Smadjad, Ayeboua L *et. al.* Benign intracranial hypertension. Retrospective study of 20 cases. *J Fr Ophthalmol* 1998 ; 21(I) : 42-50.
15. Burgett RA, Purvin VA, Kawasaki A. Lumbo-peritoneal shunting for pseudotumor cerebri. *Neurol* 1977 ; 49(3) : 734-39.
16. Corbett JJ, Thompson HS. The rational management of idiopathic intracranial hypertension. *Arch Neurol* 1989 ; 46(10) : 1049-51.