Emergency. 2018; 6 (1): e23

OPEN ACCESS

CASE REPORT

Hopkins Syndrome in a 14 Year Old Boy; a Case Report

Alireza Majidi[1], Sasan Saket[2], Zohreh Nabizadeh Gharghozar[3], Ehsan Akrami[2], Seyed Mohsen Fayyazi[1]*

1. Emergency Department, Shohadaye Tajrish Hospital, Shahaid Beheshti University of Medical Sciences, Tehran, Iran.
2. Pediatric Neurology Research Center, Shahaid Beheshti University of Medical Sciences, Tehran, Iran.
3. School of Nursing and Midwifery, Kashan University of Medical Sciences, Kashan, Iran.

Received: August 2017; Accepted: January 2018; Published online: 16 April 2018

Abstract: Hopkins syndrome (HS) is a flaccid paralysis resembling poliomyelitis that has been seen in some children who are recovering from an acute episode of asthma. This syndrome should be suspected based on clinical findings even before the occurrence of characteristic breathing patterns and epilepsy. We report a 14-year-old boy who had experienced an episode of HS.

Keywords: Pitt-Hopkins syndrome; nervous system diseases; rare diseases; asthma

Cite this article as: Majidi A, Saket S, Nabizadeh Gharghozar Z, Akrami E, Fayyazi SM. Hopkins Syndrome in a 14 Year Old Boy; a Case Report. Emergency. 2018; 6 (1): e23.

1. Introduction

In some children recovering from an acute episode of asthma, a flaccid paralysis resembling poliomyelitis has been encountered (1-3). This condition, first reported from Australia in 1974, has been termed Hopkins syndrome (HS) (4). No consistent virus has been cultured from such patients, who have generally been successfully vaccinated against poliomyelitis (5). The disorder primarily affects the anterior horn cells. A rapid progression of paralysis usually affects one limb, leaving the child with a severe and permanent weakness. Sensation is preserved; the cerebrospinal fluid (CSF) usually shows moderate mononuclear pleocytosis, and the protein content can be slightly elevated (6-8). Magnetic resonance imaging (MRI) changes in the anterior horn have been documented (9). Some of the children have shown evidence of an underlying immune deficiency (10). We report a 14-year-old boy who had experienced an episode of HS.

2. Case Presentation:

A 14-year-old boy with a history of asthma since being 2 years old was admitted to hospital with history of seizure and loss of consciousness. There was a history of common cold in the patient a week before and he had experienced severe shortness of breath 4 days before admission. No significant history of trauma was recorded. His mother had a history of abortion. His mother had no certain disease during and before being pregnant with him. He had used atrovent and salbutamol sprays. No family history of similar illness was present. At the time of admission, vital signs were as follows: temperature 37.8 °c, pulse 131 beats per minute, respiratory rate 26 per minute and blood pressure 90/50 mmHg. On physical examination, he was confused and there was bilateral expiratory wheezes as well as scattered rhonchi in the lower lobes. His thoracic x-ray was normal. The neurologic examination results were normal. His complete blood count was as follows: 25,600 white cells/mm^3, with 81% neutrophils, 2% eosinophils, 15% lymphocytes, and 3.0% monocytes, Erythrocyte sedimentation rate (ESR) was (8, 0-10 mg/dl), C-reactive protein (CRP) was (4.1 mg/dl, 0-6 mg/dl), pH= 7.59, Po_2= 96.5, HCO_3= 25.9 mmol/l, and O_2sat=94%. Other routine laboratory test results were normal. The antiviral antibodies in paired serum samples showed no significant changes in any of the examined viruses, including herpes 1 and 2, echovirus, enterovirus, coxsackievirus, and poliovirus types 1, 2, and 3. TORCH, anti-mycoplasma, and anti-borrelia titers were normal. Hbs Ag, Abs Ab, HCV Ab and HIV Ab were negative.

