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Case report

Headache with Isolated Sixth Cranial Nerve Palsy in an Overweight Woman: A Case of Idiopathic Intracranial Hypertension

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***Abstract*— Idiopathic intracranial hypertension (IIH), also known as pseudotumour cerebri, has an estimated incidence of 1 to 2 in 100,000 people per year. IIH commonly affects obese young women, where its pathogenesis, however, is not clearly understood. Patients present to the emergency department with a wide array of presentations. In light of this, we present a case of isolated sixth cranial nerve palsy in a non-obese young woman due to IIH in which brain imaging studies showed no abnormality; however, lumbar puncture reveals an opening pressure of more than 50 cmH2O. This case would help to alert physicians to consider IIH as one of the differential diagnoses when encountering such cases, as an uncommon presentation in the non-obese patient, and prompt referral is important to avoid unnecessary delay in the treatment of IIH.**

Keywords— intracranial hypertension, headache, diplopia, abducens nerve palsy

I. INTRODUCTION

Idiopathic intracranial hypertension (IIH, pseudotumour cerebri) is a neurological condition that is known to cause raised intracranial pressure without the presence of a tumour or other disease. It is a diagnosis of exclusion, occurring with higher frequency in obese, reproductive women, though it could occur in men and children too. The highest incidence, 28 per 100,000 per year, was reported in Ireland [1].

This case report highlights the importance of the identification of IIH as a diagnosis of exclusion. It also should not be missed by primary care nor emergency physicians and must be considered as a differential diagnosis in women of reproductive age presenting with a disabling headache, even if they are not obese. It must be mentioned that a prompt referral, which was applied in this case, saved the patient from unnecessary delay in diagnosis.

II. CASE

A 24-year-old Malay lady with no known medical illness and no known drug allergy was presented to our emergency department after an ophthalmology outpatient assessment. She complained of a headache for the past two weeks, which is sudden in onset and progressively worsening. The headache was throbbing in nature over the right side at the temporal region and was reported to be worse in the morning. It was also associated with double vision for a duration of two weeks, with no preceding aura. There was no recent travelling history or

head trauma. Her mental health status was normal, and the headache was not precipitated by any dairy products, caffeine, or chocolate. Otherwise, she was not on any hormonal therapy or traditional medication and denied illicit drug use, alcohol, or smoking. Her menstrual cycle was at a 28–30-day interval with normal flow. There were no constitutional symptoms such as prolonged fever, weight loss, or loss of appetite. There is no family history of malignancy, IIH, or other neuro-genetic problems. She initially sought treatment at the local health clinic, where the doctor subsequently referred her to an ophthalmology clinic for further assessment.

On examination, she was alert, conscious, and oriented from time to time, place, and person. Her blood pressure was 117/82 mmHg with a body mass index of 24.4 kg/m^2 (weight of 66, height of 1.65 meters), which was overweight, and she was afebrile. Her cranial nerves examination revealed right sixth cranial nerve palsy, as shown in Fig. 1, along with diplopia. Otherwise, the neurological examination, including tone, power, sensation, and reflexes, was normal. Signs of meningeal irritation were absent.

Fig. 1 Right eye adducted on primary position.

She underwent a plain computer tomography (CT) scan of the brain that revealed no focal lesion, as shown in Fig. 2, and was subsequently admitted to the medical ward for further necessary evaluation. A lumbar puncture (LP) was done and showed an opening pressure of more than 50 cmH_2O (high opening pressure) and a closing pressure of 12 cmH_2O. The cytology and biochemical findings of the LP were otherwise within normal range, with a total white cell of $8.6x10^9/L$. She was treated with oral acetazolamide 500 mg bd and subsequently subjected to contrast-enhanced CT and CT venogram that both reported as normal. Her treatment included a low-salt diet and weight loss advice. She was discharged home with an appointment to the neurology clinic in one month. During this visit, the headache resolved with no residual cranial nerves' abnormalities. Further documentation of the patient's case could not be retrieved to determine the duration or if any up-titration of the dose was done for acetazolamide.

III. DISCUSSION

IIH is the most common cause of papilloedema in the outpatient setting [2]. With the increasing rate of obesity around the world, the incidence and prevalence of IIH have been rising since the early 21st century [3,4].

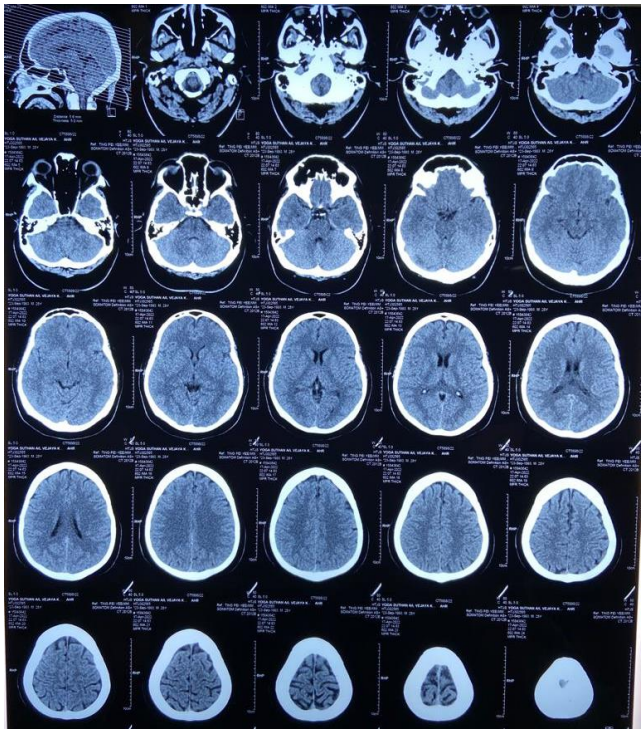

Fig. 2 A CT brain film of this patient that shows no abnormal findings.

