Ventral Brain Stem Compression Due to C1-C2 Dislocation with Odontoid Retroversion in the Absence of Chiari Malformation a Case Report

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Disclosure

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Introduction

- The odontoid retroversion is a bony malformation consisting of the backward tilt of the odontoid process, that can produces displacing and compressing of the adjacent nerve structures (ventral brain stem compression).

- The embryogenesis of this malformation can be traced to the abnormalities of the development of the axial anatomical division of the craniovertebral junction (axial sclerotomes).

- Ventral brain stem compression due to craniovertebral junction abnormalities in the pediatric population is usually reported in association with Chiari Malformation.

- The mean age of diagnosis in the pediatric population is 9 years old with the youngest patient reported being 18 months old.

- The clinical manifestations can range from a mild occipital headache to cranial neuropathies and motor disturbances.
Case Presentation

• An 11 months old female presented in the emergency room with spinal shock, consisting of respiratory failure, spasticity, inexhaustible clonus and hyperreflexia.

• The patient had a history of non-specified neuroinfection at the 10th day after birth that was managed during a 30 days hospitalization, after discharge the patient presented the next clinical manifestations before arriving to our service:
  
  • Delayed in development milestones
  • Left upper body weakness at 7 months old, treated with physical therapy
  • At 10 months old the patient presented left hemiplegia
Pre-surgical imaging.

Figure 1.
Sagittal T2 MRI showing cord compression and associated inflammatory changes.

Figure 2.
Sagittal CT showing C1-C2 dislocation and narrowness of the spinal canal.
Pre-surgical imaging.

Figure 3. CT 3D reconstruction showing C3-C4-C5 pseudofusion with facet sclerosis.
Treatment.
The patient was treated surgically doing a posterior approach and an occipitocervical fusion with placement of hybrid instrumentation.

Figure 4.
Post-op CT 3D reconstruction showing the hybrid instrumentation.

Figure 5.
Post-op Sagittal CT showing a partial reduction of C1-C2 dislocation and narrowness of the spinal canal.
Post operative evaluation.

- After surgical treatment the patient showed ventilatory improvement and hemodynamic stability.

- There was no complication during the procedure and immediate hospitalization.

- The discharge was conducted 4 weeks after surgical approach.

- 10 weeks after treatment the patient presented motor improvement of the left hemibody.

- Currently the patient does not need ventilatory assistance and has little limitation to basic motor activities.
Discussion

• The CVJ malformation in this patient was accompanied with pseudoartrosis of the C3, C4 and C5 vertebral facets, and also fusion of C7 and T9 that can be tracked to alterations in the embryological development.

• This bone malformations were treated separately by doing an occipitocervical fusion and hybrid instrumentation for C3, C4 and C5 vertebral facets.

• The surgical treatment of choice for VBC is the ventral approach to de CVJ to attempt removal of C1 arch and odontoidectomy but in this case was not considered due to the size of the patient´s oral an nasal cavities.
Conclusion

- Brainsteem compression is a rare entity in the pediatric population and is usually described in association with Chiari Malformation.

- Delays in motor development milestones should be evaluated with an integral approach even in the presence of history of neurological pathologies specially with the worsening of the symptoms.

- Bony malformations have to be considered in pediatric population with clinical manifestations of brainsteem compression.