The cerebrospinal fluid cultures were negative. No cardiac abnormalities were noted on clinical and ultrasound examinations. The sleeping EEG showed anomalies in the frontocentral regions with left prevalence and tendency toward

***Corresponding Author:** Seyed Mohsen Fayyazi; Emergency Department, Shohadaye Tajrish Hospital, Shahrdari Avenue, Tajrish Square, Tehran, Iran. Tel: 00989306224313 Email: mfayyazib@yahoo.com

contralateral diffusion. Abdominal ultrasound and computed tomography (CT) scans were normal. His brain MRI was normal. With suspition to Pitt-Hopkins Syndrome Acyclovir 400 mg was prescribed. He experienced no other asthma and seizure attack. During a 5-month follow-up from the episode there were no new clinical symptoms.

3. Discussion

Hopkins syndrome is a rare disease affecting anterior horn cells following an acute asthma attack, most often in children that have atopic asthma (11). Generally, this disease manifests from several days to a few weeks after an acute attack of asthma, presenting as the acute onset of flaccid paralysis, and in most cases progresses to severe muscle atrophy in the affected limb (12).

Based on previous studies, in such episodes, there is a poor response to corticosteroids and usually there are no recurrences (13). Some cases have predominant ventral root involvement on MRI, while other cases exhibit an extension of the lesions into the anterior columns (14-16). Therefore, the site of lesions in HS is not strictly limited to the anterior horn. In some of the previous studies it was reported that peripheral blood lymphocytes of patients with asthma express more cytokine on stimulation with allergens compared to those with skin allergy (17), but currently there is no evidence-based clinical correlation between cytokine production and Hopkins syndrome. Atopic myelitis (AM) is a clinical entity with similarity to Hopkins syndrome. Many reports have confirmed the presence of evidence in the central nervous system or peripheral nervous system in patients with a high level of total IgE and co-existing atopic disease (18). HS and AM differ from each other in the preferential age of onset, neurologic manifestations, and preferential sites of spinal cord involvement. However, both conditions are similar regarding the most important point that myelitis develops in the presence of atopic disorders, which suggests a link between atopy and the development of spinal cord inflammation. Both of them could be an allergic mechanism due to cross reactivity between an allergen and CNS or PNS. Kira et al. described 22 patients with myelitis of unknown etiology and atopic diathesis, 5 of whom showed focal amyotrophy in one or two limbs (16).

Hopkins syndrome is frequently associated with epilepsy. Previous reports do not include detailed EEG descriptions, but slow-wave activity was reported in the patients of Pitt and Hopkins (1978), whereas in Singh's (1993) patient, there was "generalized epileptogenic activity".

Horiuchi et al. described a 22-year-old woman who showed an additional episode of myelitis after another asthma attack, although no relapse has ever been reported in HS cases (4, 19, 20). Joubert Syndrome, Rett Syndrome and Angelman Syndrome should be considered as differential diagnoses in patients with the same symptoms as our patient (21).

3.1. Conclusion:

Hopkins syndrome should be suspected based on clinical findings even before the occurrence of typical breathing patterns and epilepsy.

4. Appendix

4.1. Acknowledgements

All the staff members of the emergency department of Shohaday-e-Tajrish Hospital are thanked for their cooperation throughout the study period.

4.2. Authors contribution

All authors meet the standard criteria of authorship based on the recommendations of the international committee of medical journal editors.

4.3. Conflict of interest

The authors declare that there is no conflict of interest in any phase of performing the study.

4.4. Funding

None.

References

1. Hopkins I. A NEW SYNDROME: POLIOMYELITISâĂŘ-LIKE ILLNESS ASSOCIATED WITH ACUTE ASTHMA IN CHILDHOOD. Journal of Paediatrics and Child Health. 1974;10(5):273-6.
2. Yeung SC, Antonio G, Ip KS. Flaccid paralysis of the limbs after an asthmatic attack. Pediatric neurology. 2010;42(2):133-6.
3. Leonardi M, Sartorius N, Hull HF, Ward NA, Organization WH. Acute onset flaccid paralysis. 1993.
4. Horiuchi I, Yamasaki K, Osoegawa M, Ohyagi Y, Okayama A, Kurokawa T, et al. Acute myelitis after asthma attacks with onset after puberty. Journal of Neurology, Neurosurgery & Psychiatry. 2000;68(5):665-8.
5. Fine PE, Carneiro IA. Transmissibility and persistence of oral polio vaccine viruses: implications for the global poliomyelitis eradication initiative. American journal of epidemiology. 1999;150(10):1001-21.
6. Tyler KL. Acute viral myelitis. Infections of the central nervous system. 2004:310-1.
7. Sejvar JJ, Leis AA, Stokic DS, Van Gerpen JA, Marfin AA, Webb R, et al. Acute flaccid paralysis and West Nile virus infection. Emerging infectious diseases. 2003;9(7):788.

