MedPeer Publishers
Empowering Evidence-Based Medicine Through Peer-Reviewed Excellence

Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

Hidden at the Craniovertebral Junction: Atlantoaxial Instability with Cervicomedullary Compression in a Child with Multiple Congenital Anomalies

DOI: 10.70780/medpeer.000QGT9

AUTHORS AND AFFILIATION

Mehdi Salmane[1], Alia Yassine Kassab[1], Lina Lasri[1], Lina Belkouchi[1], Siham El Haddad[1], Nazik Allali[1], Latifa Chat[1]

[1] Department of Radiology, Mother-Child Hospital, Mohammed V University, Rabat, Morocco

Corresponding author : Mehdi Salmane.

ABSTRACT

Atlantoaxial instability is an uncommon but potentially life-threatening condition in children, particularly when associated with congenital or syndromic disorders. We report the case of a 9-year-old child presenting with orthopedic malformations, encopresis, and facial dysmorphism. Cervical spine MRI revealed C1–C2 dislocation with marked widening of the atlantodental interval, associated with hypertrophic soft tissue surrounding the odontoid process causing narrowing of the foramen magnum and compression of the cervicomedullary junction. Associated T2 hyperintensity within the spinal cord suggested chronic myelopathic changes. This case highlights the importance of imaging evaluation of the craniovertebral junction in children with complex congenital abnormalities and neurological manifestations.

KEYWORDS :

Atlantoaxial instability, C1–C2 dislocation, Craniovertebral junction, Pediatric myelopathy, Cervicomedullary compression.

MAIN ARTICLE

INTRODUCTION

Atlantoaxial instability refers to abnormal mobility between the atlas (C1) and the axis (C2) resulting from congenital, developmental, inflammatory, or traumatic abnormalities. Although rare in children, it may lead to progressive spinal cord compression and irreversible neurological deficits if not recognized early [1,2].

Magnetic resonance imaging plays a crucial role in evaluating the degree of instability, associated soft-tissue abnormalities, and spinal cord involvement. Early diagnosis is essential because surgical stabilization may prevent permanent neurological sequelae [2,3].

CLINICAL PRESENTATION

A 9-year-old child was referred for MRI evaluation in the setting of multiple orthopedic malformations, chronic encopresis, and facial dysmorphism.

Neurological examination revealed gait abnormalities and progressive motor difficulties. Given the complex clinical presentation, MRI of the craniovertebral junction and cervical spine was performed.

MRI Findings

MRI demonstrated:

- Atlantoaxial dislocation with widening of the atlantodental interval measuring approximately 8 mm.
- Marked hypertrophy of the peri-odontoid soft tissues surrounding the odontoid process.
- Significant narrowing of the foramen magnum.
- Compression of the cervicomedullary junction by the hypertrophic tissue.
- Focal T2-weighted hyperintensity within the upper cervical spinal cord at the site of compression, consistent with spinal cord suffering (compressive myelopathy).

These findings were consistent with severe atlantoaxial instability complicated by chronic cervicomedullary compression and spinal cord injury.

DISCUSSION

Atlantoaxial instability in childhood may occur in association with congenital skeletal abnormalities, connective tissue disorders, syndromic conditions, and developmental anomalies of the craniovertebral junction [1,4].

The atlantodental interval is a key radiological marker of instability. In children, widening beyond normal limits suggests disruption of the stabilizing ligamentous structures and may result in excessive motion between C1 and C2 [2].

In the present case, MRI demonstrated marked widening of the atlantodental interval associated with hypertrophic peri-odontoid soft tissue causing compression of the cervicomedullary junction. The presence of spinal cord T2 hyperintensity indicated established myelopathic changes, emphasizing the chronic nature of the compression [3].

The association of orthopedic malformations, facial dysmorphism, and atlantoaxial instability raises suspicion for an underlying syndromic or genetic skeletal disorder. Several conditions, including skeletal dysplasias and connective tissue disorders, have been reported in association with craniovertebral junction abnormalities [4,5].

MRI is particularly valuable because it simultaneously evaluates:

- Bony alignment.
- Ligamentous and soft-tissue abnormalities.
- Degree of neural compression.
- Spinal cord integrity.

