MEDDOCS Open Access Publisher

ISSN: 2637-9627

Annals of Pediatrics

Open Access | Case Report

Stridor as the Initial Presentation of Guillian Barré Syndrome (GBS): Two Paediatric Cases

Banks T[1]; Bernardo Fonz R[2]; Majumdar A[1]

[1]University Hospitals Bristol, Bristol, UK.

[2]University Hospital Migel Severet, Zaragoza, Spain.

***Corresponding Author(s): Teresa Banks**

Department of Paediatric Neurology, Bristol Royal Hospital for Children, University Hospitals Bristol, Bristol, UK.

Mail: teresa.banks@uhbw.nhs.uk

Received: Nov 23, 2020

Accepted: Dec 21, 2020

Published Online: Dec 24, 2020

Journal: Annals of Pediatrics

Publisher: MedDocs Publishers LLC

Online edition: http://meddocsonline.org/

Abstract

Guillain-Barre Syndrome (GBS) is a rare diagnosis in the paediatric population and usually presents with classic post-viral ascending paralysis and Areflexia. Stridor is a common presentation to acute paediatric services and is usually caused by laryngotracheobronchitis (viral croup).

We present two case studies of paediatric patients who presented acutely with respiratory distress and stridor. The focus on acute airway compromise and treatment for croup in both cases may have detracted from more subtle neurological signs and thus causing a delay in investigation and treatment for their final diagnosis of GBS.

These cases serve as an important reminder for those caring for paediatric patients with stridor. If the symptoms seem especially refractory to conventional croup treatments, then further assessment and investigation for underlying neurological issues is warranted.

Introduction

Background

Guillian-Barré Syndrome (GBS) is a clinical syndrome characterised by post-viral ascending, bilateral weakness with hypo or Areflexia. Although clinical resolution is common in the paediatric population, it is important to recognise and treat the condition promptly to prevent acute deterioration and longstanding consequences occurring.

We report two cases of children that presented to the paediatric neurology team, initially with stridor and with a final diagnosis of Guillian-Barré Syndrome. These cases highlight several pitfalls that can occur when examining and treating a paediatric patient. They serve as a reminder to keep an open mind when faced with a child who appears to have a common illness but is refractory to conventional treatment.

Case report 1

A 22 month-old boy presented to his local hospital with acute onset stridor. There was no history of febrile illness or foreign body and the child was generally well otherwise. He had a history of multiple viral illnesses in the previous months and was unwell 2 weeks prior to this admission with fever, cough and loose stools.

Initially he was treated for croup with minimal effect on the symptoms. He was transferred to the local children's hospital for assessment under the paediatric ENT team. Microlaryngoscopy and Bronchoscopy (MLB) showed bilateral vocal cord palsy with no localised oedema or foreign body.

During the admission he become more miserable and after referral to Paediatric Neurology was found to have an ascending bilateral flaccid paralysis with Areflexia. On questioning, his par-

Cite this article: Banks T, Bernardo FR, Majumdar A. Stridor as the Initial Presentation of Guillian-Barré Syndrome (GBS): Two Paediatric Cases. Ann Pediatr. 2020; 3(1): 1047.

ents explained he had been reluctant to walk for the previous 2 days. He also had intermittent sinus tachycardia, sweating with heat rashes and ongoing swallowing difficulties.

Investigation at this point showed markedly raised CSF protein with normal cell counts, a normal MRI brain and spine, and nerve conduction studies showed a severe demyelinating motor and sensory polyneuropathy.

He received 5 days of Intravenous Immunoglobulin (IVIG) treatment for a diagnosis of GBS. He required nasogastric feeding for an unsafe swallow and gabapentin for presumed neuropathic pain. His stridor settled on day 4 of IVIG treatment and his episodes of sinus tachycardia with autonomic symptoms settled after a week. After a 3 week inpatient stay, he was discharged home self feeding, with mild lower limb weakness which was improving. He went on to make a full neurological recovery.

Case report 2

A 19 month-old boy presented to Paediatric ED with a choking episode. He had respiratory and swallowing difficulties with low oxygen saturation. He had suffered from varicella zoster infection the week prior to this presentation. There was no other relevant past history or family history.

A rigid bronchoscopy was performed and found a small piece of mashed banana in the right main bronchus. This was removed successfully and the child recovered fully and was discharged home. A day later he developed further respiratory difficulties with stridor that required tracheal intubation. A second rigid bronchoscopy at this time showed some localised, mild vocal cord oedema with bilateral vocal cord paralysis.

On day 2 of PICU admission, an attempt at extubation failed as he was found to be significantly hypotonic with poor respiratory effort. Further assessment showed absence of cough and gag reflexes, ophthalmoplegia and Areflexia. A diagnosis of Miller-Fisher syndrome was made and he received 5 days of IVIG treatment.

CSF cell count and protein levels were normal (performed day 3 of illness). All serological tests for infections in blood, CSF and respiratory aspirates were negative. Brain and spine MRI was reported as normal. Following this treatment, his symptoms began to recover and at this point extubation was possible. Lower limb weakness recovered rapidly, but the facial paralysis with bilateral ptosis and weakness in upper limbs persisted.

