2세 남아의 특발성 경부척수공동증

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Idiopathic Cervicosyringomyelia in 2 year-old Boy

Syringomyelia is a disorder in which a cavity has formed within the spinal cord. Idiopathic syringomyelia is not associated with identifiable causes such as Chiari type 1 malformation, spinal cord tumor, vascular malformation, tethered cord, arachnoiditis, hydrocephalus, or previous spinal surgery. The main neurologic symptoms of idiopathic syringomyelia are toe-walking, constipation, incontinence, abnormal reflexes, and lower extremity weakness. Patients may present with various symptoms such as scoliosis, cutaneous markers, pain in the lower extremities or back, or may be asymptomatic. Herein, we report a young child with idiopathic syringomyelia presenting with subtle neck pain. A 23-month-old boy visited the neurologic clinic after 3 months of right occipital area neck pain. He had no history of trauma or central nervous system infection, and neurologic examination results were normal except for right posterior neck hyperesthesia. Brain and spinal magnetic resonance imaging showed an ovoid intramedullary cystic lesion (9.7×5.0×4.7 mm) at C6/7 of the spinal cord. There was no evidence of Chiari malformation or other lesions that can be primary pathologies of syringomyelia. Electromyogram/nerve conduction velocity results were normal. The subject was diagnosed as idiopathic syringomyelia. His symptoms and neurologic/radiologic indications showed no change at a 1-year follow-up. Idiopathic syringomyelia symptoms are varied and may be overlooked by physicians. Pediatricians may consider syringomyelia if patients complain about persistent sensory abnormality. All patients who present with syringomyelia should undergo detailed neuroimaging of the entire neuraxis to elucidate the proximate cause of the lesion.

Key Words: Syringomyelia

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Introduction

Syringomyelia is a disorder in which a cavity has formed within the spinal cord. In most cases, syringomyelias are associated with one of several treatable conditions including type 1 Chiari malformation¹⁾, spinal cord tethering, spinal cord tumor, vascular malformation and acute transverse myelitis¹⁻⁴⁾. Idiopathic syringomyelia can be diagnosed when there are no underlying pathologies detected.

With the technical development and increased accessibility of magnetic resonance imaging (MRI), idiopathic syringomyelias have become more easily

detected. The presenting symptoms of syringomyelia include a wide range of variability from centromedullary syndromes such as pain and temperature instability to no clinical symptoms, and syringomyelia may be only detected incidentally³⁾.

There is little published guidance on how to decide when radiologic examinations should be performed in children who present with minimal symptoms. Herein, we report a case of a very young boy with idiopathic syringomyelia who presented with mild and subtle sensory symptoms.

Case report

A 23-month-old boy was brought to the Department of Pediatrics in the Seoul National University Boramae Medical Center after complaining of pain in his right occipital area for 3 months. Previously, he was healthy and developmentally normal with no history of trauma or infection. Three months before the pediatric clinic visit, he started complaining intermittently of pain in his right occipital and neck areas when those areas were touched and when turning his head to the right side.

On physical and neurologic examination, he showed normal responses except for right posterior neck hyperesthesia (Fig. 1). He exhibited neither motor weakness nor incontinence. Brain MRI and cervical-thoracic-lumbar spine MRI revealed an ovoid intramedullary cystic lesion (9.7×5.0×4.7 mm) at C6/7 of the spinal cord (Fig. 2). There was no evidence of a Chiari malformation or other lesions that can be primary pathologies of syringomyelia. Moreover, electromyogram/nerve conduction velocity findings were normal.

He was diagnosed as idiopathic syringomyelia, and the Depart-

ment of Pediatric Neurosurgery was consulted. He was followed up without intervention at the hospital outpatient clinic and no symptom progression was detected at a 1-year follow-up.

Discussion

Idiopathic syringomyelia is defined as the presence of a syrinx for which no defined etiology is evident and is reported mostly in adults that exhibit some evidence of degenerative changes apart from a syrinx¹⁾. Most syringomyelias in children are associated with Chiari malformation type 1.

Fig. 1. Sensory dermatome maps and site of sensory abnormality in this patient (black-colored circle shape) (LEE's Medical Dictionary, 2012).

Fig. 2. Initial pediatric C-T-L-spine MRI (contrast). (A) T2 TSE sagittal image revealing a cervical syrinx (ovoid intramedullary cystic lesion at C6/7 of the spinal cord, size 9.7×5.0×4.7 mm). (B) T2 TSE TRA coronal view.

