Case Report

Reports of a Conus Cauda Tumor with Holocord Syrinx in an Adolescent Girl

Smriti Sinha, Prathibha Shankar Ashwini, Pelala Nayan Baba, Rathika Damodar Shenoy

Department of Pediatrics, K. S. Hegde Medical Academy (KSHEMA), Mangaluru, Karnataka, India

Abstract

Introduction: Spinal cord ependymoma seldom presents with holocord syringomyelia in pediatric age-group. Association of ependymoma with a lipoma is also rare. The child presented critically ill with polymicrobial pneumonia, and the neurologic findings were missed until recovery. We report a case highlighting these findings. Case Report: A 16-year-old adolescent presented critically ill with respiratory failure due to severe pneumonia. Evaluation showed fungal pneumonia with secondary bacterial infection. On recovery, she had nasal regurgitation and required nasogastric tube feeding. Examination showed palatal and left vocal cord palsy. Neuroimaging of brain showed conus cauda tumor with syringobulbia and holocord multiseptate syrinx extending from medulla to filum terminale with lipoma. Histopathology examination showed myxopapillary ependymoma. She improved with excision and postoperative radiotherapy. Discussion: Our case is a rare report of an adolescent girl with conus cauda tumor and holocord syrinx.

Keywords: Fungal pneumonia, palatal palsy, syringobulbia, syringomyelia

Introduction

Syringomyelia is a spinal cord lesion involving cystic cavitation of two or more spinal segments. The prevalence is 8.4 per 100,000 population with a lesser incidence in pediatric age-group. The presentation is mainly dependent on the extent of spinal involvement. We report an adolescent girl with syringobulbia and holocord syrinx following spinal cord ependymoma for its rarity. She presented with aspiration pneumonia due to palatal palsy.

Case Report

A 16-year-old girl presented with fever in the last 10 days and severe respiratory distress and bilateral diffuse crepititation. A history of recent-onset generalized weakness, poor feeding, and weight loss was present. At presentation, she was emaciated with a weight of 27 kg and body mass index of 11.44 kg/m^2 (<third centile). Owing to the critical nature of her presentation, she required intensive care and ventilatory support. As the blood counts suggested bacterial infection, she was started on ceftriaxone and vancomycin while awaiting culture results. Arterial blood gas showed respiratory acidosis with hypoxia. Other blood parameters, including serology for human immunodeficiency virus, were normal. Chest X-ray showed multiple flocculent opacities in bilateral lung fields.

Pulmonology consultation was carried out. High-resolution computed tomography chest showed multiple bilateral nodular opacities characteristic of fungal pneumonia [Figure 1]. There was also right lower lobe collapse, air bronchogram, peribronchial interstitial thickening, and mild pleural effusion. Fluconazole was added, and the child started to show improvement.

Repeat chest X-ray, 48h after admission, showed significant clearing of the pneumonia. Cultures grew *Candida albicans* susceptible to fluconazole as well as...
**Discussion**

Most of the syringomyelia cases in children are associated with Chiari I malformation. Only 5.5% are caused by tumors, most commonly ependymomas, hemangioblastomas, and cavernomas.[2] A quarter of intramedullary spinal tumors may be associated with syringomyelia, whereas 50% of cases of ependymoma may have an associated syrinx.[3] However, ependymoma presenting with holocord syringomyelia is extremely rare with no such reported case in pediatric age-group.

The presentation of syringomyelia depends on the extent of spinal involvement, most common presentation being pain, sensory disturbances, and motor weakness. However, there have been previous cases reported where syringomyelia and syringobulbia have presented with cranial nerve involvement.[4,5] Most commonly vagus and hypoglossal followed by nerves involved in

*Acinetobacter baumannii*, and *Pseudomonas aeruginosa* sensitive to tigecycline. Gradual improvement was observed with the escalation of antibiotic, and she was weaned off ventilator within 72 h.

Following extubation, the child’s voice was hoarse; she had difficulty in swallowing and could be fed only via orogastric feeding tube. A bilateral palatal palsy was noted. No other cranial nerve involvement was observed, and jaw jerk was normal. No evidence of Horner syndrome or dysautonomia was noted. However, she had persistent generalized limb weakness with diminished deep tendon reflexes and decreased pain and temperature sensations over all limbs. Bladder and bowel sphincters were intact. Laryngoscopy showed left vocal cord palsy.

On the basis of the aforementioned findings, postinfectious polynieuritis was suspected, and a nerve conduction velocity was carried out, which was within normal limits. Magnetic resonance imaging (MRI) of brain and spinal cord showed a holocord multiseptate syrinx extending from medulla to filum terminale [Figure 2A]. An enhancing intramedullary lesion was observed in the conus medullaris region (L1-L2) [Figure 2B] along with a lipoma at filum terminale [Figure 2C].

Laminectomy with decompression and excision of the space-occupying lesion was performed. As the vascular lesion was found to be adherent to the cord, only partial excision could be performed and after surgery, radiotherapy was started. Histopathology examination showed cuboid to elongated tumor cells arranged in papillary pattern with myxomatous stroma and fine nuclear chromatin, suggestive of myxopapillary ependymoma–conus medullaris, WHO (World Health Organization) Grade I.

**Figure 1:** High-resolution computed tomography of the chest showing bilateral nodular opacities characteristic of fungal pneumonia, the largest indicated by the arrow

**Figure 2:** MRI of the brain and spinal cord, sagittal images showing (A) syringobulbia and multiseptate syrinx, (B) conus medullaris tumor (arrow) with contrast enhancement, and (C) lipoma (arrow) at filum terminale

Her voice, gag reflex, and limb weakness improved within few days, and oral feeds were gradually built up. At first follow-up a month after radiotherapy, the child had gained 7 kg weight, and her demeanor had markedly improved. A repeat MRI, conducted 6 months after radiotherapy, showed resolution of the syringobulbia and cord syrinx [Figure 3] without any recurrence of tumor. The child was doing well over 1-year follow-up.
ocular movements.\textsuperscript{[6]} However, initial presentation with only bulbar palsy is extremely rare, with only one prior similar case report, in a 20-year-old female patient.\textsuperscript{[7]}

Our case was unique as at the initial presentation to the pediatric emergency, no history or physical finding to point toward a spinal pathology was observed. A high index of suspicion and timely diagnostic imaging may help in an early diagnosis. Only after adequate stabilization, a history of dysphagia became known and related physical findings could be noted. As in our case, the initial presentation of syringomyelia and syringobulbia maybe a complication of dysphagia similar to aspiration pneumonia and may delay the identification of underlying cause. The underlying malnutrition probably predisposed her to polymicrobial pneumonia including fungal etiology.

Our case also had a concomitant filum terminale lipoma with intramedullary ependymoma and adherent cord, which is a very rare association. This finding is probably incidental. It has been postulated that a common embryonic pathway may contribute to such development.\textsuperscript{[8]} Radiotherapy after excision has been postulated in recent years to have prognostic benefits.\textsuperscript{[9]} Our case received radiotherapy after excision, as excision was incomplete in the presence of adherent cord. Over a follow-up of more than a year, no recurrences were noted.

This case, to the best of our knowledge, is the first reported case of ependymoma associated with holocord syringomyelia and syringobulbia in pediatric age-group. We present this case to highlight that spinal cord lesions may present even without apparent central nervous system involvement.

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**Conflicts of interest**

There are no conflicts of interest.

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