소아에서 급성 신부전을 동반한 자발성 두개내 저압 1예

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A Case of Spontaneous Intracranial Hypotension Accompanied by Acute Kidney Injury in a Child

Spontaneous intracranial hypotension in childhood is rare, and a few cases have been reported as a cause of headache in children. A 9-year-old boy was admitted to our hospital with a 3-day history of new-onset headache that worsened upon standing or walking, and aggravating low back pain. No medical history of injury, connective tissue disorder or migraine was detected. A neurological examination revealed neck stiffness. His initial blood tests suggested acute kidney injury by increased blood urea nitrogen (BUN) and creatinine. Brain computed tomography (CT) and cerebral spinal fluid (CSF) analysis were normal; however, opening pressure was low (<60 mm H₂O). Magnetic resonance imaging (MRI) of the spine showed a collection of cerebral spinal fluid in the dorsal extradural space throughout the entire thoracic and lumbar spine level. The patient was diagnosed as having spontaneous intracranial hypotension accompanied by acute kidney injury. Magnetic resonance myelography and spinal MRI performed 14 days later did not show any cerebrospinal fluid leak. The headache and back pain were alleviated with strict bed rest and hydration. He remained free of headache and back pain at the 2-month follow-up. Here, we report a case of a 9-year-old boy with spontaneous intracranial hypotension.

Key Words: Intracranial hypotension, Child, Headache, Acute kidney injury

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Introduction

Spontaneous intracranial hypotension is an extremely uncommon disease, with an estimated incidence of 5/100,000 adults/year¹⁾. Spontaneous intracranial hypotension occurs more frequently in women than in men, especially middle-aged women²⁾. The exact incidence of spontaneous intracranial hypotension in childhood has not been known. Spontaneous intracranial hypotension is characterized by a headache that develops on an upright position and is improved on lying down³⁾. However, its clinical manifestations have a wide spectrum in children, and it can be difficult to diagnose.

There have been few case reports on intracranial hypotension in childhood, which are usually associated with connective tissue disorder, especially Marfan syndrome^{4,5)}. In this report, we describe a 9-year-old child with spontaneous intracranial hypotension without any history of connective tissue disorders.

Case Report

A 9-year-old boy was admitted to Inje University Busan Paik Hospital with a 3-day history of new-onset headache and aggravating back pain. The headache worsened upon standing or walking. He also had vomiting and headache, but no fever, sensory or motor symptoms. There was no significant past medical history. There was no preceding history of physical exertion and trauma. There was no family history of connective tissue disorders or aortic aneurysm.

On physical examination, the patient complained of tenderness on his lower back area and neck stiffness on flexion. Focal neurological deficits were not detected. His joints showed a normal range of movement. He had no skin hyperextensibility. His height was 131 cm (10–25 percentile) and his body weight was 25 kg (25–50 percentile). His blood pressure was 90/50 mm Hg, pulse rate 92/min, body temperature 36.7°C and respiratory rate 26/min. The initial serum blood urea nitrogen (BUN) was elevated at 37 mg/dL, and creatinine was raised at 2.87 mg/dL. The urinary output was not reduced. The urine specific gravity was within the upper normal limit of 1.022. His kidney-ureter-bladder ultrasonography was normal. After hydration, the serum BUN and creatinine levels obtained 5 days later were 15 and 1.03 mg/dL respectively, both of which had been normalized. He was diagnosed as having a prerenal type of acute kidney injury caused by dehydration due to poor oral intake and vomiting.

Even though he restored renal function, his headache, back pain and neck stiffness continued to remain. The brain computed tomography (CT) showed no abnormal density or enhancing lesion in the brain parenchyma. A lumbar puncture was performed, which was suspected of meningitis. The cerebrospinal fluid (CSF) analysis revealed a white blood cell (WBC) count of 0, a red blood cell (RBC) count of 1, a protein level of 36.4 mg/dL and a glucose level of 61 mg/dL. Blood, urine and CSF cultures showed no growth, and CSF polymerase chain reaction (PCR) for herpes simplex virus types 1 and 2, varicella zoster virus, and enterovirus was negative. However, CSF opening pressure was noted to be very low, with less than 60 mm H₂O. Magnetic resonance imaging (MRI) of the whole spine was immediately performed for the evaluation of back pain and meningeal irritation signs. Spinal MRI revealed an extensive collection of CSF in the dorsal extradural space throughout the entire thoracic and upper lumbar spine level, which was not matched with the lumbar puncture site between the third and fourth lumbar vertebrae (Fig. 1A, 2A).

