# Syringobulbia with Syringomyelia Presenting as Unilateral Multiple Cranial Nerve Palsies with Ipsilateral Hemiparesis in an Adult: A Rare Case and Literature Review

To the Editor,

Syringobulbia (SB) denotes a longitudinally oriented fluid-filled cavity within the brainstem. It is usually associated with syringomyelia (SM), which is more common.[1,2]

Many conditions can lead to syringobulbia, including posterior fossa or spinal cord neoplasms and inflammatory disorders (e.g., arachnoiditis and meningitis). There are several possibilities for the pathology and a need for more agreement about what SB is. Because of the rarity of this condition, its manifestations, treatment methods, and long-term prognosis still need to be established. Moreover, unilateral multiple cranial nerve palsies due to SB have never been reported.

We describe the case of a 26-year-old man who was previously healthy. He presented with a two-year history of progressive weakness of the right upper and lower limbs, stiffness, and difficulty clearing the ground. There has been a progressive decline in his walking capacity, and now, he can walk only 100 meters without rest. Along with this, he had double vision on the right lateral gaze, deviation of the angle of the mouth toward the left side, difficulty swallowing (worse with fluids), and dysphonia in the form of stuttering and hoarseness. He had right upper limb and right lower limb stiffness with hemiparetic gait. A double vision was limited to the right lateral gaze. He had a deviation of the angle of his mouth toward the left side and difficulty blowing. There was no history of anosmia, parosmia, diminution of vision, change in taste sensation, hearing loss, or tinnitus. He did not have any occipital headache, vertigo, trigeminal paresthesia, or hypoesthesia to pain and temperature. There was no history of recent or remote trauma to the head or neck, no prior account of the posterior fossa, spinal cord neoplasms, or inflammatory disorders (e.g., arachnoiditis and meningitis).

On physical examination, he was conscious, well oriented to time, place, and person, and had higher mental functions within normal limits. On cranial nerve examination, he had gaze restriction on the right lateral gaze and right facial palsy of lower motor neuron type. There was palatal palsy on the right side. On direct laryngoscopy, he had right true vocal cord palsy. On tongue protrusion, he had tongue deviation towards the right side. On motor examination, the bulk was well preserved; there was grade 3 spasticity on the modified Ashworth scale on the right side. The power was 4/5 on the Medical Research Council (MRC) grading. On the right side, reflexes were brisk (3+), abdominal reflexes were absent, and the plantar

was extensor. He also had difficulty walking with a hemiparetic gait on the right side. There was no apparent sensory loss. On the left side, the examination was unremarkable.

A complete blood count, electrolytes, glucose, renal and hepatic function tests, and inflammatory markers were all within the normal range. Serum ESR, serum CRP, RA factor, ANA, anti-dsDNA, anti-rho, anti-La, anti-centromere, and serum ACE levels were within the normal range. We did a lumbar puncture, and the CSF opening pressure was 10 cm of CSF. CSF showed a total of 10 cells, three neutrophils, and seven lymphocytes; CSF sugar/RBS was 80/120 mg/dl; and protein was 35 mg/dl. Gram stain and bacterial culture were negative, and GeneXpert, India ink, and Cryptococcal antigen were negative. CSF cytology did not show any malignant cells. The pure tone audiometry was within normal limits—his MRI findings are shown in Figure 1. In the abovementioned patient, a diagnostic endoscopic third ventriculostomy (ETV) was planned, followed possibly by a shunt with a guarded prognosis, but the patient did not consent to the procedure. On the six-month follow-up, the patient's condition remained the same.

In all forms of SM, there appears to be a partial obstruction or constriction of the subarachnoid space. However, the mechanism and route of this fluid transport, which results in syrinx cavity formation, have not been elucidated. There are various theories to explain its complex pathophysiology. Gardner proposed that the tonsillar descent was due to the "water hammer" effect of arterial pulsations transmitted to the CSF. Bertrand showed that the dural tube pressures are transmitted into the cavity of syringomyelia or hydromyelia, thus forcing the fluid to dissect into the brainstem.[3] Williams[4,5] explained experimentally that a pressure differential exists after the tonsils have descended and that this pressure differential favors progressive tonsillar descent. He termed this as "cranial–spinal pressure dissociation." Current thinking is that in the face of an obstruction at the foramen magnum, some form of transparenchymal fluid migration occurs through the Virchow–Robin spaces of the spinal cord; a partially blocked CSF system at the craniovertebral junction leads to fluid within the syrinx cavity dissecting in an upward axial direction, leading to SB under pressure differentials generated by epidural venous distention.

There needs to be more literature regarding patients with non-ACM SB presenting as unilateral multiple cranial nerve palsies. Morgan *et al*.[6] mentioned 56 patients with syringobulbia, of whom 46 had tonsillar descent. The authors described patients having various cranial nerve palsies, but there is no tabular documentation of the combination of cranial nerve palsies that the patients had.

We postulate that this patient had a congenital or acquired arachnoid veil, which led to cranial hydrocephalus and caudal dissection of syrinx into the brainstem and spinal cord. There were many unique features in our case:

a. Age of presentation: Most of the patients present in the pediatric age group, but our patient presented at 26 years of age, and his symptoms were progressive for two years.
b. There was no association with the Arnold–Chiari malformation, a crowded posterior fossa, or any other craniovertebral junction anomaly.
c. The SB extended rostrally from the medulla up to the lower pons on the right side.

