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REVIEW

Idiopathic intracranial hypertension: diagnosis and management

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ABSTRACT

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, may occur at any age but is primarily a disease of obese women between the ages of 15 and 40. Diagnosis is made based on modified Dandy criteria, which include normal neuroimaging studies, elevated intracranial pressure (ICP) on lumbar puncture, signs and symptoms of elevated ICP, and a non-focal neurologic examination aside from sixth nerve paresis. Presenting symptoms include headache, pulsatile tinnitus, transient visual obscurations, diplopia, and visual field constriction. Diagnosis may be delayed because headache from other causes such as migraine is much more common, and patients may be treated for this condition without improvement before the actual diagnosis is recognized. Ophthalmologic signs may include papilledema and esotropia (from the sixth nerve palsy) as well as visual field and even visual acuity loss; the latter two signs are very ominous, as they indicate severe and potentially permanent visual damage. Medical and surgical options vary depending on the severity of the disease and are used to control headache as well as to prevent vision loss. Because most IIH is associated with obesity, weight loss is an essential element of any treatment regimen, as retrospective as well as prospective studies have shown disease resolution with as little as 6% weight reduction. Secondary causes of high ICP are being recognized in a greater number of IIH patients, and the term "idiopathic" likely applies to fewer patients than in the past. The underlying pathogenesis of the disorder remains elusive.

Key words: Idiopathic intracranial hypertension, lumbar puncture, papilledema, pseudotumor cerebri, visual field loss

Introduction

Patients with idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri (PTC), often present to the ophthalmologist because of the complaint of blurred vision or headache. The prompt and accurate recognition of this condition is beneficial both because it establishes the correct diagnosis with a minimum of delay, but also because PTC is a potentially blinding disorder. Recent advances in the study of the basic biology and the clinical pathophysiology of PTC have changed our approach to this disease condition. The term PTC syndrome should be used in favor of IIH, because we are now able in many cases to identify the cause of the elevated intracranial pressure (ICP) and treat it with specifically targeted therapies.

Etiology of Pseudotumor Cerebri

The major risk factors for developing PTC remain obesity and female sex [1]; although no new major epidemiological studies have appeared in the past decades, clinical experience supports their conclusions. The incidence of PTC has not exploded with the current "epidemic of obesity," and clearly, all obese patients do not develop papilledema and headache. Anatomic factors that increase venous pressure through resistance in the intra-thoracic or intraabdominal compartments may play a role in causing disease, and small cases series in which these pressures were temporarily altered have demonstrated improvement in symptoms and signs [2]. The role of weight loss in the long-term management of PTC must be emphasized to patients, and providing nutritional guidance and counseling may be of benefit. In morbidly obese patients body mass index (BMI >40, or BMI >35 with co-morbid disease), bariatric surgery should be considered seriously if conservative measures do not give adequate results [3].

Secondary causes of PTC syndrome include physically obstructive lesions or conditions (obstructive hydrocephalus, Chiari I malformation) [4], numerous drugs [5-7], vitamin A and related retinoids [8], and impaired venous drainage due to venous sinus thrombosis or stenosis [9-11]. It is especially important to consider these secondary causes of PTC in patients who do not fit the standard demographics; this population includes all men (regardless of weight), non-obese women, and women over the age of 45 (again, regardless of weight/obesity). To some extent, the workup described below applies to all patients, and the distinction in the initial workup is not too important; in other words, all patients get the same initial history and examination, followed by appropriate neuroimaging studies to look for underlying secondary causes of PTC syndrome. The purpose of mentioning the "non-traditional" PTC patients is to re-emphasize the need in

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all patients to look for a cause that might be treated specifically rather than palliatively through ICP lowering alone.

The diagnostic criteria for PTC syndrome have remained unchanged for a number of years. The modified Dandy criteria still apply and must be satisfied for the diagnosis to be made [12] [Table 1]. Certain criteria deserve special mention. The concept of "pseudotumor cerebri sine papilledema" has been described in the literature with cases of patients whose optic disc appearance is normal (except for the lack of spontaneous venous pulsations), headache is present, and the cerebrospinal fluid pressure is found to be elevated [13]. The existence (or lack thereof) of this form of the PTC syndrome is controversial and beyond the scope of this handout. This condition appears to exist with the proper anatomic variant (optic nerve sheath not being continuous to the back of the globe, thus "insulating" the nerve head from the increased ICP) but must be considered as an uncommon diagnosis of exclusion. It is more likely that a patient with another headache syndrome will be labeled with IIH because of a lumbar puncture (LP) done with poor technique, for example.