The pathophysiology of idiopathic syringomyelia is unclear, but Bogdanov et al⁵⁾, suggested that a posterior fossa with diminished compliance promotes the development of pulsatile cerebrospinal fluid (CSF) subarachnoid pressure waves and is implicated in the development of syringomyelia. Struck et al⁶⁾, also reported abnormal CSF flow velocities in idiopathic syringomyelia and suggested the presence of flow pattern similarities at the foramen magnum in both Chiari malformation type 1 and idiopathic syringomyelia. In addition, there is a report suggesting that subtle microtrauma may be a factor contributing to idiopathic syringomyelia⁷⁾. Neurological symptoms of syringomyelia arise from spinal cord compression due to the cavity in the spinal cord being a space-occupying lesion³⁾. As in other central cord syndromes, pain and insensitivity to temperature with a cape-like distribution are classic syringomyelia symptoms. But syringomyelia can present with a variety of symptoms other than its typical symptoms¹⁾. Pain is the most common syringomyelia symptom, but paresthesia, numbness, unnoticed hand injuries, long tract signs, spastic weakness of lower extremities, dysesthesia can also be neurologic symptoms of syringomyelia¹⁾. In the present case, hyperesthesia in the right posterior neck area was the main syringomyelia symptom.

Many symptoms and signs of syringomyelia can be very subtle, as was observed in our case, and thus can be easily overlooked by physicians³⁾. Regardless, a delayed diagnosis should be avoided because syringomyelia can result in irreversible neurological damage; therefore, early detection and making the correct decision regarding the optimal timing of decompression of the syrinx may prevent progression to other neurologic problems³⁾. Moreover, it is important to detect a syringomyelia early because usually syringomyelias are associated with other treatable conditions such as a Chiari malformation, cord tethering, spinal cord tumor, and vascular abnormalities, thus prompt correction of underlying conditions can be the cure of a syringomyelia. Hence, extensive neurologic investigations are recommended.

The natural course of idiopathic syringomyelia is diverse, and idiopathic syringomyelia can spontaneously resolve itself⁶⁾. Ninety-one percent of children with idiopathic syringomyelias remain asymptomatic or have stable or improved symptoms²⁾. Thus, the treatment strategy for idiopathic syringomyelia is controversial. In many cases, the size of the syringomyelia is not related with the severity of the presenting symptoms or the neurologic outcome⁹⁾, therefore, symptom characteristics may be key factors when making a decision on treatment strategy.

For treatment options, patients without neurologic signs or symptoms and in whom syringomyelia was detected incidentally may be treated conservatively. But patients that show worsening

of symptoms may be treated more aggressively with surgical decompression, comprising myelotomy, a syringosubarachnoid or syringopleural shunt, and/or spinal cord transection^{10,11)}. Shunting operations have been used as a treatment for idiopathic syringomyelia. Regardless, to be cautious, it should be confirmed that there is no evidence of other syringomyelia-related pathologies as surgery can induce several complications such as shunt failure, syrinx relapse, and catheter blockage^{11,12)}.

In conclusion, chronic mild subtle neck pain can be a presenting symptom of syringomyelia in children without overt trauma history. It is recommended that all patients with syringomyelia undergo further extensive neuroimaging investigations to look for primary causes of the syringomyelia. Serial neurological and MRI follow-ups are needed to monitor the progression of neurological damage and to prevent permanent neurologic deficits.

요약

원발성 척수공동증은 드문 질환으로 대부분 키아리증후군과 관련이 있으며, 다른 이상과도 관련 있는 경우가 많으나 때로는 원인 없이 척수공동이 나타나게 되며 이를 원발성 척수공동증이라 부른다. 특징적인 증상은 구획화 되어있는 상지의 감각이상으로 기술되고 있으나, 통증 및 감각 이상과 같은 증상이 주된 첫 증상이다. 이 증례의 환자도 감각 이상을 느껴 자기공명검사를 시행한 결과 척수공동이 발견되었다. 원발성 척수공동증은 치료에 있어서도 명확히 정해진 바는 없으나, 증상을 기준으로 치료 방법을 나누는 것이 일반적이다. 증상이 없거나 매우 작은 경우 치료하지 않고 경과를 지켜보는 방법을 주로 택한다. 이 증례와 같이 지속된 감각 이상을 호소하는 경우 척수공동과 같은 질병을 의심하여 영상의학적 검사를 고려해보는 것이 필요할 것으로 생각된다.

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