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Bilateral vocal cord palsy causing stridor as the only symptom of syringomyelia and Chiari I malformation, a case report

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ABSTRACT

INTRODUCTION: Bilateral vocal cord palsy is a condition which has many causes (Gupta et al., 2012) [1]. Syringomyelia is an uncommon condition which describes the formation of fluid filled cavity, occupying the spinal cord (Chang, 2003) [2]. It rarely manifests itself as subacute onset of stridor.
PRESENTATION OF CASE: We present the case of a three year old female who presented for evaluation of her speech and language delay, when incidentally it was made note of her loud breathing which had previously been managed as bronchiolitis by her general practitioner. In hospital she was found to have a bilateral vocal cord palsy. Further investigation revealed a large syrinx as well as an associated Arnold Chiari 1 malformation, for which she required neurosurgical decompression.
CONCLUSION: Although uncommon, formation of a syrinx should be considered for patients who present with stridor and reiterates the importance of MRI as an important investigative tool of bilateral vocal cord palsy.

1. Introduction

Stridor is a common presentation in children defined as high pitched monophonic sound associated with breathing. Multiple underlying pathologies have been attributed with causing stridor [3]. Vocal cord palsy is the second most common congenital laryngeal abnormality, accounting for approximately 6%–23% of cases and can be further sub classified, anatomically, into unilateral or bilateral paralysis [4]. Syringomyelia is the term given to define a fluid filled cavity laying within the spinal cord, most commonly occupying space between the cervicothoracic vertebral levels [2]. It is commonly associated with a Chiari I malformation and may be an incidental finding [5]. Although the association between vocal cord palsy and syrinx with Arnold chiari has been described in the literature, it is rarely the only sign. We present a case where an incidental note of stridor lead to the diagnosis of syringomyelia with Chiari I malformation.

2. Presentation of case

A three year old female presented to hospital for further investigation into her speech and language delay. Incidental note was made of her high pitched breathing, which began suddenly two months prior to presentation and had been treated as a viral upper respiratory tract infection leading to the exacerbation of asthma. Family members noted that despite her cough and rhinorrhea subsiding and otherwise being well, her stridor had progressed and become louder and consistently audible. She had been treated with multiple courses of antibiotics as well as salbutamol and various steroid inhalers with minimal relief. She had normal voice, snoring but no apneas and had not had any cyanotic episodes or any evidence of respiratory distress. There was no significant obstetric, family or social history. The rest of her otolaryngological and neurological examination was non remarkable. A semi urgent microlaryngoscopy revealed a bilateral vocal cord palsy Her vocal cords were paramedical with no active abduction and paradoxical movement with respiration. Her cricoarytenoid joints were mobile. She was admitted to the paediatric intensive care unit for further investigation and ongoing investigative oximetry. A nasogastric tube was inserted for feeding as she was deemed a high risk patient for aspiration (Tables 1 and 2).

An MRI brain and spine was ordered to further investigate a cause of the bilateral vocal cord palsy and revealed a large syrinx formation extending cranially from the C2 vertebrae to the T10 vertebrae caudally. The cerebellar tonsils were low lying, projecting 8 mm below the foramen magnum, indicating a Chiari I malformation.

She was subsequently treated operatively with a posterior fossa decompression with improvement but not resolution of her stridor and discharged home with outpatient follow up.

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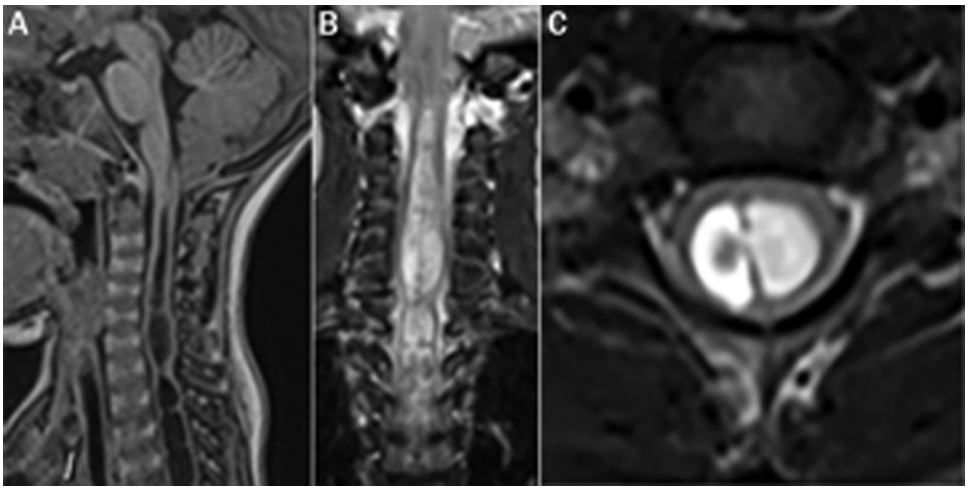

