

Cervical Myelopathy at Craniovertebral Junction Secondary to Compression by Posterior Atlanto-occipital Membrane: A Case Report

venu madhav pinninti¹, ramakrishna narayanan²

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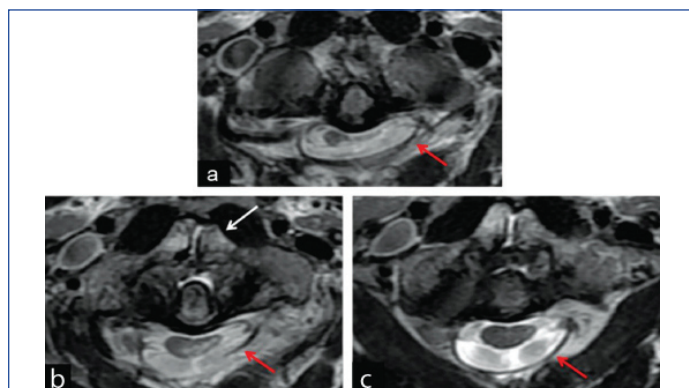
ABSTRACT

The Craniovertebral Junction (CVJ) comprises of the skull base (occipital bone), upper cervical spine (atlas and axis) and surrounding neurovascular structures, including the brainstem, spinal cord and vertebral arteries. Its stability depends on strong ligaments, membranes and bony anatomy, allowing key movements like rotation and flexion-extension. Cervical myelopathy significantly affects quality of life and can be caused by both common and rare causes. Common causes include degenerative disc disease, cervical spondylosis, spinal stenosis, herniated discs and trauma, all of which lead to compression of the spinal cord due to wear, injury, or misalignment in the cervical spine. Less common causes involve congenital abnormalities in the spinal structure, tumours, infections, rheumatoid arthritis, atlanto-occipital membrane issues and Chiari malformation, all of which lead to spinal cord compression. Despite a thorough diagnostic evaluation, around 20% of cases remain idiopathic. The authors hereby, present a case of a 59-year-old male with suboccipital and neck pain radiating predominantly to the left upper and lower limbs, accompanied by paraesthesias. Imaging revealed a congenital anomaly of the atlas, characterised by the absence of the left half of the posterior arch (Currarino type B). This structural defect led to stretching and ventral displacement of the Posterior Atlanto-occipital Membrane (PAOM), resulting in compression and myelomalacia of left hemispinal cord.

Keywords: Craniovertebral junction anomalies, Currarino congenital anomalies of atlas, Currarino type B

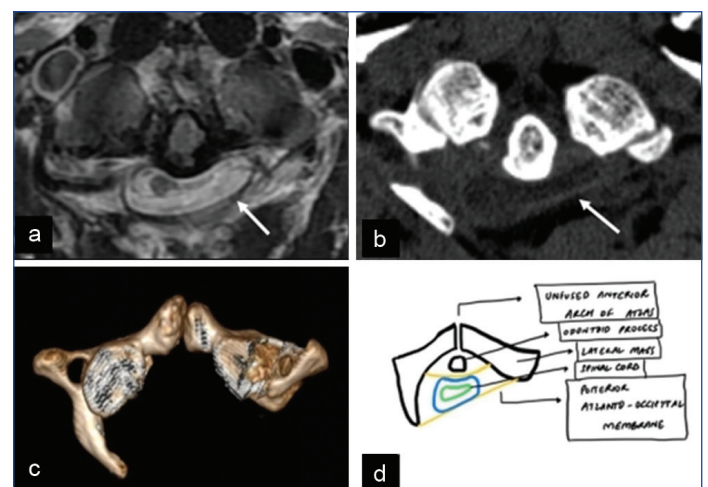
CASE REPORT

A 59-year-old male presented with a one-year history of suboccipital, neck and back pain predominantly radiating to the left upper and lower limbs. This was associated with paresthesia of left upper limb. He has a 30-pack-year smoking history (one pack per day) and reports occasional alcohol consumption. There are no known comorbidities, significant birth or family history, or prior spinal surgeries. Given the symptoms, the patient was referred for spinal imaging. Magnetic Resonance Imaging (MRI) of the cervical spine revealed ventral displacement of the PAOM, which narrowed the posterior subarachnoid space on the left side and selectively indented the left hemicord. The cord at this level exhibited myelomalacia changes, characterised by spinal cord atrophy and T2 hyperintensity [Table/Fig-1].