The patient was diagnosed with spontaneous intracranial hypotension accompanied by acute kidney injury. The headache

and back pain were alleviated with strict bed rest and hydration. Magnetic resonance myelography and spinal MRI performed 14 days later did not show any CSF leakage (Fig. 1B, 2B). He was discharged in a stable condition without any neurological deficits. He remained free of headache and back pain at the 2-month

Fig. 1. Sagittal T2-weighted magnetic resonance imaging (MRI) of the spine shows a collection of cerebrospinal fluid in the dorsal extradural space throughout the entire thoracic and lumbar spine level (A, arrow). Follow-up spinal MRI taken 2 weeks later shows improvement (B).

Fig. 2. Axial T2-weighted MRI of the thoracic spine reveals a collection of cerebrospinal fluid in the dorsal extradural space (A, arrow). Follow-up of spinal MRI taken 2 weeks later shows no cerebrospinal fluid leak at the same level (B).

follow-up.

Discussion

Spontaneous intracranial hypotension is an uncommon cause of headache in children⁽⁶⁾. In the past, spontaneous intracranial hypotension was considered an exceedingly rare disorder, so that it was frequently underdiagnosed⁽²⁾. Since introduction of MRI, it has been increasingly diagnosed and considered a potential cause of postural headache^(1,2,7). To the best of our knowledge, this is the first case report of spontaneous intracranial hypotension in childhood in Korea.

Spontaneous intracranial hypotension is due to the leakage of CSF from a dural tear or dural weakness without any history of critical trauma or dural punctures^(4,8). Although the cause of spontaneous CSF leak is unknown, dural fragility can be induced as a result of underlying conditions such as connective tissue disorders or minor traumas^(1,9). Most of the dural spontaneous tears often occur at the exit zone where the cervical and thoracic spinal roots leave the subarachnoid space^(1,10,11). Other leakage sites may include the cribriform plate or the pituitary fossa, and the leakage can occur from one or more sites^(1,3,10). The tear sites tend to have a larger diameter than those caused by iatrogenic needle injuries that can result in extensive CSF leakage involving several spine levels⁽⁹⁾. Our patient also showed a collection of fluid extending along several spinal levels, which was not matched with spinal puncture site.

The most common symptom of spontaneous intracranial hypotension is postural headache that is relieved by lying down, but rapidly worsens when the patient sits or stands. Other symptoms include posterior neck pain and stiffness, nausea, vomiting, diplopia, photophobia, hearing loss, ataxia, short-term memory loss, or neck pain^(1,4,6). Our patient presented with typical clinical features including postural headache, neck stiffness and vomiting. CSF depletion is the main cause of the symptoms. It allows for brain descent leading to traction of pain-sensitive structures: consequently, the pain worsens in an upright position because the brain sags more in that position^(4,12). Low intralabyrinthine pressure is considered to be associated with auditory and vestibular symptoms^(8,13).

The diagnosis can be confirmed by MRI or when CSF opening pressure is low (less than 60 mm H₂O) in the presence of an orthostatic headache^(1,3). Brain MRI has some characteristic imaging features of spontaneous intracranial hypotension such as subdural fluid collection, diffuse pachymeninges enhancement, engorgement of venous structures, decrease in the size of the

ventricles, enlargement of the pituitary and sagging of the brain^(2,11). However, about 20–30% of the patients can have normal findings on cranial MRI⁽³⁾. In our case, although brain MRI was not taken, brain CT showed no evidence of subdural fluid collection, obliteration of subarachnoid cisterns, ventricular collapse or any lesions in the brain parenchyma. The abnormal findings of brain MRI can be improved within days to weeks with treatment of CSF leak⁽²⁾. Spine MRI abnormalities in spontaneous intracranial hypotension are extra-arachnoid fluid collection, extradural extravasation of fluid, meningeal diverticula, spinal dural enhancement and engorgement of the spinal epidural venous plexus^(11,13-17). Our patient's whole spine MRI showed an extensive retrospinal fluid collection throughout the entire thoracic and lumbar spine levels. Radioisotope cisternography and CT myelography can also be helpful in localizing the site of cerebrospinal fluid leak⁽³⁾.

