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Case Series

Ear, Nose and Throat Section

Bilateral Vocal Cord Palsy with Arnold Chiari Malformation: A Rare Case Series

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ABSTRACT

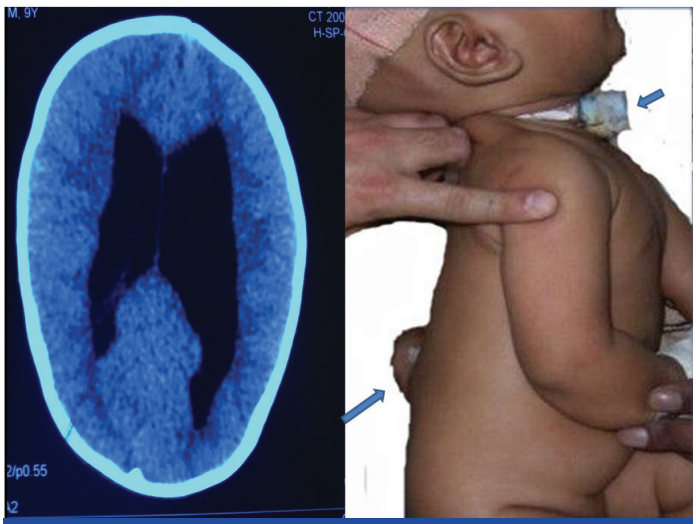
Stridor in paediatric age group is not an uncommon presentation to the ENT emergency. The range of differential diagnosis is vast. The presentation may vary from noisy breathing to severe respiratory distress and apnea. Early and meticulous diagnosis is crucial for the management as the condition may be life threatening. We report a rare case series of 3 infants with Arnold Chiari Malformation who presented to the hospital with stridor and were diagnosed with bilateral vocal cord palsy. These 3 infants had similar underlying neurological condition with hydrocephalus and raised intracranial pressure. Chiari malformation is the one of the most common congenital central nervous system anomaly associated with bilateral vocal cord paralysis. However, the presentation is rare. This article, thus, emphasizes the significance of early diagnosis and immediate management of this condition.

Keywords: Hydrocephalus, Myelodysplasia, Stridor

CASE SERIES

Case 1

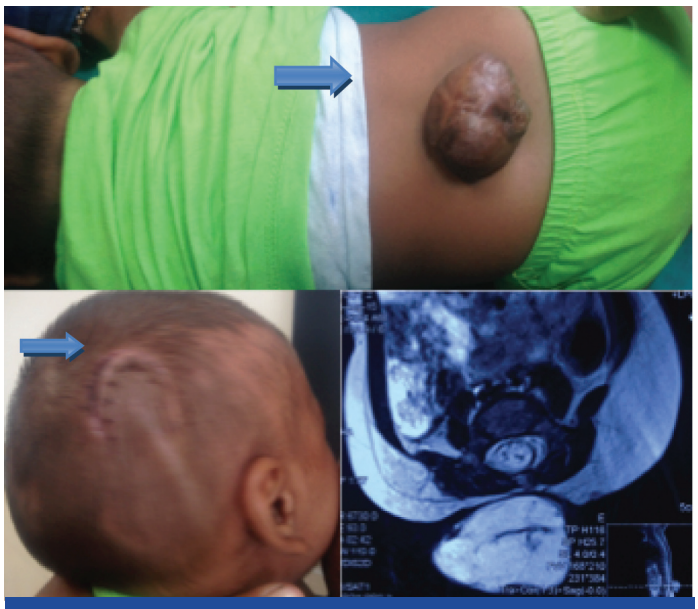
A four-month-old infant presented to the ENT emergency with an acute episode of difficulty in breathing for last 5 days. According to the parents, the child was previously taken to a local practitioner who misdiagnosed the respiratory distress as pneumonitis. The child was managed conservatively and failed to show any signs of improvement. On examination, he had inspiratory stridor, but there was no history of fever, trauma or upper respiratory tract infection. Examination revealed a lumbar meningomyelocele without any apparent neurological deficit. CT scan of head demonstrated severe hydrocephalus and tonsillar herniation due to Arnold Chiari malformation type II. Direct laryngoscopy revealed bilateral vocal cord palsy and the child was intubated with 3.5mm uncuffed endotracheal tube for an emergency ventriculo-peritoneal shunt procedure which was done to relieve the increased intracranial tension. After extubation, the stridor was persistent, due to which the child required reintubation, although he was breathing spontaneously without assisted mechanical ventilation. After observing for an hour, the patient was shifted to ICU and second trial of extubation was planned. After 48 hours, second trial of extubation also failed. Flexible laryngoscopy was carried out to confirm bilateral cord palsy and then the child was tracheostomized [Table/Fig-1]. The child was relieved of stridor and respiratory distress. Repeat direct laryngoscopy after 15 days showed improvement with return of right vocal cord movement and bilateral mobility of vocal cords were demonstrable after another week. Later, the child was taken up for repair of the lumbar meningomyelocele. The tracheostomy tube was removed after endoscopic laser fulguration of the suprastomal granulations.



