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ORAL COMMUNICATIONS

Headache in patients with idiopathic intracranial hypertension: a pilot study to assess applicability of ICHD-2 diagnostic criteria

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Abstract Headache is one of the most common symptoms of idiopathic intracranial hypertension (IIH). The aim of this study was to investigate the applicability of the diagnostic criteria for "Headache attributed to IIH" included in the current classification of headache disorders, particularly as far as the main headache features. A consecutive clinical series of IIH patients with demonstration of increased intracranial pressure by lumbar puncture in the recumbent position were enrolled. Among a total of 22 patients, headache was reported by 14. The proportion of patients reporting the main headache features required by diagnostic criteria were: 93 % for daily or nearly-daily occurrence; 71.5 % for diffuse/non-pulsating pain; 57 % for aggravation by coughing/straining. Thus, these three headache features, at least one of which is required for diagnosis of headache attributed to IIH, were present in the vast majority of our sample, suggesting that their inclusion should be regarded as appropriate. The analysis of our results may suggest possible changes in the current ICDH-2 criteria for headache attributed to IIH, based on the following considerations: the existence of remarkable differences as far as the relative frequency of each headache feature; the fact that diffuse and non-pulsating pain—included in the current classification as a single requirement—were not always found together; the high frequency of migrainous associated symptoms (nausea or photophobia–phonophobia were present in 71.5 % cases).

Keywords Idiopathic intracranial hypertension (IIH) · Headache · International Classification of Headache Disorders (ICHD-2)

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Introduction

Idiopathic intracranial hypertension (IIH) is the syndrome of elevated intracranial pressure in the absence of space occupying lesions or other brain disorders [1]. According to modified Dandy criteria for IIH [2], increased opening pressure on lumbar puncture and normal CSF composition together with absence of any known specific cause of increased intracranial pressure are the needed diagnostic requirements.

A diagnosis of IIH is a diagnosis of exclusion, after extensive diagnostic evaluations meant to rule out specific causes. Secondary intracranial hypertension occurs in a large variety of disorders, such as intracranial mass lesions (tumor, abscess), dural venous sinus thrombosis, traumatic brain injury, ischemic or hemorrhagic stroke, dural arteriovenous fistula, hydrocephalus [3]. An intracranial pressure at lumbar puncture in the recumbent position higher than 200 mmH_2O in adults is usually accepted as a marker of IIH diagnosis. Values higher than 100 mmH_2O and than 250 mmH_2O are required in young children and in obese patients, respectively.

Although IIH may be heterogeneous as far as clinical presentation, visual disturbances and headache are the most common symptoms [1–4]. For this reason operational

diagnostic criteria for "Headache attributed to IIH" have been included in the second edition of the international classification of headache disorders, the ICHD-2 [5] (Table 1).

The aim of this study was to investigate the applicability of the ICHD-2 criteria of headache attributed to IIH, particularly as far as the main clinical features of headache, in a sample of patients who were sent to our Neurology Department to verify a suspected diagnosis of IIH.

Patients and methods

We enrolled a consecutive series of patients attending our Neurology Department suspected to have IIH on the basis of their clinical history and/or previous ophthalmological examinations. The presence of headache, and its main characteristics, were systematically assessed and recorded. The presence of papilledema or other visual alterations was assessed by neuro-ophthalmologic examination. To rule out possible causative diseases, all patients underwent: lumbar puncture in the recumbent position, with measurement of intracranial pressure; CSF analysis; neurological and general examination; blood tests; MRI and MRI venography.

Results

Twenty-two consecutive patients (18 women, 4 men; mean age 38.5 years) were enrolled. Among them, 14 (63 %) reported headache. Papilledema was evident in all patients. Increased CSF pressure (<200 mmH_2O in normal weight patients, <250 in obese patients) was found in all the enrolled patients. A normal CSF chemistry was present in all cases. Intracranial mass lesions, dural venous sinus thrombosis, dural arteriovenous fistula, stroke, traumatic brain injury, hydrocephalus were absent in all the enrolled patients.

