Case Report

Acute Flaccid Monoplegia After an Asthmatic Attack

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Abstract

Background: Hopkins syndrome is a polio-like paralytic illness following a few days after asthmatic attack. Although very rare, the disease has devastating clinical outcomes.

Case Report: The patient was a 4-year-old girl presenting with acute monoplegia of her right arm six days after an event of acute asthmatic attack. Magnetic resonance imaging revealed increased T2 signal intensity within the cord from C1 to C6-7 levels, mainly at anterior horns. The patient did not gain motor power improvement after two years of treatment and follow up.

Conclusions: We report a rare case of Hopkins syndrome presented with acute flaccid monoplegia and magnetic resonance imaging pathology. This is the first case report in Southeast Asia.

Keywords: Hopkins syndrome, Monoplegia, Acute asthmatic attack, Asthmatic amyotrophy

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Introduction

Hopkins syndrome (HS) is a rare polio-like acute flaccid paralysis. It occurs a few days to few weeks following an asthma attack in children.[1,2] Hopkins, I. J. first reported this disease entity in 1974 and since then, less than 40 cases have been reported worldwide.[3,4] Immunologic or infectious pathologic process of the anterior horn cells was proposed as a cause of the disease, but these theories need further elucidation.[5,6]

Here, we are reporting a case of a 4-year-old girl presenting with acute flaccid monoplegia of the right arm. The diagnosis of HS was confirmed by prior history of acute asthmatic attack and magnetic resonance imaging (MRI) of the spinal cord. This is the first case report of HS in southeast Asia.

Case Presentation

The patient was a 4-year-old Thai girl presented with acute weakness of the right arm. The weakness occurred while she was waking up from sleep. She had no prior history of fever or trauma of the affected limb. Six days prior to the onset of weakness, she had been hospitalized due to third episode of moderate to severe asthmatic attack with the lowest oxygen saturation of 88%. She was treated with a course of inhaled beta-2 agonist, intravenous corticosteroid, and intravenous magnesium sulfate. Respiratory symptoms subsided without the need of intubation.

Upon arrival, physical examination revealed flaccid tone of the right arm. Motor power was graded 0/V by medical research council scale. Sensation was intact in pinprick sensation, and light touch. Deep tendon reflex was absent at the affected limb. Neurological examination other than the right arm was within normal limits.

Laboratory investigation revealed high erythrocyte sedimentation rate 43 mm/hour (0-20 mm/hour), high C-reactive protein 18.01 mg/L (0-0.75 mg/L). Serum antibodies for herpes simplex virus, Epstein-barr virus, and varicella zoster virus were negative. Serum antinuclear antibody, anti-dsDNA, and anti-aquaporin-4 antibodies were negative. Anti-aquaporin-4 antibodies in cerebrospinal fluid was negative. Complement 3 and 4 level were normal for age. Serum Mycoplasma IgM and IgG were positive at onset of weakness with a titer of 1:200 and 1:320, respectively. The titers turned negative on the subsequent testing after 2 weeks, rendering the diagnosis of mycoplasma less likely.

Cerebrospinal fluid analysis revealed pleocytosis with white blood cell 39 cell/mm^3 all mononuclear cells. Red blood cell 100 cell/mm^3 protein 30.9 mg/dL, CSF/serum glucose ratio is 0.57. Cerebrospinal fluid polymerase chain reaction (PCR) for enterovirus, coxsackie virus, herpes simplex virus, adenovirus and cerebrospinal fluid bacterial culture were negative.

Magnetic resonance imaging was performed on the fifth day after the onset and revealed swelling of cervical cord with increased T2 signal intensity within the cord from C1 to C7 levels, mainly at anterior horns and right side of the white matter of the cervical cord (Figures 1 and 2).