Typical symptoms include chronic severe headaches, visual disturbance, and rarely permanent visual loss occurring in young women of reproductive age [5]. Pulsatile tinnitus and brief visual disturbance are two possible symptoms of IIH [6]. Some risk factors known to be related to IIH include obesity, female gender of reproductive age, including medications such as growth hormones, tetracyclines, and retinoids [4]. However, special attention should be given to this case as the patient is clinically not obese, although she was overweight and was not taking any hormonal treatment or other possible drugs. Only a small number of literature reports about IIH in non-obese young women. Other instances of rare cases of IIH are familial or genetic predisposition, including atypical demographics such as men and older people [7, 8]. One study reported that IIH may be diagnosed in older patients with normal Body Mass Index (BMI) because there is a lower suspicion index in the normal BMI population [8]. Associated conditions related to IIH were also reported, such as systemic diseases, vitamin deficiencies, or excess and anaemia; however, they were not directly linked. The modified Dandy criteria can be applied to diagnose IIH. Headache, pulsatile tinnitus, papilloedema, and high CSF pressure (more than 25 cmH2O) are among the classical findings. However, as in this case, not all criteria are usually fulfilled [9, 10, 11].

Three primary intracranial processes have been proposed as aetiologies of elevated intracranial pressure, which are increased venous sinus pressure, reduced CSF outflow, and increased CSF production in the choroid plexus. Female gender and obesity are two risk factors for idiopathic intracranial hypertension, implying that sex hormones and substances generated by adipose tissue, such as adipokines (cytokines released mostly by adipose tissue), may have a role in CSF dynamics. Other processes, such as an increase in the production of inflammatory or thrombophilic substances, might also be at work. Obesity has also been linked to an

increase in intrathoracic pressure, which, when combined with venous sinus stenosis, might lead to an increase in venous sinus pressure. However, stenosis is more likely to occur as a result of increased venous pressure than as a result of a blockage [11].

There is little information available on the aetiology of IIH, which causes abducens nerve palsy. However, it is thought that the abducens nerve's participation in IIH is owing to its anatomical predisposition. Dorello's canal, also known as the petroclival segment, is located in the transition between the posterior and middle cranial fossa. This canal is fused superiorly by a bony arch, the union of the petrous apex (spine), and the accessory clinoid process (ACP). The non-union of the petrous apex and ACP is replaced by an elastic fibro-osseus ligament, the petrosphenoidal ligament (PSL), covering the roof of the foramen. High ICP squeezes and compresses the PSL, constricting the sixth cranial nerve in Dorello's canal, resulting in a sixth nerve palsy [10].

The sixth cranial nerve is unique: it is a pure motor nerve, the second-longest cranial nerve after the trochlear nerve, entirely dedicated to supplying the lateral rectus muscle, thereby allowing abduction of the eye. When ICP is elevated, there is a downward displacement of the brainstem that stretches CN VI as it crosses over the petrous ridge and enters Dorello's canal. Approximately 12% of adults with IIH develop sixth nerve palsy [10].

In the approach to patients with suspected IIH, imaging investigations need to be done to rule out any structural lesions. Imaging studies of the brain must demonstrate the absence of any focal parenchymal lesion concurrent with elevated intracranial pressure by a lumbar puncture. Furthermore, the CSF results, i.e., cytology and biochemistry, must be normal, which was seen in this case that aided in the diagnosis of IIH [12].

The mainstay of the treatment is actually to alleviate the symptoms and prevent the worsening of the symptoms. In this case, the patient was treated with oral acetazolamide, and the symptoms resolved within one month. Final visual outcomes are usually similar between typical and atypical presentations, but visual field loss at presentation plays a crucial role in predicting the prognosis of visual outcomes in all patients [8]. There are no specific protocols or guidelines for the management of IIH; however, it is concluded that acetazolamide is effective and safe for IIH treatment and that the benefits of acetazolamide can be seen within the first 4-6 weeks of therapy [9, 13-15]. Some physicians do keep patients on acetazolamide for a few months before tapering off the dose despite seeing good results early on [15].

Iv. CONCLUSIONS

Young, overweight women presenting with headaches, temporary visual complaints, with cranial nerve abnormality should be suspected of IIH. There is a possibility of genetic predisposition of IIH rather than the usual obesity or gender-related. Emphasis on IIH as a diagnosis of exclusion must be reinforced; hence, imaging investigations are generally recommended to rule out other structural and obstructive problems. A high index of suspicion for IIH must be observed in the outpatient clinics so that early investigation and treatment could be initiated. The prognosis of the final visual outcome is dependent on whether the patient presented with visual field loss or not, as in this case, the patient did not, leading to good treatment response and recovery.

CONSENT

Written informed consent was obtained from the patient for the anonymized information and partially covered picture to be published in this article.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this paper.

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