What is normal intracranial pressure?

The definition of normal ICP has been controversial but has been clarified in a number of well-designed studies [14,15]. It was argued that ICP is elevated by obesity alone, thus calling into question the meaning of elevated readings in many patients with pseudotumor cerebri (the majority of whom are obese; see below). In adults, regardless of body habitus, normal opening pressure is <20 cm H_2O. From 20 to 25 cm H_2O is a "grey zone" of indeterminate significance, although many neurologists would consider these numbers normal. Values above 25 cm H_2O are clearly abnormal; however, it is crucial to ensure the opening pressure was measured in the lateral decubitus position with the legs extended and the patient breathing normally. Flexion of the legs, curvature of the spine, and breath holding all may falsely elevate the measured opening pressure and lead to an inaccurate diagnosis. Measurement in the prone position (often done when the procedure is performed under fluoroscopic control) has not been studied in a large, comparative series [16]. The available data show that readings are comparable to those obtained in the lateral decubitus position, but concern remains that in obese patients, the increased intra-abdominal pressure when prone could elevate the measured pressure. We are currently studying this question in more detail. Normal ICP in infants and pre-pubertal children was thought to be significantly lower, but a recent study demonstrated that opening pressures to 28 cm H_2O should be considered normal [17], and that positioning seems to have little importance [18]. Moderate sedation should not markedly affect these values, but falsely low readings may be obtained under general anesthesia [19].

Table 1: Modified Dandy criteria for diagnosis of PTC syndrome

Signs and symptoms of increased intracranial pressure	Papilledema, headache, transient visual obscurations
Normal neuroimaging study	MR brain with gadolinium; MR venography; no evidence of obstructive hydrocephalus
Normal neurologic examination	Sixth nerve palsy (unilateral or bilateral) is allowed
Elevated intracranial pressure	Normal cerebrospinal fluid contents also must be documented or confirmed

Diagnosis

Lumbar puncture is mandatory for the diagnosis of PTC, not just to confirm that the opening pressure is elevated, but also to ensure that the cerebrospinal fluid formula is normal. A number of inflammatory or infectious conditions may present with disc swelling and signs and symptoms of elevated ICP, and diagnosis of these conditions when they exist is imperative. Some patients will have a normal opening pressure at the time of LP but have signs/symptoms of elevated ICP. Such patients may benefit from ICP monitoring (inpatient study, done for 48–72 h). Like blood pressure and intraocular pressure, ICP does vary diurnally.

Neuroimaging

All patients with newly diagnosed bilateral optic disc swelling must have a neuroimaging study done prior to LP. The study is done not only to ensure the absence of a space-occupying lesion that could cause brain herniation upon LP; it also can identify hydrocephalus, hemorrhage, and other anomalies that change the diagnostic workup. Magnetic resonance imaging (MRI) with MR venography (MRV) is the most complete study to obtain in patients with suspected PTC (see below). However, in the acute setting, it may be difficult to get an MRI that will be done with the specific sequences and quality desired. Thus, for most patients, a computed tomography (CT) scan of the head should be performed on the day of the diagnosis of bilateral optic disc swelling. It is imperative to obtain a cranial imaging study of some sort the same day; if it cannot be done, then a reason should be documented and patient understanding of the potential risk noted. In the modern age, it is very unlikely that a brain tumor with obstructive hydrocephalus will present to the ophthalmologist first. However, malpractice cases have been brought successfully against physicians who failed to diagnose brain tumors in such patients who subsequently collapsed before their MRI or CT scheduled for days in the future could be obtained.

Assuming the head CT is normal, both LP and MRI/MRV can be done on a less urgent basis unless there are other neurological signs or symptoms to prompt earlier evaluation (meningeal signs, focal deficits, etc.). Such patients by definition do not have PTC and are excluded from this discussion. MRI should be ordered with gadolinium contrast unless there is a specific contraindication to its use. Such conditions include allergy to the contrast substance and, possibly, renal insufficiency. Nephrogenic systemic fibrosis (NSF) was initially described

in patients with severe diabetes-related renal insufficiency or failure [20-22]; it is an extremely rare but potentially devastating condition related to gadolinium infusion. Concern over the potential for causing NSF has led many radiologists to restrict the use of gadolinium contrast in all patients with any degree of renal dysfunction. Communication with the radiologist will help to forestall any difficulties. As most PTC suspects are young and often healthy (aside from obesity), NSF is rarely a concern or issue.