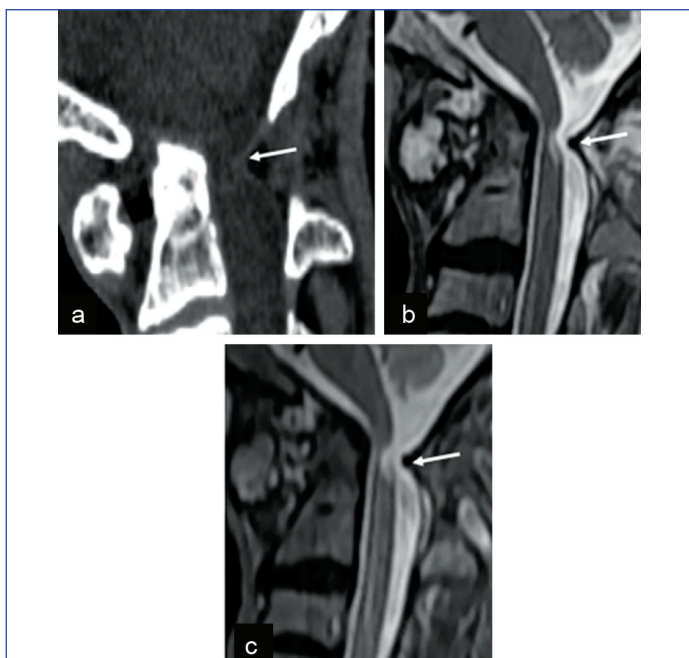


[Table/Fig-1]: Limited axial T2-weighted MRI sections of the Craniovertebral Junction (CVJ) from cranial to caudal views: a-c) Show a stretched Posterior Atlanto-occipital Membrane (PAOM) attached to the remnant right half of the posterior arch and the left lateral mass, causing compression of the spinal cord (red arrow), predominantly over the left hemicord. Associated hyperintensity within the cord suggests focal oedema or myelomalacia; b) Demonstrates non fusion of the anterior arch of the atlas (white arrow) and absence of the left half of the posterior arch.

To identify the cause of ventral displacement of the PAOM, a limited Computed Tomography (CT) scan of the cervical spine was performed with the neck in mild flexion. The scan revealed non fusion of the anterior arch of the atlas and absence of the left half of the posterior arch, consistent with a Currarino type B posterior arch anomaly. Additionally, there was ventral displacement of the PAOM on the left-side, which was seen directly attaching to the left lateral mass of the atlas. This displacement resulted in severe narrowing of the spinal canal on the left-side [Table/Fig-2,3].



[Table/Fig-2]: a, b) Representative axial T2-weighted MRI and CT sections demonstrating a stretched and ventrally displaced Posterior Atlanto-occipital Membrane (PAOM) attached to the right half of the posterior arch of the atlas and the left lateral mass, compressing the spinal cord (white arrow), predominantly over the left hemicord. Hyperintensity within the cord along with atrophy suggests myelomalacia; c) 3D reformatted CT image showing an unfused anterior arch and Currarino type B anomaly of the posterior arch of the atlas; d) Schematic diagram illustrating corresponding anatomical and pathological findings.



[Table/Fig-3]: a) Sagittal CT image showing a defect in the posterior arch of the atlas; b, c) Sagittal T2-weighted and Short Tau Inversion Recovery (STIR) MRI images of the Craniovertebral Junction (CVJ) demonstrating a stretched and tented Posterior Atlanto-occipital Membrane (PAOM) (white arrow) compressing the spinal cord.