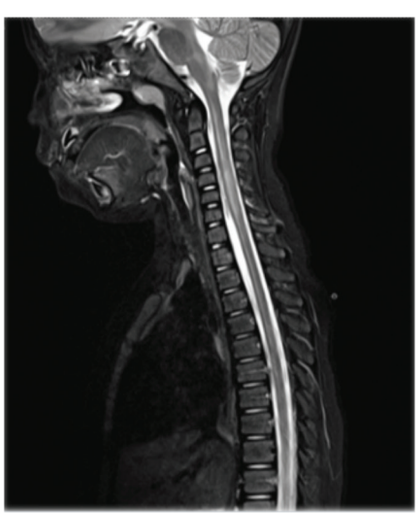

Figure 1 MRI sagittal T2 weighted view of the cervical segment of spinal cord revealed swelling of cervical cord with increased T2 signal intensity within the cord from C1 to C6-7 levels.

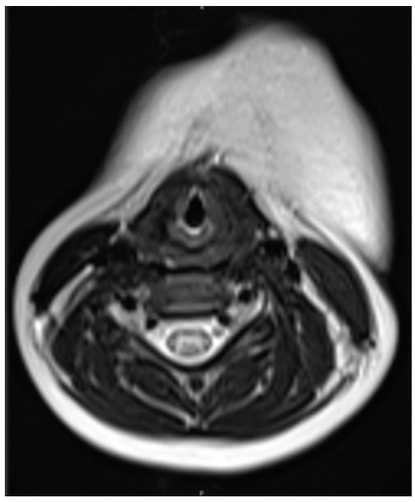

Figure 2 MRI axial T2 weighted view of cervical spinal cord revealed anterior horns and right side of the white matter of the cervical cord were affected.

With the suspicion of Hopkins syndrome, pulse methylprednisolone 30 mg per kg/day was given intravenously for 5 days, followed by oral prednisolone tapering for one month. An early extensive rehabilitation program was initiated and continued regularly. Unfortunately, after two years of follow up, the patient did not have significant motor power improvement. The deep tendon reflex at the affected limb was still absent.

Discussion

Hopkins syndrome, or asthmatic amyotrophy, is a paralytic illness following an asthmatic attack. Like poliomyelitis, the syndrome commonly attacks anterior horn cells, causing flaccid paralysis of the limb, sparing sensory and autonomic modalities. Monoplegia is the most common type of weakness, followed by diplegia, and hemiplegia.[7] Although firstly and more commonly described in patients under 13 years old, there was also a study reporting HS in up to elderly.[2,8]

Pathoetiology of the disease is still unclear. Both immunologic process and viral direct invasion to the spinal cord has been proposed.[5,6] No study, including in our patient, has shown an evidence of poliovirus infection.[1,2,4] Hayashi et al. reported detection of enterovirus D68 in endotracheal aspirates during the attack but fail to detect it in cerebrospinal fluid.[3] The role of immune-mediated mechanism was also suggested due to an elevation of cerebrospinal fluid oligoclonal bands in a study.[9] Manson et al. proposed that immunosuppression from asthma treatment rendered patients more susceptible for viral invasion and attack the anterior horn cells.[5]

Spinal cord MRI is critical for diagnosis of Hopkins syndrome. Anterior horn cells are the most affected part.[4,7] In our patient, and in several previous studies, the MRI showed bilateral unequal involvement.[1-3] The area of involvement may extend to posterior horn cells in more severe cases.[3] But the spinal MRI can also be normal.[10]

There is no treatment of choice in Hopkins syndrome. Courses of corticosteroid or intravenous immunoglobulin failed to significantly improve the weakness at 1-2 years after onset.[2,6-8] However, one study reported treating a patient with intravenous immunoglobulin and resulted in complete weakness recovery.[6]

Although this syndrome is very rare, it is important for clinicians to suspect the diagnosis of this disease when acute flaccid paralysis follows an asthmatic attack. A newer strategy is still needed to improve weakness outcome of the patients suffering from HS.

Ethical Considerations

This case report article was approved by the ethic committee of the Faculty of Medicine, Thammasat University, Pathum Thani, Thailand. Written informed consent was obtained from the guardian of the patient.

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None reported.

Potential conflicts of interest

All authors report no conflicts of interest relevant to this article.

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