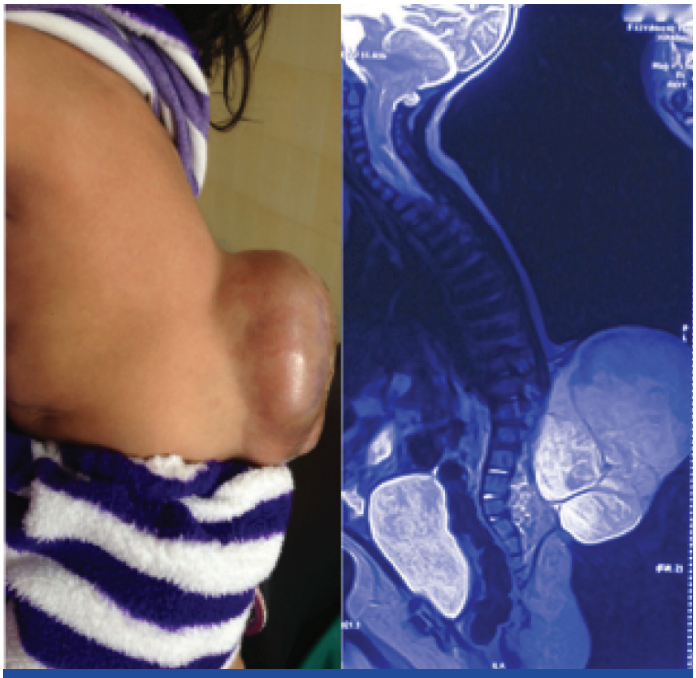
[Table/Fig-1]: Showing the hydrocephalus as in NCCT head of the child and clinical picture with tracheostomy tube in situ and the lumbar meningomyelocele.

Case 2

A nine-month-old child [Table/Fig-2] was brought to the ENT OPD with complaints of noisy breathing since last 2-3 weeks without any respiratory distress, no previous history of upper respiratory tract infection, fever and trauma. Patient had a lumbar meningomyelocele without any neurological deficit. MRI revealed type II Arnold Chiari malformation with lumbar meningomyelocele with syringohydromyelia with tonsillar herniation and hydrocephalus. On fibre optic laryngoscopic examination, vocal cords were found lying in paramedian position with no lateral movement. Child was



[Table/Fig-2]: Showing clinical and radiological picture of the second patient with lumbar meningomyelocele with theventriculo-peritoneal shunt scar.



[Table/Fig-3]: Showing the clinical photograph of the patient along with the MRI film suggestive of a large lumbar meningomyelocele.

taken up for emergency ventriculo-peritoneal shunt to relieve the raised intracranial tension, after which the stridor was relieved within 15 days. Repeat laryngoscopy showed improvement in vocal cord mobility. The child was taken up for repair of meningomyelocele in the second stage under general anaesthesia.

Case 3

Another nine-month-old child was brought to the ENT OPD with complaints of noisy breathing since 4-5 days with a previous history of upper respiratory tract infection. However, the child did not have any respiratory distress and maintained normal oxygen saturation on room air. On examination, the child had a large lumbosacral meningomyelocele with inspiratory stridor [Table/Fig-3], without any signs of neurological deficit. Right sided pre-auricular tags were also observed. MR brain and spine revealed type II Arnold Chiari malformation, with moderately dilated lateral and third ventricles with tonsillar herniation and compression of the spinal cord. On fibre optic bronchoscopic examination, patient was found to have bilateral abductor cord palsy. Before the child could be taken up for surgery, the meningomyelocele sac burst and there was cerebrospinal fluid leak, relieving the raised intracranial tension. Stridor resolved spontaneously after a week. Laryngoscopy was repeated and the vocal cord mobility was found to have improved. Later, the meningomyelocele was repaired.

