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Hopkins Syndrome and Phantom Hernia: A Rare Association

KE ELIZABETH, CS GURUPRASAD AND TG SINDHU

From Department of Pediatrics, SAT Hospital, Government Medical College Trivandrum.

Correspondence to:

Dr KE Elizabeth,
Professor of Pediatrics,
SAT Hospital, Government Medical
College, Trivandrum 695 011,
Kerala, India. elizake@hotmail.com
Received: November 09, 2009;
Initial review: January 1, 2010;
Accepted: March 29, 2010.

Acute flaccid paralysis (AFP), other than paralytic poliomyelitis, are usually due to demyelination like Guillian Barre syndrome (GBS), transverse myelitis and traumatic neuritis. Poliomyelitis like illness, Hopkin's syndrome or Post Asthmatic Amotrophy, associated with bronchial asthma and hyperIgEemia has been reported in literature. We present a two and a half year old child who developed AFP with phantom hernia following an episode of bronchial asthma.

Key words: *Acute Flaccid Paralysis, Phantom hernia, Hopkin's syndrome, HyperIgEemia.*

Subsequent to decline in cases of paralytic poliomyelitis, demyelination is the most common cause of AFP followed by traumatic neuritis. AFP has also been reported with asthmatic attack and hyperIgEemia (Hopkin's syndrome or Post asthmatic amotrophy) and following ICU admission for critical illness, called 'critical illness polyneuropathy/myopathy [1-3]. Phantom hernia refers to unilateral bulging of abdominal wall due to patchy paralysis of abdominal wall muscles. It was first described in paralytic poliomyelitis by Achar [4]. It is derived from the word 'phatasm' meaning mental imagery produced by fantasy [5]. It has also been reported with non-polio conditions like hypokalemia complicating gastroenteritis [6].

We report a case of AFP with phantom hernia associated with an asthmatic attack and hyper-IgEemia. Several cases of poliomyelitis like illness following bronchial asthma have been reported [7-10], but associated phantom hernia has not been documented.

CASE REPORT

A two-and-a-half year old male child with normal

growth and development was admitted in PICU with acute severe asthma. He had intermittent asthma for the past one and a half years. He was fully immunized including pulse polio immunizations. He had three days of ICU stay and was given nebulization with salbutamol, ipratropium and steroids, ampicillin, IV methyl prednisolone and MgSO₄ infusion. He responded and did not require any ventilator support or muscle relaxants.

On the 4th day of hospital stay, he developed acute flaccid paralysis of the left lower limb, which progressed and involved the right lower limb in the next two days. His higher mental function and upper limbs were normal. He had hypotonia, grade 0 power, areflexia and down going plantars in both lower limbs. He had no sensory involvement except for myalgia. Abdominal reflex on the left lower quadrant was absent. There was transient bladder involvement also. Blood counts were within normal limits and there was no eosinophilia. CSF study one week after paralysis showed 0-2 lymphocytes/mm³ and protein of 20 mg/dL and sugar of 80 mg/dL. Mantoux test was negative. Chest X-ray showed increased bronchovascular markings. Serum CPK and LDH were normal. Mycoplasma antibody was

negative. There was ten fold rise in serum IgE level, 630 IU/dl (normal <60 IU/dl). MRI spine and brain were normal. Nerve conduction study showed decreased amplitude of complex motor neuron action potential with greater involvement of the left lower limb compared to right. This asymmetrical pattern and absence of albumin-cytological dissociation in the second week were inconsistent with GBS. Eventhough fibrillations were not seen on surface EMG, anterior horn cell involvement could not be ruled out .The child was non-cooperative for a needle EMG study.

Para infectious demyelination was considered and IV Immunoglobulin (IVIG) 2g/kg was given for two days, but there was no improvement. AFP reporting was done and child was initiated on physiotherapy. He was discharged on oral steroids. At the time of discharge, he had severe hypotonia, areflexia and grade 0 power of both the lower limbs.

During follow up visit 2 weeks later, he was found to have phantom hernia in the left lumbar region. His stool culture for poliovirus was negative. He is currently on follow up for the last 6 months with only very little recovery. Phantom hernia has resolved, but with only Grade 1 power in left lower limb and grade 2 power in right lower limb with muscle atrophy, areflexia, weakness and severe physical handicap.

