Intracranial hypertension: Is it primary, secondary, or idiopathic?

Intracranial hypertension is a rare disorder that, if not properly diagnosed, can result in morbidities such as vision loss and chronic pain. The case report by Reddy *et al*. in this issue of the *Journal* is a classic presentation of a patient suffering from intracranial hypertension.[1] She is a middle-aged female with a recent onset of progressively worsening headache, optic edema, transient visual blurring and documented peripheral vision loss. The patient received the proper work up, with initial testing done to rule out a space occupying lesion or infection as the cause of her headaches. The authors then appropriately question whether her intracranial hypertension can be considered secondary to use of dexamethasone, 6 months prior to presentation.

The terminology associated with intracranial hypertension is confusing and frequently used incorrectly. Yet incorrect use can have an impact on a patient's work-up and treatment course. There are two types of intracranial hypertension: Primary and secondary,[2] also known as idiopathic intracranial hypertension (IIH) and intracranial hypertension secondary to (a specific condition), respectively. The differentiation between these two extends beyond merely being an academic exercise.

Primary intracranial hypertension has gone by numerous names since it was first described by Quincke in 1897 and labeled "meningitis serosa".[3] In 1904, Nonne coined the term pseudotumor cerebri because patients often present with symptoms similar to those found in a patient with an intracranial mass.[4] In 1955, Foley suggested the notion of benign intracranial hypertension so as to avoid negative connotations associated with a "pseudo-cancer" diagnosis.[5] In the 1980s, following a series of reports describing vision loss, the syndrome was renamed IIH. Other past names have included otitic hydrocephalus, hypertensive meningeal hydrops, and meningeal hypertension.

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The older terms of pseudotumor cerebri and IIH remain in use, especially among the lay public. Unfortunately both names are often incorrectly applied by medical professionals as well. This misuse is partially the result of the ambiguity inherent in both terms, and has resulted in suggestions to clarify both types by use of the terms primary or secondary intracranial hypertension (PIH and SIH, respectively).[2] Any condition that would result in increased intracranial pressure would be characterized as secondary intracranial hypertension. The expectation is that the current increased pressure would rapidly resolve once the causative condition is corrected. Examples include cessation of tetracycline or growth hormone use, or recanalization of a thrombosed transverse venous sinus. Additional medications and work up may be required in cases of SIH: For example the use of enoxaparin injections and hematologic work up in the case of sinus venous thrombosis. Use of acetazolamide or furosemide should still be considered for a short course to reduce the risk of permanent visual deficits. Treating physicians should monitor these patients closely as once the causative condition is corrected patients can develop intracranial hypotension due to overcorrection from acetazolamide, or similar treatments.

Primary intracranial hypertension is then reserved for individuals who do not have a clear cause for increased intracranial pressure. These individuals may have risk factors for increased intracranial pressure, such as the frequently associated links with puberty, female gender, obesity and polycystic ovarian syndrome.

The patient presented by Reddy *et al*., raises the appropriate question of whether this patient should be considered primary or secondary. The use of steroids has been reported to cause SIH.[6,7] However, in terms of the relation to steroid use and symptom onset in SIH, symptoms appear during treatment or shortly after the steroids are tapered. In this case, there was a 2-month window between the abrupt withdrawal of dexamethasone and development of symptoms. Therefore, as the authors suggest, it would be appropriate to label this patient as having PIH. Moreover, this patient also has the well-known associated risk factor

of obesity with recent weight gain, which in this case was the intended result of self-medication with dexamethasone and cyproheptadine.

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