Venography

Several methods are available to obtain MRV images. Most commonly used is time-of-flight (TOF), which captures changes in magnetic orientation occurring within the veins over the course of a specific scan sequence [23]. The source images are then post-processed to create a two-dimensional or three-dimensional reconstruction of the cerebral sinuses and large veins. TOF images are obtained rapidly and convey useful information. The reconstructions can be misleading, with artifacts induced by in-plane scanning that occurs with certain vessels (the transverse and sigmoid sinuses are most frequently affected) [23,24]. False narrowing or venous discontinuity may be seen on the reconstructed images and must be confirmed either by examining the source images or by viewing a reconstruction done in an orthogonal plane. For example, the transverse sinus runs in a plane such that an axial image may show only part of its diameter or fail to demonstrate it entirely due to slice thickness. It would thus appear abnormal on the reconstructed images regardless of its real condition. Coronal imaging also may miss a major portion of the transverse sinuses, and sagittal imaging adds a third view. Not unexpectedly, three-dimensional TOF MRV is much more accurate than two-dimensional methods for finding real venous abnormalities. Even more sensitive is gadolinium-enhanced MRV, which helps to reduce some of the artifacts inherent in the non-contrast technique, and time-resolved methods have been explored recently in addition to contrast use [24-26]. For optimal results the contrast must be given as a bolus. Contrast-enhanced techniques should be considered as the study of choice for most patients.

A less-widely used but very powerful MRV method is the auto-triggered elliptic centered oriented technique [27]. Few papers have been published on this technique, and only one report exists on its use in PTC. The physics of the technique helps to compensate for the artifacts noted above with the TOF method and reported data show very high sensitivity for detecting venous sinus abnormalities in PTC patients. However, the paucity of published literature, given the potential power of the method, suggests that its use is quite challenging. We do not employ this method presently at our institution but continue to be interested in its development and future utility. Instead, we have been using multidetector CT venography (MDCT) after LP (detailed below) to help in the identification of "real" venous stenoses and to prepare a treatment strategy.

Multidetector computed tomography venography

Non-invasive venous sinus imaging has been advanced significantly with the advent of 64-, 256-, and 320-MDCT scanning. The use of multiple detectors permits the rapid acquisition of information from a larger anatomic area than would otherwise be possible. While both 64- and 256-MDCT scanners are able to capture the venous structures in high resolution, they cannot capture the complete transit of contrast material from the beginning of the arterial phase until the end of the venous phase. Thus, the detection of fistulas or venous sinus thrombosis becomes more difficult. The advent of 320-MDCT and its popularity in cardiac CT studies makes this powerful tool much more widely available today [Figure 1].

Catheter venography

Use of this method is limited almost entirely to therapeutic maneuvers based upon the non-invasive study findings.

Use of Clinical Imaging

Almost all PTC patients at presentation will lack spontaneous venous pulsations. The re-appearance of spontaneous venous pulsations may be a useful clinical sign and shows that ICP has been lowered after treatment [28-30]. It however, cannot be the sole sign and improvement of other signs and symptoms such as diplopia, headache, and visual fields also should be documented.

Both the diagnosis and clinical management of PTC patients is enhanced by the use of photographic and ultrasonographic methods. Stereo optic disc photos assist in the documentation of the disc swelling and in serial clinical examination [Figure 2]. The clinical utility of photography has not been studied in a systematic fashion in patients with papilledema, but expert opinion and longstanding

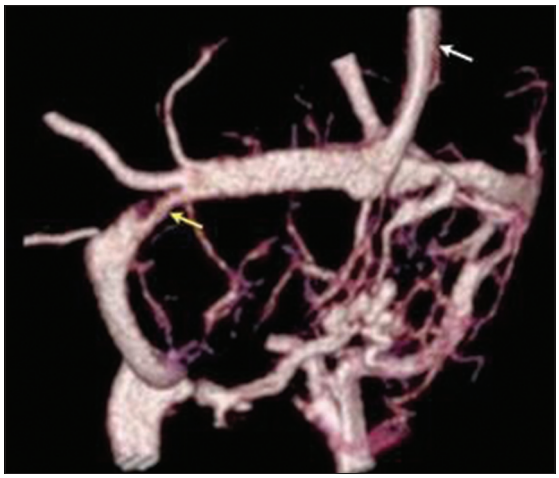

Figure 1 Left transverse sinus stenosis (yellow arrow) in a patient with pseudotumor cerebri, demonstrated by 320-multidetector computed tomography after lumbar puncture to lower intracranial pressure to 15 cm H_2O White arrow shows patent superior sagittal sinus

