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

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

Table 1: Diagnostic criteria for GBS [1].
GBS is often grouped into variants depending on 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**