Fig 1. Chiari malformation and spinal cord syrinx. Sagittal T1 (A), coronal T2 (B) and axial T2 (C) images show mild descent of the cerebellar tonsils, to just below the foramen magnum, though associated with an extensive spinal cord syrinx—from obex to lower thoracic cord (lower limit not shown).

Table 1
Common causes of stridor in children.

Acute onset	Subacute/Chronic onset
Foreign body aspiration	Laryngomalacia
Epiglottitis	Vocal cord palsy—unilateral/bilateral
Burns	Subglottic stenosis
Laryngotracheitis	Vascular ring
Retropharyngeal abscess	Respiratory papilloma
Peritonsillar abscess	Tumor

Table 2
Age correlation with stridor.

Neonates and inants	Toddlers	Adolescents
Laryngomalacia	Foreign body aspiration	Supraglottitis
Subglottic stenosis + subglottic cysts	Croup	Paradoxical cord movement
Vocal cord palsy	Epiglottitis	Trauma
Tracheobronchomalacia	Respiratory papilloma	Bacterial tracheitis
	Tracheitis	

This patient was followed up in the outpatient clinic six weeks post-operatively with further improvement in her inspiratory stridor at rest but persistence during sleep (Figs. 1–4).

Due to the nocturnal stridor and the association of central apneas with Arnold Chiari malformations, a sleep stufy was performed. This reealed mixed sleep disordered breathing with borderline gas exchange, high work of breathing as well as sleep fragmentation. CPAP (continuous positive aitway pressure) was initiated and was well tolerated. Airway evaluation at three months showed no resolution of cord mobility but clinically she has no audible stridor at rest or with exertion.

3. Discussion

Stridor, originating from the latin term, '*stridere*', meaning to creak, is a common presentation in the paediatric population. However the multitude of underlying pathologies associated with stridor means that it may be difficult to distinguish a clear cause. Stridor is commonly distinguished by age, acuity and anatomically.

In the initial evaluation, emphasis must be placed on the acuity of the symptom in the history as well as pattern of stridor, i.e. whether it is an inspiratory or expiratory stridor. Acute onset of stridor is classically associated with an upper airway obstruction. This may be due to a foreign body aspiration, burn, epiglottitis,

