Case Report

Cervical Myelopathy in a Child: A Rare Cause of Hypoventilation Syndrome Presenting with Type 2 Respiratory Failure

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Abstract

Hypoventilation syndrome leading to Type 2 respiratory failure is not a rare cause of Pediatric Intensive Care Unit admission and mechanical ventilation. Common causes in pediatric population are Guillain–Barre syndrome and various central nervous system disorders such as encephalitis, traumatic brain injury, and drugs. Any injury or disease in the cervical cord can also produce respiratory paralysis causing respiratory failure. Here, we present two cases of mixed cerebral palsy with cervical myelopathy due to compression effect of fractured segments of first and second cervical vertebrae. Both of them presented with Type 2 respiratory failure.

Keywords: Cerebral palsy, cervical fracture, cervical spondylotic myelopathy, choreoathetoid cerebral palsy

Introduction

The center of respiration lies in the medulla and efferent fibers travel along the anterolateral part of the cervical cord. A chronic fracture or posterior dislocation of the odontoid process gradually compresses on the anterior segment of cervical canal, producing myelopathic changes in it, which may lead to respiratory muscle paralysis resulting in hypoventilation and eventually Type 2 respiratory failure.[1,2] Cerebral palsy (CP) patients with chronic choreoathetoid movement of the neck region are at greater risk of cervical instability.[3] The prolonged motion-related microtrauma might lead to fracture of the cervical vertebra resulting in a compressing injury to the medullary fibers and ultimately respiratory failure.[4] Here, we describe two such cases of cervical myelopathy in CP patients who presented to us in a similar manner.

Case Reports

Case 1

A 7 year old boy who is a known cerebral palsy patient, mixed type (spastic and dystonic) was admitted to the pediatric ward with chief complaints of fever and difficulty in breathing for last 1 day. The child was a known case of bilirubin encephalopathy with involvement of bilateral basal ganglia region. Quadriplegic since birth, he also had a history of abnormal neck movements noticed by the parents for the last 4 months. Initially, the child was admitted and managed in general pediatric ward; however, due to progressively increasing distress and labored breathing, he was shifted to Pediatric Intensive Care Unit (PICU). After 2 days of conservative management in the PICU, the condition of the patient further worsened, and there were very sluggish respiratory efforts with severe respiratory acidosis in arterial blood gas (ABG) analysis; hence, he was intubated and mechanically ventilated. Gradually, the acidosis was corrected, and the blood gas parameters improved. He was weaned off after 3 days but within a couple of hours following extubation, the child started hypoventilating and desaturating again. The ABG revealed profound respiratory acidosis. Thus, he was intubated. The Chest X-ray did not reveal any pulmonary pathology, and sepsis was also ruled out. He always needed minimum ventilator support. Keeping in mind, these repeated episodes of hypoventilation, and a repeat magnetic resonance imaging of the brain and cervical spine was planned to pickup any new central nervous system insult on the previous pathology. MRI of the cervical spine revealed a fracture

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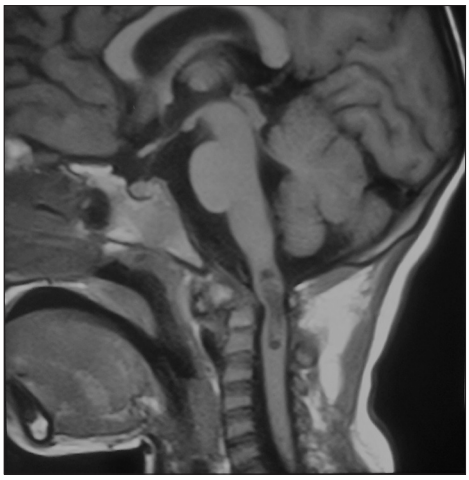

Figure 1: Magnetic resonance imaging of the brain and cervical spine showing a fracture involving the odontoid process and the fractured segment displacing the body of C2 vertebra posteriorly compressing the cervicomedullary canal