Most symptoms can resolve spontaneously after conservative management such as bed rest, hydration, and use of intravenous caffeine, steroid or theophylline⁽¹²⁾. If these approaches fail, an autologous epidural blood patch can be considered^(1,3). The epidural blood patch improves the symptoms by replacing CSF volume with blood volume within the spinal canal and by forming a dural tamponade which can seal the leak⁽²⁾. If the exact site of CSF leak is detected, a direct epidural blood patch or fibrin sealant can be considered. However, if nonsurgical measures fail, surgical repair of CSF leak can also be considered. Our patient improved after strict bed rest and hydration. The patient's magnetic resonance myelography and spinal MRI taken 2 weeks later did not show any CSF leakage.

Our patient had typical clinical features including postural headache, back pain and neck stiffness before the dural puncture. Opening pressure was less than 60 mm H₂O, and an immediate spinal MRI revealed an extensive CSF leak involving several spine levels. He also had 3-day history of vomiting, nausea and poor oral intake. His serum BUN and creatinine levels presented prerenal type acute kidney injury and after hydration, the serum BUN and creatinine levels had been normalized. His kidney-ureter-bladder ultrasonography was normal. He presented initially prerenal type acute kidney injury due to poor oral intake and vomiting which were the symptoms of spontaneous intracranial hypotension. The cause of his dural leak was not identified; however, it is common in children with spontaneous intracranial hypotension. His symptoms and imaging findings improved after conservative measures. Spontaneous intracranial hypotension is not commonly observed in children; however, it can be well recognized by neuroimaging and clinical suspicion. Therefore, our case suggests that spontaneous intracranial hy-

potension should be considered a possible cause in children who present with orthostatic headache and meningismus without any infection signs.

요약

자발성 두개내 저압은 주로 중년 여성에서 발병하고 앉거나 일어설 때 악화되는 두통이 주 증상으로 나타나며, 소아에서는 흔하지 않은 질환이다. 대개는 결제조직 질환에서 발생하는 경우가 많으나 이 증례의 환아와 같이 특별한 원인을 밝힐 수 없는 소아 환아가 전체 환아들의 반 수 이상을 차지한다. 본 증례의 환아에서는 3일간 지속된 두통을 주소로 내원하였다. 두통은 주로 서 있거나 걸을 때 악화되었으며 허리와 뒷 목의 통증을 동반하였다. 목을 앞으로 굽혔을 때 목과 허리의 통증이 악화되었으며 뻣뻣하였고 늑골척추각의 압통을 보였다. 뇌컴퓨터단층촬영 및 뇌척수액 분석검사에서는 특이소견이 없었으나, 뇌척수액 초압이 60 mmH₂O 미만으로 매우 낮았다. 척수의 자기공명영상에서 흉추와 요추를 따라서 광범위하게 등쪽 경막외공간에 뇌척수액이 저류되어 있었다. 환아의 혈액검사에서는 급성신손상의 소견이 있었으나 신장초음파에서 구조적 이상은 없었다. 환아는 자발성 두개내 저압의 증상으로 탈수가 진행하면서 발생한 급성신부전으로 진단되었다. 절대적인 침상 안정과 수액 요법으로 환아의 증상은 호전되었다. 약 2주 후 시행한 척수 자기공명영상에서 더 이상 뇌척수액의 누출은 없었다. 외상이나 특이 병력이 없는 소아 환아에서 자세 변화에 따른 기립성두통이 발생하였을 때 두통의 드문 원인인 하나 자발성 두개내 저압을 의심하여 뇌척수액의 압력 측정 및 영상의학적 검사를 고려해야 할 것이다.

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