The progressive character of headache (i.e. a tendency to higher headache frequency during the course of illness) was present in all. As far as the main headache features, we found the following percentages: daily or nearly daily occurrence in 93 % cases (13 patients out of 14); diffuse/non-pulsating pain in 71.5 % (10 patients); aggravation by coughing/straining in 57 % (8 patients). Overall, at least one of these three features was present in all the studied patients: all the three features in 28.5 % (4 patients); two of them in 57.1 % of the sample (7 patients), and only one in 7.1 % (2 patients).

As far as the diffuse and non-pulsating quality of pain, these two characteristics were both present in eight patients, both absent in two, and only one feature was present in four patients.

Associated symptoms were present in many patients: nausea in 71.5 % (10 patients 14); photo-phonofobia in 71.5 % (in 10 patients); both nausea and photo-phonofobia in 21.4 % (in 3 patients).

Discussion

The main headache features required for diagnosis of headache attributed to IIH, were present in the vast

Table 1 International Headache Society diagnostic criteria for "Headache attributed to idiopathic Intracranial hypertension (IIH)" (code 7.1.1 of ICHD-2) [5]

A. Progressive headache with at least one of the following characteristics and fulfilling criteria C and D:
  1. Daily occurrence
  2. Diffuse and/or constant (non-pulsating) pain
  3. Aggravated by coughing or straining

B. Intracranial hypertension fulfilling the following criteria:
  1. Alert patient with neurological examination that either is normal or demonstrates any of the following abnormalities:
    a. Papilloedema
    b. Enlarged blind spot
    c. Visual field defect (progressive if untreated)
    d. Sixth nerve palsy
  2. Increased CSF pressure (>200 mmH_2O in the non-obese, >250 mmH_2O in the obese) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
  3. Normal CSF chemistry (low CSF protein is acceptable) and cellularity
  4. Intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
  5. No metabolic, toxic or hormonal cause of intracranial hypertension

C. Headache develops in close temporal relation to increased intracranial pressure

D. Headache improves after withdrawal of CSF to reduce pressure to 120–170 mmH_2O and resolves within 72 h of persistent normalization of intracranial pressure

majority of our sample, suggesting that their inclusion should be regarded as appropriate.

However, the analysis of the results of this preliminary survey may suggest possible changes in the current ICDH-2 diagnostic criteria for "Headache attributed to IIH", based on the following considerations.

Remarkable differences were found as far as the three headache features, at least one of which is required for diagnosis, as daily occurrence was reported by almost all patients, while aggravation of headache by coughing/straining and the diffuse/non-pulsating quality of pain were present in a lower proportion (<60 and around 70 %, respectively).

In the current diagnostic criteria, the presence of diffuse/non-pulsating quality of pain is included as a single requirement. We note that the term "diffuse" may be not appropriate, and could be changed in "bilateral". Furthermore, the two characteristics could be split in two different features, as in our sample they were not always found together (4/14 patients reported only one of them).

Moreover, as far as the "progressive" nature of headache, this feature should be better defined. It is not clear if an episodic presentation before the daily occurrence of headache must be reported in patient's history, or if a progressive increase in headache severity is needed to make a diagnosis.

It is also interesting to note that some associated symptoms—the presence of which is not included among the current criteria—were present in many patients: nausea and-or photo-phonofobia were reported by around 70 % patients, although both symptoms were reported in 21.4 %. The high frequency of these "migrainous" accompanying symptoms, together with the pulsating quality and unilateral distribution of pain in some patients (in 28.5 and 14 %, respectively), may suggest a migraine-like presentation of headache in (a relevant proportion of) IIH patient, as previously reported [2, 4]. This topic is very intriguing, also considering the possible correlations between IIH and migraine progression postulated in the literature. Both clinical and instrumental findings indicate IIH—particularly IIH without papilledema—as a risk factor for migraine progression to chronic, often refractory forms [6, 7]. In fact, considering all the above-reported symptoms together with the usual daily occurrence of pain in 50 % of our IHH patients with headache, we note that a misdiagnosis of "primary" chronic migraine might be possible in patients who may in fact suffer from a "secondary" headache form.

We think that our findings will be useful in promoting further studies meant to assess headache characteristics on larger samples of IIH patients, to reach a better understanding of IIH and to evaluate a possible revision of current ICDH-2 diagnostic criteria for "Headache attributed to IIH".

Conflict of interest The authors certify that there is no actual or potential conflict of interest in relation to this article.

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