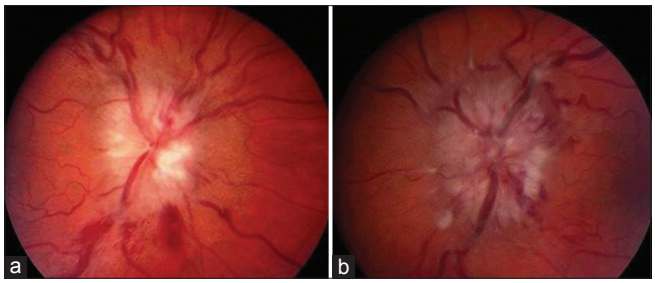

Figure 2 (a) Left eye of pseudotumor cerebri patient with acute Frisen grade 3 papilledema shown with peripapillary hemorrhages and nerve fiber layer infarct ("cotton wool spots"). (b) Right eye of the same patient with grade 4 papilledema. Note engorgement of the retinal veins

clinical practice support its use. Digital imaging provides adequate quality, but undilated photos may not give proper stereopsis to allow the physician to see changes in the degree of swelling.

Optical coherence tomography (OCT) of the retinal nerve fiber layer has been studied as a means of diagnosing true papilledema vs pseudopapilledema [31-33]; unfortunately, the results are mixed. One study reported that the consideration of optic disc contour as well as the size and configuration of the subretinal space allowed accurate discrimination of the optic disc swelling and disc drusen [34]. Additional studies in progress using spectral domain OCT show that this technique may have an even greater ability to tell these entities apart. OCT also can be very useful in following patients with papilledema to determine clinical improvement or to worsen [32]. OCT measurements are accurate to ±7 μ; therefore, changes in excess of this amount between clinic visits can be considered as significant. Research is ongoing to define the correlation between the absolute nerve fiber layer thickness and degree of visual field loss. The relationship is likely non-linear and may be dependent upon the quadrant(s) in which swelling and thickening occur.

B-scan ultrasonography remains a very useful tool in distinguishing papilledema from intrinsic disc swelling or elevation (pseudopapilledema or disc swelling from other causes) [35]. Anomalous or elevated optic discs without swelling can be differentiated from truly swollen discs by measuring the retrobulbar optic disc diameter. The presence of fluid in the subarachnoid space of the optic nerve sheath is measured using the 30° test. Like all methods, its sensitivity and specificity are not 100%, and it does require a skilled operator for reliable results. Ultrasound also may reveal buried optic disc drusen that give the appearance of papilledema.

Treatment

Since most cases of PTC syndrome are still in the idiopathic category, treatment is directed at lowering the ICP. Short-term medical management is most commonly initiated with acetazolamide (Diamox), which must be prescribed at the proper frequency and dose to have any real effect on ICP. At a minimum, adult patients of normal body weight or greater (the vast majority of patients) will require a daily dose of at least 1000 mg, although some patients may respond to less; headache relief may occur at lower doses without appreciable ICP lowering effect. Acetazolamide has a short half-life, and if given in standard tablet form, is usually dosed four times a day. Twice daily dosing is possible with extended-release capsules (Diamox Sequels). Doubling up the tablet dose is currently being used as part of a randomized, prospective clinical trial (Idiopathic Intracranial Hypertension Treatment Trial) being conducted in North America to evaluate the role of acetazolamide in disease treatment along with weight loss.

One of the frequent concerns that arise with acetazolamide is its use in patients with sulfa allergy. Almost all patients who report this allergy have previous exposure to sulfonamide antibiotics. While acetazolamide also is a sulfonamide derivative, its chemical structure lacks the antigenic moiety associated with allergy [36]. Many patients and even physicians believe that the reaction is to the sulfur-containing portion of the molecule, but this is not correct. Reports of true allergy to acetazolamide in patients with previously documented sulfonamide antibiotic allergy are rare to nonexistent in the literature. It is thus reasonable to offer patients with potential allergy the opportunity to take the first dose of acetazolamide in the office. On rare occasions, consultation with an allergist may be required.