Recognition of spinal cord signal abnormalities is crucial because they may correlate with neurological impairment and influence surgical decision-making [3].

CONCLUSION

This case illustrates severe atlantoaxial instability with cervicomedullary compression and spinal cord suffering in a child with multiple congenital anomalies. MRI provided comprehensive evaluation of the craniovertebral junction and demonstrated the extent of

neural compromise. Early recognition of these findings is essential to guide timely neurosurgical management and prevent irreversible neurological deterioration.

FIGURES:

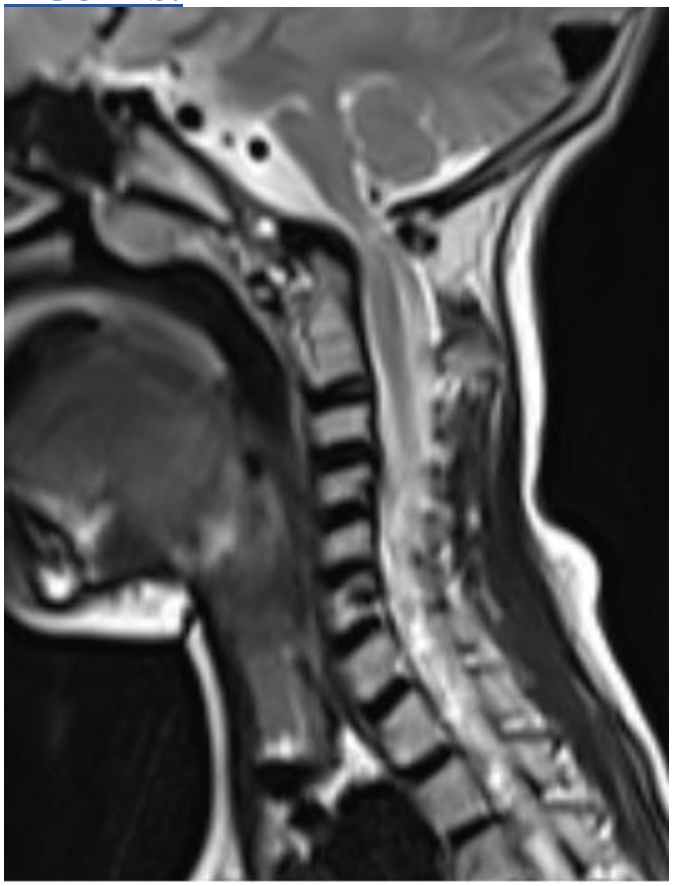

Figure 1: Sagittal T2-weighted MRI demonstrating atlantoaxial dislocation with widening of the atlantodental interval, and compression of the cervicomedullary junction with associated spinal cord T2 hyperintensity consistent with myelopathy.