DISCUSSION

In 1974, Hopkin described ten cases of poliomyelitis like illness following acute asthma [1]. Subsequently, similar cases were reported [2,3,7-10]. All these cases had hyperIgEemia in common. In these cases, no single organism was isolated, but microbes like adenovirus, ECHO, Coxsackie B 5 and mycoplasma were documented. All the reported children had received various drugs in the recommended limits and no adverse drug reactions were noted. The unusual combination of symptoms and striking similarity in these reported cases argue against a chance association between asthma and paralysis [7,8].

According to a hypothesis put forward by Manson and Thong, patients who develop poliomyelitis like illness after an attack of acute asthma,

do so because of an underlying immunological deficiency, albeit of a minor nature [7]. It is possible that due to further immunosuppression in these patients at the time of stress, intercurrent infections or corticosteroid therapy, the host resistance is reduced to such a level that a neurotropic virus, not usually considered pathogenic, may invade the anterior horn cells [7].

In view of the persistent clinical course of the child, with poor response to IVIG and steroids, presence of phantom hernia, and NCV and CSF findings not consistent with GBS; we thought of other differential diagnosis of AFP. Paralytic poliomyelitis was ruled out in this case as the child was fully immunized and stool culture for poliovirus was negative. Rare association of AFP with mycoplasma was also ruled out in this case [9]. Due to the association with bronchial asthma and hyper-IgEemia, Hopkin’s syndrome was considered. The physical and laboratory findings were comparable to the previously reported cases of Hopkin’s syndrome, except for phantom hernia [10] (**Table 1**).

Acknowledgment: Dr MK Mohandas, Professor of Pediatrics, Dr SM CSI Medical College, Karakonam, Trivandrum, Kerala for his suggestions and, Dr PA Mohammad Kunju and Dr D

TABLE I PREVIOUS CASES OF POLIOMYELITIS LIKE ILLNESS AFTER ACUTE ASTHMA

	Reported cases (n-22)
Gender - M:F	1.75:1
Age at onset	13 months-11 years
Immunized against poliovirus	Yes in all cases
Days between wheezing and paralysis	4 – 11
Meningism	Yes in 3/22
Myalgia	Yes in 10/22
Neurological findings (acute phase)	Paralysis of arm or leg
Reduced Motor NCV	Yes in 3/14
Denervation on EMG	Yes in 13/13
Residual weakness with muscle atrophy	Yes in all cases
Microbial association	Adeno/ ECHO/ CoxsackieB5/ <i>Mycoplasma</i>

NCV: nerve conduction velocity; EMG: electromyography.

Kalpana, Department of Pediatric Neurology, SAT Hospital Trivandrum for their help in the management of the case.

Contributors: EKE was the consultant in charge, critically evaluated the manuscript and will act as the guarantor of study. GCS managed the case, collected data and drafted the paper. STG helped in management and revision of the manuscript.

Funding: None.

Competing interest: None stated.

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Prolonged Cholestasis due to Hepatitis A Virus Infection

PAWAN KUMAR AND VIDYUT BHATIA

From the Department of Pediatrics, All India Institute of Medical Sciences, New Delhi 110 029, India.

Correspondence to:

Dr Vidyut Bhatia,

Senior Research Associate,

Department of Pediatrics,

AIIMS, New Delhi 110 029, India.

drvidyut@gmail.com

Received: January 25, 2010;

Initial review: February 26, 2010;

Accepted: March 29, 2010.

We present a 12-year old boy with jaundice for 2 weeks. The child was deeply icteric and had hepatomegaly. IgM antibodies for hepatitis A virus were positive. However this child had prolonged cholestasis and cholestyramine was started. The child responded only after prednisolone was started.

Key words: *Hepatitis A, Clinical features, Cholestasis.*

Acute viral hepatitis due to Hepatitis A virus is usually a self limiting illness in children with complete recovery within two months of onset of symptoms [1]. Occasionally, the clinical syndrome of cholestasis, characterized by intense pruritus and prolonged conjugated hyperbilirubinemia, may persist for several months [2].

CASE REPORT

A 12-year old male child was admitted with

complaints of jaundice for 6 weeks and itching for 2 weeks. The onset of jaundice was preceded by low grade fever and nausea for 4-5 days. There was no history of bleeding from any site or any symptoms of encephalopathy. There was no past history of jaundice. On examination, the child was deeply icteric. Scratch marks were present all over the body and there was no pallor. His weight was 39 kg and height was 142 cm. The vital parameters were stable, and examination of cardiovascular, respiratory and neurological systems was unremarkable. Liver was