Many neurologists, but not necessarily ophthalmologists, will prescribe topiramate (Topamax) as first-line therapy for PTC. Topiramate is also a sulfonamide derivative whose primary indications are antiepileptigenesis and migraine prophylaxis [37]. Its efficacy in PTC probably derives from its beneficial effects on headache (many PTC patients have migraine-like headache that is not pressure-dependent) and body weight (weight loss occurs in many patients from decreased appetite and increased metabolism) rather than its weak carbonic anhydrase inhibitory activity. A common side-effect of topiramate is cognitive slowing, especially at higher doses (>150 mg/day), and patients will often complain of this effect [38]. Serious adverse ophthalmologic events include acute angle-closure glaucoma from uveal effusion [37].

The best long-term management of primary PTC is weight loss [39]. Loss of just 6% of body weight has been reported to be beneficial [40,41], but for a number of poorly-understood reasons, PTC patients seem to have trouble losing weight despite being highly motivated and undertaking weight loss programs [42]. Bariatric surgery has been used in morbidly obese PTC patients (BMI > 40 or > 35 with obesity-related co-morbidities) with marked success [3,43] although the procedure carries significant short- and long-term risks. We

currently refer all of our PTC patients for nutritional and dietary counseling, and all morbidly obese patients are offered a bariatric surgery consultation.

Surgical management remains second-line treatment in patients and is reserved for those who fail medical therapy or weight loss. Usually it is the patients with acute loss of vision or intractable headache who become surgical candidates. Both optic nerve sheath fenestration (ONSF) and ventriculoperitoneal (VP) shunting should be considered based on the clinical picture. We rarely use lumboperitoneal (LP) shunts at our institution, as our experience demonstrated that VP shunting is more successful in alleviating symptoms and less prone to long-term failure [44]. Others have reported good results with LP shunting, however [45]. ONSF is the procedure of choice where vision loss is the primary concern [46]. Others have reported headache relief after ONSF [47], but the mechanism by which this might occur is unclear. Unilateral ONSF may lead to bilateral improvement in papilledema [48], but this effect is variable, and bilateral sequential surgery is often necessary. Papilledema should start to resolve within days, and complete resolution may be seen by 4-6 weeks. When headache is a more prominent (or the primary) issue, then VP shunting should be favored. A shunt has the advantage of potentially treating both headache and papilledema; headache is more likely to respond to shunting if it is not longstanding (<2 years) [44].

Both ONSF and VP shunting tend to fail with time; persistent CSF drainage in ONSF probably lasts 6 months or less [49-50]. Concerns about optic nerve toxicity have limited the use of mitomycin C or other agents to prevent scarring. It is possible that the scarring itself is beneficial as the optic nerve head would then become insulated from the CSF compartment pressure. Since ONSF is used to prevent acute vision loss, its long-term persistence is less important. VP shunts also tend to fail with time, as almost 50% of patients will require one or more shunt revisions within 5 years. The failure rate does not differ significantly with placement of the distal catheter (peritoneal, pleural, or atrial). Whenever a PTC patient with a shunt experiences recurrent symptoms, shunt failure must be considered and shunt patency studies ordered.

Venous sinus stenting shows promise in the treatment of secondary PTC from venous outflow obstruction. Stenting is most successful where there is focal narrowing of the venous channel without extrinsic compression [51]. For example, patients with superior sagittal sinus meningioma may have marked venous obstruction, but stenting within a mass lesion will not be effective (the tumor will compress the stent, and the lumen will not open significantly). Demonstration of a pressure gradient across the stenosis should precede the stenting procedure, as this measurement helps to confirm that the stenosis is in fact hemodynamically significant [52]. The stent is deployed and the gradient re-measured. Patients must remain on antiplatelet therapy for at least 6 months after the procedure; pregnancy likely should be avoided during this time because of the drugs being used (aspirin and/or clopidogrel).

Conclusion

Pseudotumor cerebri remains a diagnosis of exclusion. When presented with a patient who has bilateral optic disc swelling, presume tumor or other intracranial process until proven otherwise. Make the diagnosis in a timely and proper fashion. Do not ignore secondary causes of PTC (drugs, abnormal anatomy, etc.), and obtain baseline ophthalmic imaging- photos or OCT/HRT/GDx. Do not hesitate to intervene aggressively (medically) or to act surgically (or refer) when vision loss is occurring. Delay can lead to irreversible vision loss. Modern diagnostic methods are taking the "idiopathic" out of IIH- use them!

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Competing Interests

Nil

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REVIEW

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