8. Marx A, Glass JD, Sutter RW. Differential diagnosis of acute flaccid paralysis and its role in poliomyelitis surveillance. Epidemiologic reviews. 2000;22(2):298-316.
9. Chen C-Y, Chang Y-C, Huang C-C, Lui C-C, Lee K-W, Huang S-C. Acute flaccid paralysis in infants and young children with enterovirus 71 infection: MR imaging findings and clinical correlates. American journal of neuroradiology. 2001;22(1):200-5.
10. Shahmahmoodi S, Mamishi S, Aghamohammadi A, Aghazadeh N, Tabatabaie H, Gooya MM, et al. Vaccine-associated paralytic poliomyelitis in immunodeficient children, Iran, 1995âĂŞ2008. Emerging infectious diseases. 2010;16(7):1133.
11. Piero P, Longo MR, Scalia F, Polosa R, Kira J-i, Falsaperla R. Recurrent Hopkin's syndrome: A case report and review of the literature. Journal of the neurological sciences. 2010;297(1):89-91.
12. Shapiro GG, Chapman JT, Pierson WE, Bierman CW. Poliomyelitis-like illness after acute asthma. The Journal of pediatrics. 1979;94(5):767-8.
13. Shahar EM, Hwang PA, Niesen CE, Murphy EG. Poliomyelitis-like paralysis during recovery from acute bronchial asthma: possible etiology and risk factors. Pediatrics. 1991;88(2):276-9.
14. Danta G. Electrophysiological study of amyotrophy associated with acute asthma (asthmatic amyotrophy). Journal of Neurology, Neurosurgery & Psychiatry. 1975;38(10):1016-21.
15. Liedholm LJA, Eeg-Olofsson O, Ekenberg BEK, Nicolaysen RB, Torbergsen T. Acute postasthmatic amyotrophy (Hopkins' syndrome). Muscle & nerve. 1994;17(7):769-72.
16. Kira J-i, Isobe N, Kawano Y, Osoegawa M, Ohyagi Y, Mihara F, et al. Atopic myelitis with focal amyotrophy: a possible link to Hopkins syndrome. Journal of the neurological sciences. 2008;269(1):143-51.
17. Kimura M, Tsuruta S, Yoshida T. Differences in cytokine production by peripheral blood mononuclear cells (PBMC) between patients with atopic dermatitis and bronchial asthma. Clinical and experimental immunology. 1999;118(2):192.
18. Osoegawa M, Ochi H, Yamada T, Horiuchi I, Murai H, Furuya H, et al. High Incidence of Subclinical Peripheral Neuropathy in Myelitis with HyperlgEaemia and Mite Antigen-specific IgE (Atopic Myelitis): an Electrophysiological Study. Internal medicine. 2002;41(9):684-91.
19. Kira J-i, Kawano Y, Yamasaki K, Tobimatsu S. Acute myelitis with hyperIgEaemia and mite antigen specific IgE: atopic myelitis. Journal of Neurology, Neurosurgery & Psychiatry. 1998;64(5):676-9.
20. Kira J-i, Kawano Y, Horiuchi I, Yamada T, Imayama S, Furue M, et al. Clinical, immunological and MRI features of myelitis with atopic dermatitis (atopic myelitis). Journal of the neurological sciences. 1999;162(1):56-61.
21. Taddeucci G, Bonuccelli A, Mantellassi I, Orsini A, Tarantino E. Pitt-Hopkins syndrome: report of a case with a TCF4 gene mutation. Italian journal of pediatrics. 2010;36(1):12.