Two weeks into the illness he started to swallow and say some words. He was able to walk at this time although his gait was unsteady. The facial and upper limb weakness gradually improved and after three months, the patient had made a complete neurological recovery.

Outcomes

Both patients were reviewed in clinic 6 months after discharge. Each had made a full neurological recovery with no findings on examination. Interestingly, both sets of parents describe a level of tiredness and fatiguability in the patients after discharge which was still present at follow up.

Discussion

Stridor is an abnormal, high-pitched sound produced by turbulent airflow through a partially obstructed upper airway. In the paediatric population, it is usually caused by a respiratory infection such as croup, retropharyngeal abscess or epiglottitis. However, stridor can be caused by many other conditions. Neurological disorders with involvement of the recurrent laryngeal nerve leading to partial or complete vocal cord paralysis is an unusual cause for stridor in this population.

Guillian-Barré Syndrome (GBS) is a monophasic, immune mediated polyradiculoneuropathy. It is characterised by rapidly progressing ascending weakness, with hypo or Areflexia and mild sensory loss. The pathophysiology of damage is not fully understood but is thought to be an autoimmune process damaging myelin associated proteins or glycoproteins [1]. Patients can present with varied symptomology which is related to the location and type of nerves that are damaged and the extent of demyelination with or without indirect axonal damage. (See Table 1 for diagnostic criteria) In children it has a worldwide incidence of 0.34-1.34 per 100,000 [2].

Table 1: Diagnostic criteria for GBS [1].

Required	Supportive	Exclusionary
Progressive symmetric weakness of > 1 limb	Sensory symptoms or signs	Other causes excluded (botulism, toxins, diphtheria, porphyria)
Hyporeflexia or areflexia	Cranial nerve involvement especially bilateral VII	
Progression < 4 weeks	Autonomic dysfunction	
Symmetric weakness	CSF protein elevation	
	CSF cell count < 10/mm3	
	Electrophysiological features of demyelination	
	Recovery	

GBS is often grouped into variants depending of the precise symptomology. But many case reports have shown that there can be a significant overlap between these variants.

GBS and common variants [1,2,3]

- Acute Inflammatory Demyelinating Polyneuropathy (AIDP) is the classic bilateral ascending paralysis with areflexia and can have bulbar and autonomic involvement.
- Miller-Fisher syndrome usually comprises of ophthalmoplegia, weakness, Areflexia and ataxia.
- Pharyngeal-Cervical-Brachial variant (PCB) comprises of ptosis, facial palsy and neck, arm and pharyngeal muscle involvement [4].
- Bickerstaff's brainstem encephalitis involves altered conscious level, paradoxical hyperreflexia and ophthalmoplegia [5].

CSF albuminocytological dissociation is common to all GBS variants but may not be evident, especially early on in the illness (as seen in the second case). In both these described cases, the patient initially presented with stridor, breathing difficulties, and intermittent swallowing difficulties with choking.

The first case, although having a rare presenting symptom, seems to then follow the typical GBS pattern and recovery when given appropriate treatment and supportive management.

The second patient demonstrated upper and lower limb weakness with absence of cough and gag reflexes which, along with ophthalmoplegia, led to the diagnosis of the Miller-Fisher variant. In this case the limb weakness resolved quickly leaving the patient with a more pharyngeal-cervical-brachial variant clinical picture. This could represent part of what is thought to be an overlap between Miller Fisher and PCB [4].

Both cases, after appropriate treatment with IVIG responded well and clinical improvement was appropriate. Both patients made a full neurological recovery.

Learning points from these cases

- GBS is a rare in the paediatric population
- Stridor can be the initial presenting complaint of GBS and is caused by damage to the nerves supplying the vocal cords
- The urgency of acute airway issues in a paediatric patient can lead to the overlooking more subtle clinical findings which may point towards a different diagnosis
- It is important to consider alternative diagnoses for croup if it appears refractory to traditional treatment.

References

1. Dimachkie MM, Barohn RJ. Guillain-Barré syndrome and variants. Neurol Clin. 2013; 31: 491-510.
2. McGrogan A, Madle GC, Seaman HE, de Vries CS. The epidemiology of Guillain-Barré syndrome worldwide. A systematic literature review. Neuroepidemiology. 2009;32(2):150-63. Epub. 2008.
3. Lin J-J, Hsia S-H, Wang H-S, Lyu R-K, Chou M-L, Hung P-C, et al. Clinical variants of Guillain-Barré syndrome in children. Pediatr Neurol [Internet]. 2012; 47: 91–96.
4. Wakerley BR, Yuki N. Pharyngeal-cervical-brachial variant of Guillain-Barré syndrome. J Neurol Neurosurg Psychiatry. 2014; 85: 339–344.
5. Michev A, Musso P, Foiadelli T, Trabatti C, Lozza A, Franciotta D, et al. Bickerstaff Brainstem Encephalitis and overlapping Guillain-Barré syndrome in children: Report of two cases and review of the literature. Eur J Paediatr Neurol [Internet]. 2019; 23: 43–52.