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Case Report

Hypoplasia of C1's posterior arch: Is there an ideal anatomical classification?

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Received : 15 August 2021
Accepted : 10 November 2021
Published : 20 December 2021

DOI
10.25259/SNI_820_2021

Quick Response Code:

ABSTRACT

Background: Congenital anomalies of the atlas are rare and usually occur in conjunction with other congenital variants. They include a wide spectrum of anomalies ranging from clefts to hypoplasia or aplasia of its arches that may contribute to spinal cord compressive syndrome.

Case Description: A 54-year-old male presented with the sudden onset of a severe quadriparesis and loss of proprioception after a minor fall. The magnetic resonance (MR) scan showed cord compression at the C1 level attributed to C1 arch hypoplasia. Two months following a decompressive C1 laminectomy without fusion, and the patient was symptom free.

Conclusion: Posterior C1 arch hypoplasia is a rare anomaly that can contribute to cervical cord compression and myelopathy. The optimal surgical management may include, as in this case, a posterior decompression without fusion.

Keywords: Atlas hypoplasia, C1 hypoplasia, Cervical stenosis, Craniocervical junction, Spine surgery

INTRODUCTION

Congenital anomalies of the posterior arch of the atlas are rare (i.e., 0.69–4%) and may vary from clefts to hypoplasia or aplasia [Table 1]. Atlas defects may be associated with the following additional congenital anomalies: Arnold-Chiari malformations, gonadal dysgenesis, Klippel-Feil syndrome, and Turner and Down syndromes.[1,8,10]

When present, posterior arch anomalies typically do not alter the biomechanical stability of the craniocervical junction and are mostly asymptomatic (i.e., incidentally detected on imaging). Nevertheless, these must be considered among the differential diagnoses when, following mild cervical trauma, patients acutely present with cervical pain and/or myelopathy.[2,8]

Currarino five types of posterior C1 arch anomalies

Currarino *et al.* described five malformations of the posterior arch of C1 (i.e., from A to E) [Table 1].[2] This classification is divided in 4 categories, that include: (A) Failure in the fusion of hemi-arches; (B) Unilateral cleft; (C) Bilateral cleft; (D) Complete absence of the posterior arch with persistent isolated tubercle; (E) Complete absence of posterior arch, including the tubercle. Here, we describe a

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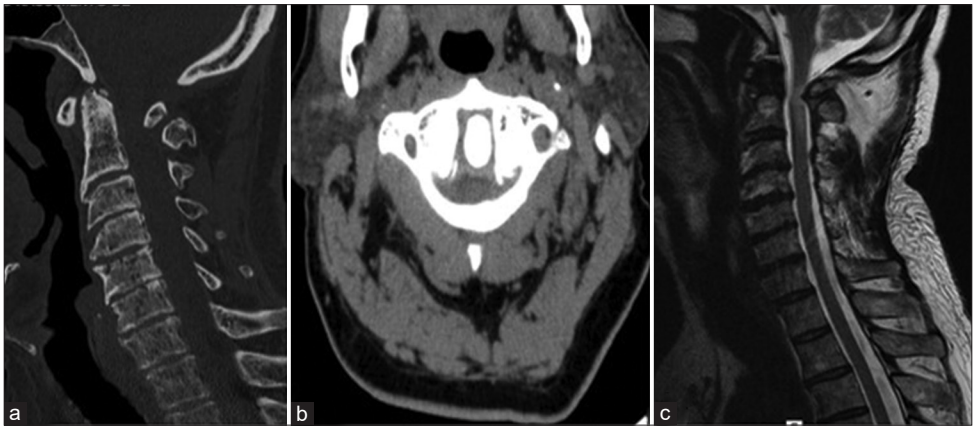

Figure 1: CT scan (image a and b) reveals stenosis of the canal at the level of the first cervical vertebra, with no fissures along the entire extension of the posterior arch of CMRI (image c) reveals compression in the cervical segment of the spinal cord.

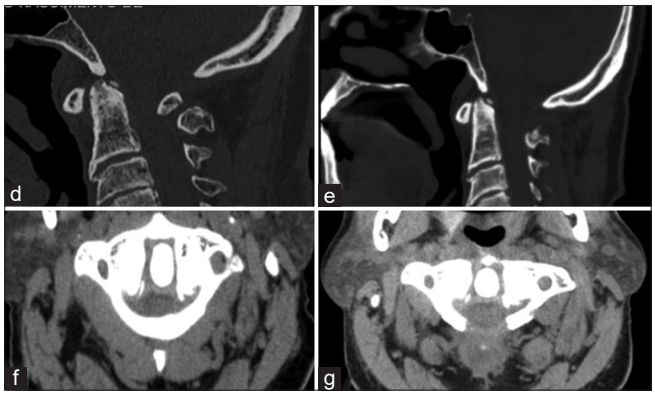

Figure 2: Comparative images of the C1 posterior arch hypoplasia (d and e) and after its surgical laminectomy (f and g).