Figure 2: Cervical column stabilization with rigid cervical collar

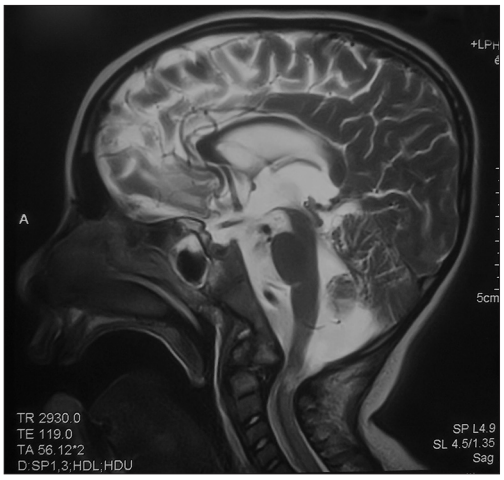

Figure 3: Magnetic resonance imaging of the brain and cervical spine showing a fracture of C2 vertebra with posterior dislocation of the odontoid process impinging on the cervicomedullary junction

involving the odontoid process and the fractured segment displacing the body of C2 vertebra posteriorly compressing the cervicomedullary canal, thereby producing myelomalacic changes in it [Figure 1]. The cervical spine of the child was stabilized and immobilized with a rigid cervical collar [Figure 2]. Surgical options with poor outcomes were discussed with the parents. Realizing a poor prognosis, the parents took the child to their hometown and he expired there after a couple of days.

Case 2

A 12-year-old boy, known case of quadriplegic CP, presented to emergency with complaint of difficulty in breathing for the last 1 day. The patient was seen to have a gasping respiration with abnormal neck movements and hence he was intubated in emergency and shifted to the PICU and mechanically ventilated. His ABG before intubation revealed severe respiratory acidosis with Type 2 respiratory failure. Similar choreoathetoid neck movements were also noticed in this child and the parents had also noticed them for the last 6 months. The chest X-ray showed collapse of the right lung which recovered with nebulization and chest physiotherapy. The respiratory acidosis resolved gradually, and ventilator settings required were also minimal. After 5 days of intubation, we tried to wean off the patient gradually and extubated the child. He was put-on noninvasive ventilation, but he was having very poor respiratory effort with repeated desaturations; a blood gas analysis at that time revealed a severe respiratory acidosis; hence, he had to be intubated again. Considering a similar background and presentation, a similar pathology behind his hypoventilation was anticipated, and MRI of the brain with cervical spine was planned. MRI of this child also revealed an old fracture of C2 vertebra with posterior dislocation of the odontoid process impinging on the cervicomedullary junction [Figure 3]. This finding was quite identical to the previous case, and this patient was also immobilized with a rigid cervical collar. The boy was ventilated for 4 weeks in our PICU and later shifted to another low-cost hospital for financial constraints where he expired after 2 weeks.

Discussion

Hypoventilation syndrome due to cervical myelopathy as a result of cervical vertebral fracture in pediatric patients with choreoathetoid type of CP is rarely reported. CP patients are vulnerable to fracture of cervical region due to continuous movement of their neck. Such movements produce microinjuries at the level of the cervicomedullary junction which contains all the vital neural pathways.[4] These patients may go undiagnosed for a long period because of very slow progression of the disease process and also due to masking by preexisting neurological insults as seen in our patient with bilirubin encephalopathy.[5] Cervical myelopathy, secondary to choreoathetoid CP, has been reported to occur at an earlier age group usually in the 4th decade compared to the general population,[3] but we got such changes in the pediatric age group (age <15 years). Both of our cases presented to us with Type 2 respiratory failure, and there was a definite preceding

history of abnormal neck movements noticed few months before admission. Clinical presentations are reported to be quite varied depending on the level of spine affected and does not seem to vary much in different age groups.[6] In studies with middle-aged cervical spondylotic myelopathy patients, various symptoms of cervical myelopathy have been reported including sensory changes, reflex abnormalities, motor disturbances, autonomic instabilities, and even acute spinal injuries in some.[3,6] Therefore, clinical features such as any new onset abnormal neck movements should not be overlooked, and one should also periodically check for the development of any additional sensory or motor symptoms to have an earlier diagnosis. In a radiological study of 180 patients of athetoid CP by Harada *et al*., involvement was noted to be at the C3/4, C4/5, and C5/6 levels mostly whereas in both of our cases, C1/2 junction was affected.[6-8] Surgical intervention had a better outcome than conservative management in such patients as reported by Lee *et al*.[9] Earlier surgical procedures such as anterior and posterior spinal fusions of different methods also yield a better outcome,[9,10] but overall prognosis remains poor. Multilevel anterior decompression and fusion, however, was reported to have good results but long-term complications were also reported during follow-up.[3] Postoperative application of halo vest has been used for effective immobilization. Conservative management with cervical collar and medicines such as botulinum toxin-induced denervation were also tried but with poor outcomes.[9,11] Both of our cases were conservatively managed with cervical immobilization as scope for surgical intervention was not there.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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