DISCUSSION

Bilateral vocal cord palsy is the second most common cause of stridor, after laryngomalacia, accounting for 10-15% of cases [1]. Bilateral vocal cord palsy is often idiopathic; however it may be associated with neurological anomalies like Arnold Chiari malformation, intraventricular haemorrhage, meningo-encephalocele and hydrocephalus. It may also follow a traumatic forceps delivery, mediastinal surgery, or ligation of Patent ductus arteriosus [1]. Chiari first described different variations of hindbrain herniation through the foramen magnum in 1891 [2]. Type I Arnold Chiari malformations typically involve downward displacement of the cerebellum and cerebellar tonsils that produces a small posterior fossa. This malformation is often an isolated finding. Arnold Chiari II malformation involves downward displacement of the cerebellar vermis, medulla and fourth ventricle. This is strongly associated with hydrocephalus and myelodysplasia. These patients often have intrinsic brainstem histopathological changes that can produce symptoms of bulbar dysfunction (hindbrain dysfunction). Both types of Chiari malformation scan be easily identified on radiological imaging (computed tomography or magnetic resonance imaging) of the craniovertebral junction. Type III Chiari malformation is an encephalocervical-meningocele and type IV refers to cerebellar hypoplasia [3].

The Chiari II malformation is the leading cause of death in children with treated myelodysplasia during first 2 years of life [4]. They usually present with cranial nerve and brain stem dysfunction with dysphagia (92%), stridor or bilateral vocal cord paralysis (69%) or apnoea (54%) [5]. Charney and co-workers classified patients on the basis of their initial brain stem dysfunction and found that the patients with stridor alone, who were classified as Grade I, had minimum mortality. Grade II patients exhibiting stridor and apnoea possessed a 50% chance of recovery. Grade III patients who exhibited stridor, apnoea, cyanotic spells and dysphagia had minimal chances of recovery and a 40% chance of survival. Grade II, III are caused by more extensive brainstem damage due to haemorrhage, infarction and necrosis [6]. Holinger et al., have described a series of 21 children who had stridor associated with meningomyelocele, Arnold chiari malformation and hydrocephalus. Thirteen of these patients also presented with episodes of apnoea. Temporal association of vocal cord paralysis with raised intracranial pressure was noticed during the study [7].

Vocal cord paralysis with inspiratory stridor during infancy is a complication of the Chiari II malformation associated with meningo-myelocele, though rarely found with type I Chiari malformation [8]. This develops with acute downward or upward displacement of the malformed brainstem secondary to increased or decreased intracranial pressure. The treatment of these symptoms requires a decompression of the raised intracranial pressure, which reduces the pressure difference between the intracranial and the intraspinal compartments. Yamada et al., have reported the importance of immediate ventriculo-peritoneal shunting in 12 patients of Arnold Chiari malformation with stridor. This procedure provided symptomatic relief to the patients [9]. Solan and Glaisyer had described a case of a neonate who was born with meningomyelocele and had stridor. The cause of stridor was ascertained to be due to evolving hydrocephalus. Symptoms were relieved immediately after direct tapping of the cerebrospinal fluid from the right coronal horn via the coronal suture [10].

The diagnosis of bilateral vocal cord palsyis established by direct laryngoscopy performed without general anaesthesia. Chaten et al., have documented 9 patients who developed unilateral or bilateral vocal cord paralysis in a PICU setting. It was either due to iatrogenic trauma during thoracic surgery or underlying neurological condition with documented or suspected intracranial hypertension. They also discussed the significance of early visualisation of larynx in cases of children who became stridulous post extubation and had underlying neurological disorders associated with raised intracranial tension. Four patients out of seven with neurological disorder regained cord mobility in 4 months [11].

It is to be emphasized that in case of an emergency, endotracheal intubation or tracheotomy should be considered to establish an airway with immediate ventricular puncture. Ventricular shunting procedures to reduce intracranial pressures should ideally be performed within the first 48 hours after the onset of features of hindbrain dysfunction [5]. Delayed procedure reduces the likelihood of recovery of vocal cord paralysis. The degeneration of nucleus ambiguous leads to irreversible paralysis of vocal cords [6,12]. The bilateral vocal cord paralysis may be totally reversed if the intracranial pressure is decreased on time [5]. Glossopharyngeal, vagus and accessory nerves may be involved due to hindbrain dysfunction.

CONCLUSION

A possibility of bilateral vocal cord palsy should be speculated if stridor is observed in an infant with meningomyelocele. Laryngoscopic examination is imperative for diagnosis in children with stridor. Early decompression of the raised intracranial tension can reverse the palsy, alleviating the stridor and thus, the need for a tracheostomy. Hence, early recognition, diagnosis and management of these abnormalities may be lifesaving.

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