Table 1: Description of Currarino classification of C1's congenital hypoplasia.

Types A-E	Description
A	Hyperossification of the fourth tubercle with premature complete fusion of hemi-arches.*
B	Failure in the fusion of hemi-arches
C	Unilateral cleft
D	Bilateral cleft
E	Complete absence of the posterior arch with persistent isolated tubercle
F	Complete absence of posterior arch, including the tubercle

*Type A: New type of C1's hypoplasia proposed

rare case of hypoplasia of the posterior arch of C1, emphasizing the anomalous anatomy and embryological etiology of these lesions also found in the literature.

CASE REPORT

A 54-year-old male presented with a severe quadriparesis and loss of proprioception in the lower extremities following a minor fall.

Diagnostic studies

Dynamic X-rays of the cervical spine showed no instability, but magnetic resonance (MR) and computed tomography (CT) scans demonstrated C1 posterior arch hypoplasia without fissures or clefts and significant dorsal cord compression [Figure 1].

Surgery

The patient underwent a minimally invasive C1 midline laminectomy (i.e., resection of the C1 posterior arch) without a fusion (i.e., facet joints and soft tissues preserved) [Figure 2]. The patient was discharged 2 days later, neurologically intact; there were no complications. Over the next 3 years, he remained symptom free and did not develop radiological signs of cervical instability.

DISCUSSION

Cervical myelopathy is usually attributed to subaxial degenerative disease. Rarely, cervical canal stenosis may be attributed to hypoplasia of the atlas resulting in spinal cord compression/myelopathy.

Anatomy of C1

The body of the atlas is derived from three ossification centers, which extend to and fuse dorsally to form the posterior arch.[8] Currarino five categories are based on defects of these centers of ossification [Table 2].[1,4-7,9] A fourth defect involves the fourth hyperossification center in 2% of the population (i.e., responsible for the posterior tubercle) that warrants that a new category should be added to Currarino classification.[3]

Surgery

A C1 laminectomy without fusion, as performed minimally invasively in this case, is the typical treatment of choice. Notably, no fusion is warranted if the facet joints are preserved.

Table 2: Case reports of myelopathy due to hypoplasia of the atlas look at other tables this is too verbose-Cut-Edit-Shorte n.

Reference journal	Age/ sex	Preoperative deficit	Defect stenosis	Surgery	Outcome
Sawada *et al.*, Neuroradiology 1989	38/Male	Quad	Atlantal stenosis	Lam C1	Improvement of neurological deficits
Phan *et al.*, Neurosurgery 1998	80/Male	Bilateral hand paresthesia, leg stiffness, and urinary incontinence	Atlantal hypoplasia	Lam C1/C2	Improvement of neurological deficits
	75/Male	Quadriparesis and hypereflexia	Atlantal Hypoplasia	Lam C2	
Liliang, *et al.* Journal of neurosurgery 2000	3/Male	Quadriparesis and respiratory distress	Atlantal stenosis	Lam C1 + fusion occiput to C2	Neurological status gradually improved after 3 weeks
May *et al.*, Journal of neurosurgery 2001	66/Male	Upper limb numbness and gait difficulty	Atlantal stenosis	Lam C1	Gait Improvement, sustained pyramidalism
Hsu, *et al.*, J Chin Med assoc. 2007	38/Male	Upper limb, abdominal and perineal paresthesia	Atlantal Stenosis	Lam C1 + Duroplasty	Improvement of all Neurologic symptoms
Bhattacharjee *et al.*, J craniovertebr junction spine 2011	10/Male	Progressive Quadriparesis and Respiratory distress	Atlantal stenosis + syringomielia	Lam C1	Imediate mprovement in his respiratory distress gradually
The spasticity came down. Meng *et al.*, Medicine 2016	39/Male	Lower extremities paresis and intermittent urinary incontinence	Atlantal Hypoplasia + Ossification of atlantoaxial membrane	Lam C1/ C2	Preoperative symptoms were alleviated

Other factors contributing to need for surgery include stenosis, extensive cord compression, high intrinsic cord signals/edema/ myelomalacia on MR, abnormal sagittal alignment, ankylosis of the anterior spinal column, and motion on flexion/extension cervical films (i.e., dynamic instability).

CONCLUSION

Here, we propose an additional classification to Currarino five A-E classifications. This should be labeled "A" and would be defined as C1 arch hypoplasia with hyperossification of the fourth tubercle with premature complete fusion of hemi-arches.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Junior MG, dos Santos NA, Ribeiro RT, Landeiro JA, Pessoa BL. Hypoplasia of C1's posterior arch: Is there an ideal anatomical classification? Surg Neurol Int 2021;12:623.