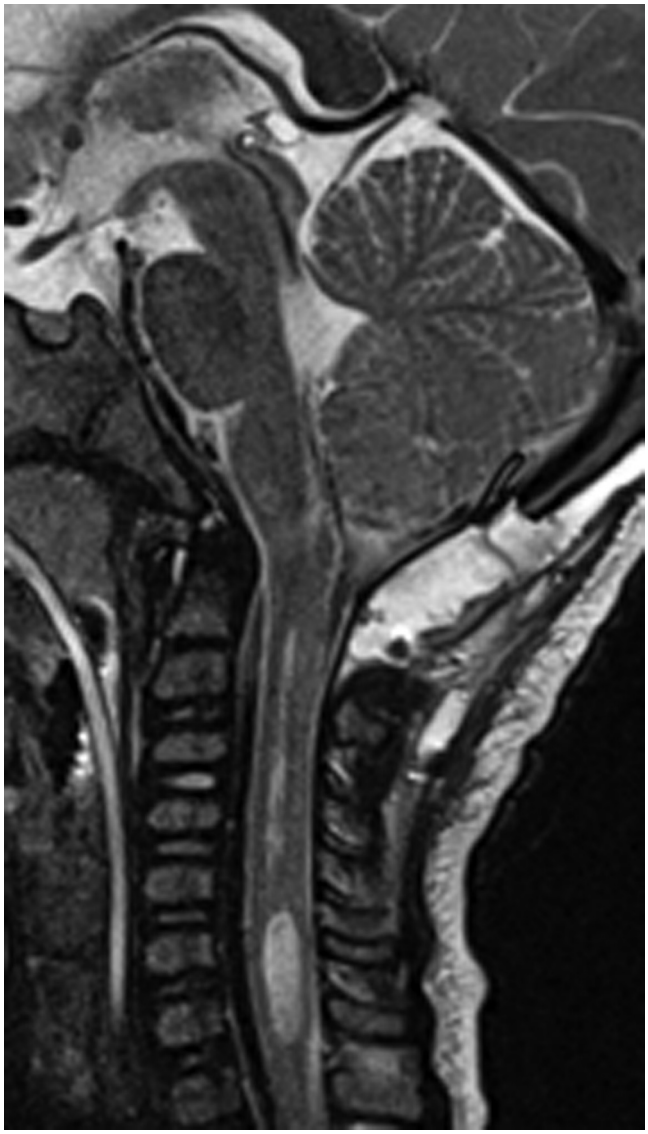

Fig. 2. Posterior fossa decompression. Sagittal T2 image shortly after a suboccipital craniotomy shows significant reduction of the cervical cord syrinx. Note the low signal 'CSF flow voids' at the craniocervical junction that also indicate improved CSF dynamics.

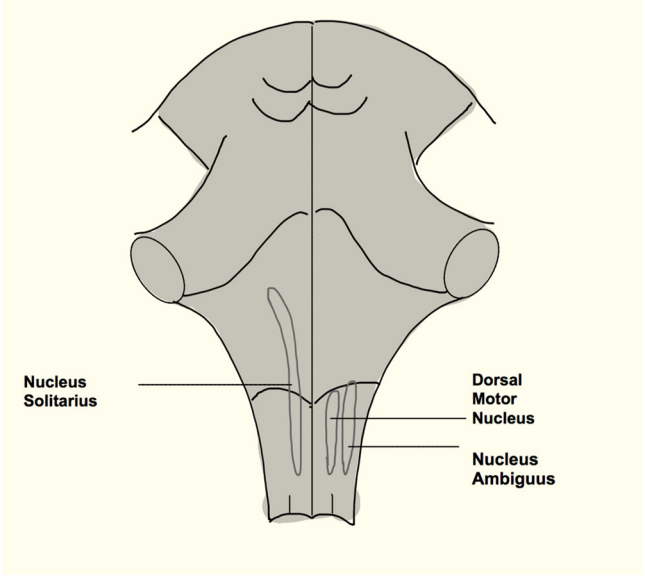

Fig. 3. The vagus cranial nerve nuclei. With the decent of the brainstem in Chiari I malformation, these nuclei can become compressed. As labelled, the nucleus solitarius gives sensory fibres for taste, nucleus ambiguous gives branchiomotor supply to the larynx and the dorsal motor nucleus gives general efferent visceral fibres. Further along the course of the vagus nerve, the recurrent laryngeal nerve branches off to innervate the larynx, a commonly injured nerve causing vocal cord paralysis.

laryngotracheitis or an abscess formation, being either retropharyngeal or peritonsillar abscess. This necessitates urgent attention and management by the clinician. It is the chronic stridor which is more likely in accordance with a structural abnormality. This may be persistent, intermittent or progressive in nature. Common causes include laryngomalacia, the most common cause of stridor in the neonatal population, as well as vocal cord palsy, subglottic stenosis subglottic cysts, vascular rings, respiratory papilloma and tumor causing compression of the airway. Chronic stridor can be misdiagnosed for long periods as asthma or bronchiolitis and hence mismanaged as in the case presented above.

Vocal cord palsy is one of the leading causes of stridor [6], the most common being laryngomalacia [7]. It is the second most common congenital anomaly of the larynx. It commonly manifests as an inspiratory stridor which may be progressive with time. It may or may not be associated with respiratory distress. Vocal cord paralysis, anatomically is defined as either being unilateral or bilateral. De Gaudamar et al. found similar rates of unilateral to bilateral vocal cord palsy. Out of the 113 patients evaluated with a vocal cord palsy, 61 had a unilateral palsy versus 52 who had a bilateral palsy. Unilateral cord paralysis can often be idiopathic although can also be attributed to mechanical injury to the left recurrent laryngeal nerve during cardiac and neck surgery as well as birth trauma.

Bilateral vocal cord palsy is also multi-causal. De Gaudamar also described that out of the 113 cases of vocal cord palsy, the aetiology varied, with 37% being idiopathic, 25% being secondary to a central nervous system dysfunction (most commonly an Arnold Chiari malformation), 13% due to obstetric complication and approximately 5% iatrogenic post cardiac surgery [8].