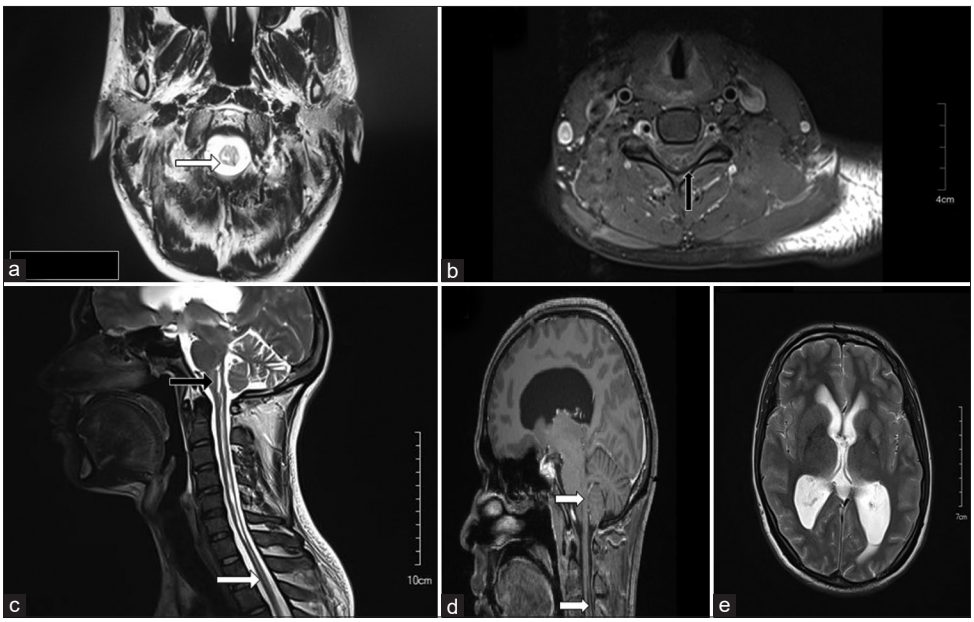


**Figure 1:** (a) T2-weighted axial images showing syrinx in the cervical cord at the C5 level. (b) Post-contrast T1-weighted axial images showing syrinx in the cervical cord at the C5 level. (c) T1-weighted sagittal cervical spinal cord MRI showing linear T2 hyperintensity involving the cervical cord extending up to the medulla and lower pons consistent with syringomyelia with syringobulbia (arrows). (d) T1-weighted sagittal brain MRI with gadolinium contrast showing no abnormal enhancement along the cord and visible brain parenchyma (arrows). (e) T2-weighted axial brain MRI showing mild dilation of ventricular system with no periventricular ooze (arrows). *Black arrow shows syringobulbia, while the white arrow shows syringomyelia

d. Unilateral simultaneous involvement of the 6th, 7th, 9th, 10th, and 12th nerves: This is theoretically possible but has never been reported previously in a single patient, to the best of our knowledge.

We hypothesize that CSF flow dynamics were disturbed because of a congenital or acquired arachnoid obstruction within the third ventricle. This resulted in a rostral communicating hydrocephalus involving both lateral ventricles and a caudal syrinx cavity dissecting into the midbrain, pons, and medulla and extending into the central canal of the spinal cord up to the lower cervical level. The syrinx tends to dissect to any particular side; in our patient, it is preferably dissected toward the right side when moving cranially, causing such a unilateral presentation.

## Ethics approval and consent to participate

The institutional review board does not require ethics approval for case reports.

## Consent to participate

Written informed consent was obtained from the patient to publish clinical details, photographs, videos, and imaging.

## Consent for publication

Not applicable.

## Authors' contributions

AA and AKK designed and conceptualized the study. CDS, AA, and AKK analyzed or interpreted the data. All authors drafted and revised the manuscript, acquired the data and revised the manuscript for intellectual content, and read and approved the final manuscript.

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**Abbreviations:** ACM = Arnold–Chiari malformation; ETV = endoscopic third ventriculostomy; MRI = magnetic resonance imaging; SB = syringobulbia

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## Conflicts of interest

There are no conflicts of interest.

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## References

1. Menezes AH, Greenlee JDW, Dlouhy BJ. Syringobulbia in pediatric patients with Chiari malformation type I. J Neurosurg Pediatr 2018;22:52-60.
2. Shen J, Shen J, Huang K, Wu Y, Pan J, Zhan R. Syringobulbia in patients with chiari malformation type I: A systematic review. BioMed Res Int 2019;1-8. doi: 10.1155/2019/4829102.
3. Bertrand G. Dynamic factors in the evolution of syringomyelia and syringobulbia. Neurosurgery 1973;20:322-33.
4. Williams B. Progress in syringomyelia. Neurol Res 1986;8:130-45.
5. Williams B. On the pathogenesis of syringomyelia: A review. J R Soc Med 1980;73:798-806.
6. Morgan D, Williams B. Syringobulbia: A surgical appraisal. J Neurol Neurosurg Psychiatry 1992;55:1132-41.

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