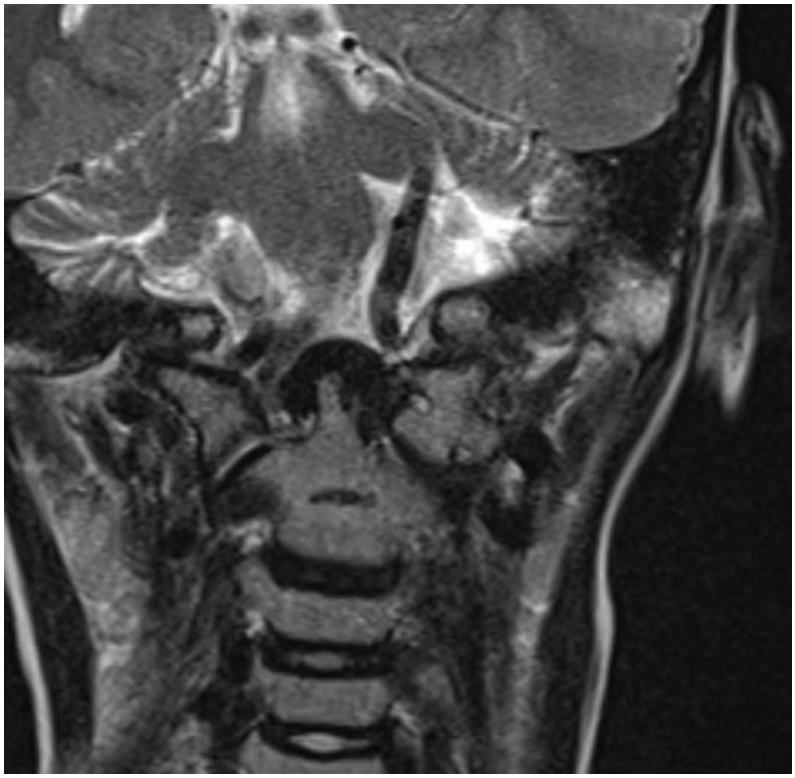

Figure 2: Coronal T2-weighted MRI showing hypertrophic peri-odontoid soft tissue.

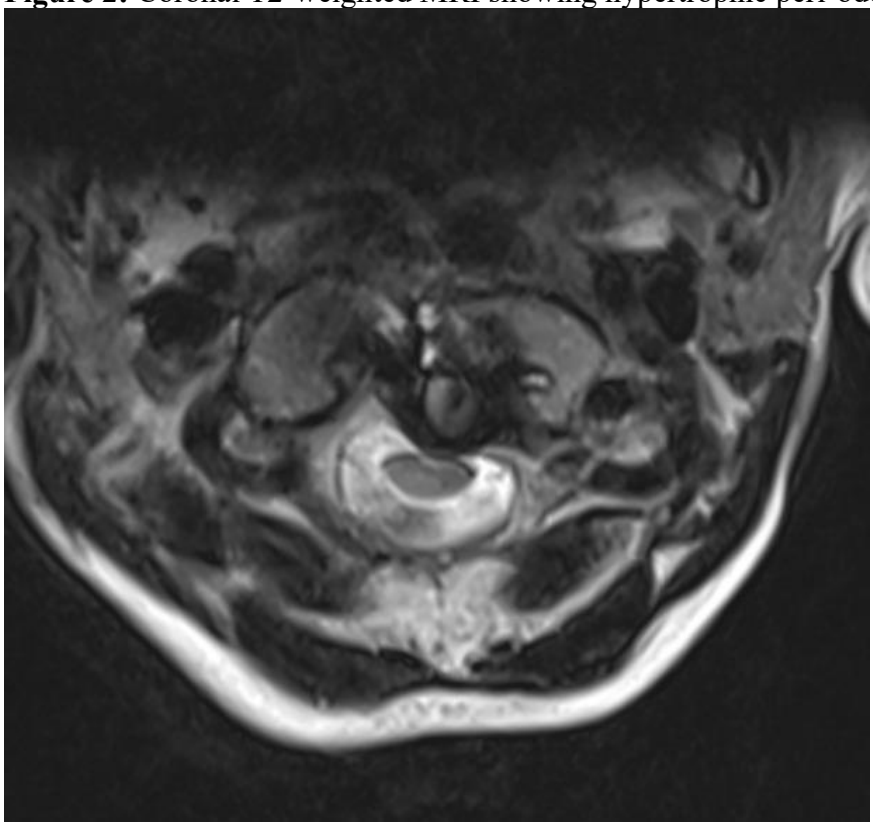

Figure 3: AxialT2-weighted MRI showing hypertrophic peri-odontoid soft tissue causing narrowing of the foramen magnum.

ACKNOWLEDGEMENTS

The authors would like to acknowledge the neuroradiology and pediatric teams involved in the diagnostic evaluation of this patient

REFERENCES

[1] Joaquim AF, Patel AA. Craniovertebral junction abnormalities in children: current concepts and management. Neurosurg Rev. 2023;46(1):56.

[2] Goel A. Atlantoaxial instability: contemporary concepts in diagnosis and treatment. J Craniovertebr Junction Spine. 2022;13(4):245-252.

[3] Martin MD, Bruner HJ, Maiman DJ. MRI evaluation of cervical spinal cord compression and pediatric myelopathy. Neuroimaging Clin N Am. 2022;32(3):401-417.

[4] Brockmeyer DL. Congenital abnormalities of the craniovertebral junction in children. Childs Nerv Syst. 2021;37(7):2089-2101.

[5] Menezes AH. Craniovertebral junction anomalies: diagnosis and management in pediatric patients. World Neurosurg. 2024;182:122-131.