Bilateral vocal cord palsy is most commonly diagnosed in the neonate. Presentation at age three in unusual and bilateral cord palsy as the etiology for new onset stridor is an unexpected diagnosis. All children with bilateral vocal cord immobility, and those with unexplained unilateral lesions should undergo an MRI to exclude this and other central causes.

In addition to stridor, the child with vocal cord palsy may present with a weak/soft cry, recurrent aspiration pneumonia or respiratory distress. In the case presented above, it is common that a syringomyelia and/or Chiari I malformation in an otherwise well patient may not yet be diagnosed. Nohria et al. showed that the average age of diagnosis of a Chiari I malformation is 18 [9]. In addition to this, previous studies have estimated that up to 30% of cases of Chiari I malformation is diagnosed incidentally [10]. Hence a high degree of suspicion is required. 30% of cases of Chiari I malformation have an associated syrinx with the most common presentation being a headache, likely as a result of meningeal irritation. A neurological examination will usually reveal other focal neurological signs although, however we emphasize that this is not always the case.

Arnold Chiari malformation is the name given to a group of anatomic variances which describe an inferiorly displaced cerebellum/brainstem into the spinal cord [11]. Four subtypes exist, as classified by Dr Hans Chiari in the 19th century [12]. Chiari I malformation, the most common of the Chiari subtypes, defines a low lying cerebellar tonsil, beneath the foramen magnum. It is this displacement, which causes compression of the cranial nuclei, resulting in vocal cord palsy.

The risk of aspiration with vocal cord paralysis is well proven and we do recommend the insertion of a nasogastric tube as a tool of prevention and speech therapy review. Up to one third of vocal cord palsy cases reviewed by Bhattacharyya et al. [13] showed radiologic evidence of aspiration, regardless of the cause of the paralysis.

The association between central sleep apnea and type I Chiari malformation is also well known [14]. Hershberger and Chidekel showed that sleep disordered breathing can be the only manifestation of a type I Chiari malformation. After neurological decompression, this patient had improvement but not resolution of her sleep disordered breathing. This highlighted the importance of a detailed sleep history when assessing these patients. Tools such as the Children's Sleep Habits Questionnaire by Owens JA, Spirito A et al. have been formed to assist in identifying this [15]. A sleep study should be considered in all individuals with a diagnosis of Arnold Chiari. This again portrays the variable presentations of Chiari malformations and high clinical suspicion required in cases such as this.

Management of this condition includes initially securing a safe airway. Daya et al. found that over 50% of cases of bilateral vocal cord palsy will require a tracheostomy to maintain a safe airway [16]. Posterior fossa decompression remains as the mainstay of therapy for symptomatic Chiari I malformation although variances do remain in management. In the presence of hydrocephalus, ventricular shunting may be deemed necessary. It is still unclear over the best timing of surgery, with early operative management now being favored [17]. Operative decisions should be tailored to the individual. Although despite this treatment, it is difficult to predict outcomes post-surgical intervention. Past literature shows that prognostically, bilateral palsy has worse outcomes compared to unilateral palsy. 73% of unilateral vocal cord palsy cases will resolve spontaneously compared to only 52% of bilateral vocal cord palsy estimated to resolve [8].

4. Conclusion

Stridor is a common symptom in young patients which can often be mistaken for reactive airways disease. Syrinx formation and Chiari I malformation, although uncommon in presenting solely as stridor, should certainly be considered. A posterior fossa decompression resulted in improvement of symptoms but not resolution of vocal cord immobility. It is expected that neurological recovery may take months if at all to resolve. This case highlights the importance of MRI as a tool for diagnosis in children with stridor and advise that is should be highly considered for all patients with vocal cord palsy of unknown etiology.

Fig. 4. Stridor workup.

Conflict of interest

We, the authors, have no conflicts of interest.

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Ethical approval

Ethics approval was not required as patient consent was obtained.

Consent

Written consent for use of de-identified information and photos has been obtained.

Author contribution

Saif Yousif - corresponding author.

Mark WAlsh - co-author, primary radiologist.
Hannah Burns - supervisor, primary surgeon and co-author.

Guarantor

Saif Yousif

Statement

I, Saif Yousif, would like to state that I am not the recipient of a research scholarship and this paper is not based on a previous communication to a society or meeting.

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