Based on these findings, the authors proposed the possibility of dynamic left hemicord compression caused by a stretched and ventrally displaced PAOM, secondary to a unilateral defect in the left half of posterior arch of the atlas. The patient was referred to the Neurosurgery Department for further management and was planned for elective decompression with occipito-cervical fusion for spine stability.

DISCUSSION

Cervical myelopathy is a progressive spinal cord disorder that significantly impairs quality of life, commonly presenting with motor, sensory and autonomic dysfunction. It is most frequently caused by degenerative changes in the cervical spine, which account for approximately 55% of cases. These include cervical spondylosis, degenerative disc disease, spinal stenosis, herniated discs and traumatic injuries. Less commonly, cervical myelopathy arises from congenital or acquired structural abnormalities, including congenital cervical canal stenosis, spinal cord tumours, infections (e.g., tuberculosis, schistosomiasis), rheumatoid arthritis affecting the cervical spine, atlantoaxial or atlantooccipital instability and Chiari malformation- together comprising about 25% of cases. Despite a thorough diagnostic evaluation, around 20% of cases remain idiopathic [1].

Among the causes of spinal cord compression and myelomalacia are congenital anomalies of the atlas. The atlas forms from three ossification centres: one anterior, which gives rise to the anterior tubercle and two lateral centres that develop into the lateral masses and posterior arch [2]. Because the posterior arch originates from two separate centres, anomalies in this region are more frequently observed than those in the anterior arch [3].

Currarino G et al., classified posterior arch anomalies of the atlas into five types, based on the degree of arch absence and the presence or absence of the posterior tubercle. These types include:

Type A: Median clefts of the posterior arch;

Type B: Unilateral defects of varying extent;

Type C: Bilateral defects;

Type D: Absence of the posterior arch with a persistent posterior tubercle;

Type E: Complete agenesis of the posterior arch, including the posterior tubercle [4].

The PAOM is a broad, fan-shaped ligament extending from the posterior arch of C1 to the posterior margin of the foramen magnum. Its central fibres are thicker, forming a distinct band from the posterior tubercle of C1 to the opisthion. On sagittal MRI, it appears as a thin V-shaped structure that blends with the ligamentum flavum below C1 [5].

Cervical myelopathy is typically associated with etiologies such as degenerative disc disease, cervical spondylosis, spinal stenosis, herniated discs and trauma. However, the present case is unique in that the myelopathy arises from an aberrant anatomical relationship between the PAOM and the left lateral mass, secondary to the congenital absence of the left half of the posterior arch of the atlas. This anomaly has resulted in the PAOM becoming pathologically stretched and tented, exerting localised pressure on the left hemicord. The resultant focal cord compression has led to spinal cord atrophy and myelomalacia at this level. Such a mechanism is exceedingly rare and emphasises the critical role of high-resolution cross-sectional imaging and detailed anatomical evaluation in diagnosing atypical causes of cervical myelopathy.

CONCLUSION(S)

The authors present an exceptionally rare case of unfused anterior atlas arch with posterior arch hypoplasia (left-sided), associated with a type B Currarino variant. This complex anomaly led to stretching and compression of the PAOM, resulting in dynamic spinal cord compression. The present case also underscores the radiologist's pivotal role in diagnosing congenital cervical anomalies, where CT-MRI fusion is essential for identifying the structural aetiologies of myelopathy.

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PARTICULARS OF CONTRIBUTORS:

1. Junior Resident, Department of Radiodiagnosis, Nizam's Institute of Medical Sciences, Hyderabad, Telangana, India.
2. Additional Professor, Department of Radiodiagnosis, Nizam's Institute of Medical Sciences, Hyderabad, Telangana, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Venu Madhav Pinninti,
Plot No. 93, Road No. 16, Snehapuri Colony, Hyderabad-500076, Telangana, India.
E-mail: venumadhav94922@gmail.com

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Nov 06, 2025
- Manual Googling: Feb 07, 2026
- iThenticate Software: Feb 09, 2026 (1%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

Date of Submission: Oct 07, 2025

Date of Peer Review: Jan 13, 2026

Date of Acceptance: Feb 10, 2026

Date of Publishing: May 01, 2026