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Case Report

Idiopathic Intracranial Hypertension With Unusual Presentation in an Adolescent Girl: A Case Report

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ABSTRACT

Background: Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri is a rare disorder. Regarding the latest diagnostic criteria for this disorder, we present a rare case of IIH.

Case presentation: The case was an 11-year-old girl referred to our center with generalized body pain and mild left-eye deviation with no other visual symptoms. She had a severe generalized pain especially in her back and prevertebral area. In the neurological examination, there was a mild left sixth nerve palsy and bilateral papilledema. We treated the patient with acetazolamide and to lumbar puncture, which reduced intracranial pressure. The generalized body pain was reduced gradually in ten days, eye deviation in three weeks, and papilledema in two months after treatment.

Conclusion: Based on the serious complications of IIH, a thorough assessment of suspicious cases, including ophthalmoscopic examinations, is mandatory. Furthermore, clinicians should consider even rare and unknown specific symptoms in these patients.

Keywords: Pseudotumor cerebri, Idiopathic intracranial hypertension, Child

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Highlights

• An 11-year-old girl had idiopathic intracranial hypertension (IIH), with no symptom or sign of increased intracranial pressure, except 6th nerve palsy.

• Final diagnosis of IIH was done due to papilledema, normal biochemistry and cytological cerebrospinal fluid (CSF), increased CSF pressure, and normal neuroimaging.

• The chief complaint of the patient with IIH was severe generalized body pain causing sleep disturbances.

Introduction

Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri is an rare disorder in children with a prevalence of 1 in 100,000-150,000 children [1]. This syndrome includes elevated intracranial pressure with no mass lesion, hydrocephaly, and cerebrospinal fluid (CSF) pleocytosis in the absence of systemic, infectious, endocrinological, metabolic, and hematological diseases or consuming specific drugs [2]. The definite pathogenesis of IIH is still unknown, which can have no underlying causes or be secondary to the aforementioned parameters [3]. Children with IIH often have the symptoms of elevated intracranial pressure, such as papilledema; however, they sometimes may have the symptoms without papilledema [4]. Headache is the most common symptom of IIH, with a prevalence of more than 90% [5]. Other symptoms are vomiting, nausea, diplopia, and blurring [6].

The IIH is diagnosed by the modified DANDY criteria including symptoms and signs of increased intracranial pressure (e.g. headache, papilledema, visual symptoms, vomiting, nausea, tinnitus), no focal neurological signs (except the 6th nerve palsy), increased CSF pressure >25 cm H_2O, and no other causes for increased intracranial pressure [7]. Considering the improvement of technology in recent years and a more precise understanding of the pathophysiology of IIH, these criteria were revised, and the diagnosis is made by the updated criteria [4]. In this study, we aim to report a rare case of IIH in a girl. This report can help clinicians become familiar with this disorder to prevent its serious complications.

Case Presentation

In this case report, we presented an 11-year-old girl referred to our center with generalized body pain and mild left-eye deviation in the absence of other visual symptoms. In her past medical history, she had an influenza-like symptom three weeks before her admission. In the recent week, she was afebrile, with no headache, and any constitutional symptoms. There was no considerable personal and family medical history. In the physical examination, she had stable vital signs and normal body mass index. She had a severe generalized pain especially in her back and prevertebral space. In the neurological examination, there was a mild left sixth nerve palsy and bilateral papilledema. There were no laboratory findings in favor of inflammatory process such leukocytosis, increased C-reactive protein, or erythrocyte sedimentation rate. There was a normal brain, spinal, and orbital magnetic resonance imaging (MRI) results, as well as a normal brain magnetic resonance venography. The patient had undergone an MRI with and without gadolinium. The MRI scan showed small ventricle without any other abnormality. In the lumbar puncture, despite a normal cytological and biochemical CSF, increased CSF pressure was noted (42 cm H_2O). The detailed results of CSF were as follows: White and red blood cell counts=zero, protein=30 mg/dL, glucose: 80 mg/dL (simultaneous blood sugar was 100 mg/dL). The CSF culture was negative. The perimetry showed no significant finding. Therefore, we performed the diagnostic procedures for ruling out other secondary causes of intracranial hypertension [8], and consequently IIH was indicated as a definite diagnosis.

We treated patient with acetazolamide, which led to decrease in intracranial pressure due to lumbar puncture (LP) and treatment. The generalized body pain decreased gradually in 10 days, eye deviation decreased in three weeks, and papilledema decreased in two months after treatment. We did not perform second LP due to the improvement of clinical symptoms after administering acetazolamide and the first LP and the parents' unwillingness to allow performing another LP.

Discussion

The IIH is an uncommon disorder in children with an unknown pathophysiology. Its possible causes are aberrant CSF flow, CSF production, or both of them [9]. Headache is the most common symptom in children with IIH. Other common symptoms are related to increased intracranial pressure which can be different in terms of age [10]. Based on the updated criteria for diagnosing IIH, it can be occurred without headache [11], or with neck stiffness [12] and torticollis [13]. Since the patient in our study had a generalized body pain without headache, we considered aberrant CSF flow, CSF production, or both of them as a possible pathophysiology for the absence of headache with high intracranial pressure.

In this case report, there was no symptom or sign of increased intracranial pressure, except 6th nerve palsy. In the beginning, we considered isolated cranial nerve palsy due to left painless nerve palsy and the female gender; however, we finally diagnosed IIH due to papilledema, normal biochemistry and cytological CSF, increased CSF pressure, and normal neuroimaging. It is noteworthy that the chief complaint of our patient was severe generalized body pain which caused sleep disturbances. Regarding the pathogenesis of unusual symptoms, it should be noted that these associated symptoms are due to forced filling of the nerve roots with CSF irritating or compressing the nerve root fibers inside.

All symptoms in the patients were improved after lumbar puncture and administering acetazolamide. This finding not only show the promising effect of the drug, but also can be a possible result of lumbar puncture changing CSF dynamics to readjust the pressure. The results of this case report are consistent with the findings of previous studies reporting uncommon symptoms for IIH [11-13].

Conclusion

Based on the serious complications of IIH, a thorough assessment of suspicious cases including ophthalmoscopic examinations, is mandatory. Furthermore, clinicians should consider even rare and unknown specific symptoms in these patients.

Ethical Considerations

Compliance with ethical guidelines

This study was approved by the Ethics Committee of Guilan University of Medical Sciences (Code: IR.GUMS.REC.1402.262).

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Authors contributions

The authors contributed equally to preparing this article.

Conflict of interest

There is no conflict of interest to be declared.

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