

CASE REPORT

Pseudotumor Cerebri Presenting by Neck Rigidity and Torticollis

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Abstract

Pseudotumor cerebri syndrome (PTCS) is an uncommon disease in children. On-time diagnosis and treatment can prevent irreversible visual loss. Although headache is the most common complaint of children with this syndrome, the present case report reported a child with neck rigidity and torticollis, declined by the reduction of intracranial pressure. Despite the importance of torticollis and neck rigidity presented in various significant neurological disorders in need of thorough investigations, in the case of unexplained symptoms of those disorders, it is recommended to consider fundoscopic examinations for PTCS to prevent its vital complications.

Keywords: Pseudotumor Cerebri; Torticollis; child

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Introduction

Pseudotumor cerebri syndrome (PTCS) is a clinical syndrome that has normal brain structure and cerebrospinal fluid (CSF) analysis, even with the presence of signs and symptoms of increased intracranial pressure (ICP). PTCS may occur after certain medical conditions or medications. However, in the case of no definite cause for PTCS, it is defined as idiopathic intracranial hypertension (IIH) (1). Headache is the most common symptom of PTCS (2). Other symptoms are visual complaints (3), tinnitus, dizziness, and ataxia (4). Besides, abducens nerve palsy is the most common sign of cranial neuropathy (5). Neck rigidity is a common complaint in children. Consequently, it occurs in an abnormal or normal neck position without the ability to move it, with various causes ranging from benign to life-threatening conditions, such as meningitis, subarachnoid hemorrhage, and posterior fossa tumor (6). Torticollis is a pathologic condition with a Latin root encompassing tortus (twisted) and collum (neck) (7) that can be congenital or acquired.

In addition, torticollis can be classified as dynamic (paroxysmal) and non-dynamic (non-paroxysmal) types. The causes of non-dynamic type are congenital muscle, bone, central nervous system, peripheral nervous system, ocular, and soft tissue disorders. Conversely, the dynamic type includes paroxysmal benign torticollis, Sandifer syndrome, medications, and increased ICP. Other substantial causes are the brain and spinal tumors (8). In this case report, consistent with previous limited case reports, the researchers aimed to present a patient with neck rigidity and torticollis as the symptoms of increased ICP to emphasize early diagnosis and treatment of patients with PTCS along with these rare and uncommon symptoms.

Case presentation

A 7-year-old girl with pain and neck stiffness from seven days before hospitalization, a history of two times vomiting, eye deviation, and diplopia from two days before hospitalization and torticollis from a day before hospitalization, was referred to the pediatric hospital. In the general physical examination, limitation of the motion of the neck

on each side was only noted. In the neurological examination, the patient was alert, had papilledema grade 3 and bilateral abducens nerve palsy in the cranial nerve examination, and the remaining neurological assessments were normal. She had a normal brain CT, MRI, MRV, and cervical spine MRI. CSF analysis was normal, but CSF opening pressure was 50 cmH₂O. Based on table 1, she was diagnosed with PTCS, and thorough assessments of the etiology were performed based on Table 2 (9). As no abnormalities were indicated based on the above tables, IIH was mentioned as the definite diagnosis. Directly after the diagnosis of PTCS, acetazolamide with a dose of 10 mg/kg was initiated and continued to 30 mg/kg/day to prevent visual loss. Consequent to taking 6cc of CSF and administering acetazolamide, neck stiffness and torticollis improved considerably in three days. Diplopia disappeared utterly, and no sign of papilledema was mentioned after three months. Considering the disappearance of all signs and symptoms, repeated LP and measuring CSF pressure were not necessary.

Table 1. Diagnostic criteria for idiopathic IIH

Diagnosis of IIH	Diagnosis of IIH without papilloedema
Diagnosis of IIH is definite if the patient fulfils A–E A. Papilloedema. B. Normal neurological examination except for sixth cranial nerve abnormalities. C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass or structural lesion, and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used. D. Normal CSF composition. E. Elevated lumbar puncture opening pressure (≥ 250 mm CSF in adults) in a properly performed lumbar puncture.	In the absence of papilloedema, a diagnosis of IIH can be made if B–E are satisfied, and in addition the patient has unilateral or bilateral abducens nerve palsy. In the absence of papilloedema or sixth nerve palsy, a diagnosis of IIH can be suggested but not made if B–E are satisfied, and in addition at least 3 of the following are present on neuroimaging: 1. Empty sella. 2. Flattening of the posterior aspect of the globe. 3. Distension of the perioptic subarachnoid space with or without a tortuous optic nerve. 4. Transverse venous sinus stenosis.

Table 2. The secondary causes of PTCS

Pharmacological agents	Systemic conditions
Antibiotics: tetracycline and derivatives, vitamin A derivatives: isotretinoin, all-trans-retinoic acid (for acute promyelocytic leukaemia) Hormonal agents: corticosteroid withdrawal, growth hormone, thyroxine replacement in children Other agents: lithium, nalidixic acid, rofecoxib, cimetidine	Haematological: anaemia Respiratory: obstructive sleep apnoea Renal: renal failure Endocrine: obesity, weight gain, polycystic ovarian syndrome, Cushing's disease, Addison's disease, hypoparathyroidism Genetic: turner syndrome Autoimmune: systemic lupus erythematosus Nutritional: hypervitaminosis A Venous: cerebral venous sinus thrombosis, superior vena cava obstruction, increased right-sided heart pressure

Discussion

PTCS, with an estimated incidence of 0.63/100000, is an uncommon disease in children (10). Although headache is the most common symptom, other uncommon symptoms have been reported by previous case reports worldwide, emphasizing the early diagnosis and treatment of PTCS. Neck rigidity (11-12) and torticollis (13) are uncommon symptoms. It has a complex pathophysiology, but it seems to occur due to aberrant CSF flow, CSF production, or both (9). In the presented case, the main symptoms were neck rigidity, torticollis, and

the limitation of neck mobility, and all of these symptoms were improved by lumbar puncture and acetazolamide administration. Although the primary mechanism of neck rigidity as a symptom of PTCS in children is unknown, the alleviation of neck pain, motion limitation and rigidity, and finally, torticollis consequent to taking CSF and administering acetazolamide (reducing the CSF production) can strongly demonstrate the direct relation between symptoms and increased ICP (13). In other words, a mechanical mechanism was probably available following intracranial

hypertension, indicating the reduced severity of symptoms due to the decreased ICP. Regarding the role of serotonin and cytokines in neck pain and rigidity in meningitis (due to the congestion and dilatation of meningeal vessels and the stimulation of pain receptors by the mentioned chemical mediators), possibly, their consumption and the cessation of production may be noted as a related mechanism (6).

This study, similar to limited previous case reports, indicated that torticollis and neck rigidity could be the symptoms of ICP. Therefore, it is recommended to consider PTCS in the case of unexplained symptoms mentioned above, even in the absence of a headache. Besides, the physician has to emphasize performing fundoscopic examination to rule out PTCS because on-time diagnosis and treatment can prevent irreversible blindness.

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Author's contribution

Afagh Hassanzadeh Rad: Substantial contributions to the conception, design of the work; the acquisition, analysis, interpretation of data for the work, Drafting the work, Final approval of the version to be published, Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy
Vahid Aminzadeh: Substantial contributions to the conception, design of the work; the acquisition, analysis, interpretation of data for the work, Drafting the work, Final approval of the version to be published, Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy

Conflict of interest

The authors declare that there is no conflict of interests.

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