Clinical Case Reports International

Case Report
Published: 01 Aug, 2023

C1-C2 Segmentation Defect: A Case Report

Meluzio MC[1], Borruto MI[1], Genitiempo M[1] and Tamburrelli FC[1,2]*

[1]Department of Aging, Neurological, Orthopedic and Head-Neck Sciences, Fondazione Policlinico Universitario Agostino Gemelli IRCCS, Italy

[2]Department of Geriatrics and Orthopedics, Catholic University of the Sacred Heart, Italy

Abstract

Background and Aim: C1-C2 segmentation defect has rarely been reported. Clinical onset occurs usually in the first years of life, but sometimes in the following. Treatment is still an argument of debate.

Case Report: This case report describes the diagnosis and surgical treatment of a cervical stenosis due to a C1-C2 deformity treated with a midline laminectomy without fusion.

Conclusion: There are no clear indications to what the treatment options might be, but at 1-month clinical follow-up, the patient had recovered walking autonomy and partially improved fine finger movements.

Keywords: C1-C2 segmentation defect; Cervical myelopathy; C1-C2 deformity; Cervical stenosis

Background

Cervical myelopathies can be associated with congenital anomalies or syndromic pictures as in S. of Arnold-Chiari, S. of Turner, Klippel-Feil syndrome and anomalies of segmentation of the vertebra. The clinical onset in some cases occurs in the first year of life in others in the following years [1,2]. The proven correlation between congenital anomalies and neurological symptoms is well described in the literature [3].

Sometimes the diagnosis of aplasia or congenital stenosis of the medullary canal goes unnoticed, and patients become symptomatic when a degenerative disorder is associated. Congenital anomalies of the posterior arch of the atlas are rare (0.69%-4%) and may vary from clefts to hypoplasia or aplasia. Currarino et al. produced an anatomical classification of malformations of the posterior arch of C1 (from A to E). This classification is divided in 4 categories [4]. We describe a case of C1-C2 deformity with a particular diagnostic-therapeutic procedure.

Case Presentation

An 82-year-old woman, presented with fatigue in the lower limbs and difficulties while walking, for about 2 years. Performs a first evaluation by lumbo-sacral spine MRI that does not show a picture of spinal canal stenosis. The symptoms worsened in about a year, with the onset of loss of balance, reduced walking autonomy and dysesthesia in the upper limbs, the appearance of clones in the lower limbs, worsening weakness in the lower limbs. She underwent MRI of the cervical spine showing C1-C2 stenosis. To complete the diagnostic process, she performs a CT scan of the cervical spine showing a segmentation defect at the C1-C2 level. The patient underwent a midline laminectomy without fusion: Resection of the C1 posterior arch, removing the posterior arch sketch, which is responsible for the compression effect. The sick woman was transferred to a rehabilitation ward after three days. At 1-month clinical follow-up, the patient had recovered walking autonomy and partially improved fine finger movements.

Discussion

Abnormalities in the formation of vertebrae during embryogenesis can ultimately result in hypoplasia or aplasia. According to Currarino's classification, the patient falls into the type C, unilateral cleft. Because she has a defect in the formation of the posterior part of the arch with preservation of the dorsal part (Table 1).

These patients often manifest their first complaints following trauma or in young adulthood [5-7].

OPEN ACCESS

Correspondence:
Maria Ilaria Borruto, Department of Aging, Neurological, Orthopedic and Head-Neck Sciences, Fondazione Policlinico Universitario Agostino Gemelli IRCCS, Rome, Italy

Received Date: *07 Jul 2023*
Accepted Date: *26 Jul 2023*
Published Date: *01 Aug 2023*

Citation:
Meluzio MC, Borruto MI, Genitiempo M, Tamburrelli FC. C1-C2 Segmentation Defect: A Case Report. Clin Case Rep Int. 2023; 7: 1576.

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

Types	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

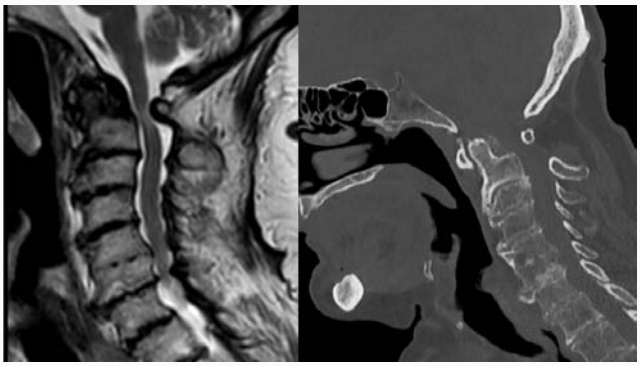

Figure 1: a) RM T2-w; B) CT scan.

Furthermore, on MRI, there were no signs of myelomalacia, edema or syringomyelia, as is the case in most of the cases described. 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).

Several authors agree that stenosis, myelomalacia and syringomyelia require surgical treatment such as isolated decompression [7,8]. Jong Kyu Kwon et al. confirms that many of the formation defects are an occasional finding during investigation of neck mass, neck pain, radiculopathy, and after trauma [9]. Posterior atlas arch defects are attributed to the defective or absent development of the cartilaginous preformation of the arch rather than a disturbance of the ossification [10].

This is supported by findings at autopsy or intraoperatively that connective tissue bridges the bony defect [11] (Figure 1).

References

1. Phan N, Marras C, Midha R, Rowed D. Cervical myelopathy caused by hypoplasia of the atlas: Two case reports and review of the literature. Neurosurgery. 1998;43(3):629-33.
2. Torriani M, Lourenco JL. Agenesis of the posterior arch of the atlas. Rev Hosp Clin Fac Med Sao Paulo. 2002;57(2):73-6.
3. Kumar R, Kalra SK, Vaid VK, Sahu RN, Mahapatra AK. Craniovertebral junction anomaly with atlas assimilation and reducible atlantoaxial dislocation: A rare constellation of bony abnormalities. Pediatr Neurosurg. 2008;44(5):402-5.
4. Currarino G, Rollins N, Diehl JT. Congenital defects of the posterior arch of the atlas: A report of seven cases including an affected mother and son. AJNR Am J Neuroradiol. 1994;15(2):249-54.
5. Ahn KW, Hong SK, Whang K, Pyen JS, Kim HJ, Han YP, et al. Transient quadriparesis due to dysgenesis of the posterior arch of the atlas: Case report. J Korean Neurosurg Soc. 1999;28(4):565-9.
6. Connor SE, Chandler C, Robinson S, Jarosz JM. Congenital midline cleft of the posterior arch of atlas: A rare cause of symptomatic cervical canal stenosis. Eur Radiol. 2001;11(9):1766-9.
7. Devi BI, Shenoy SN, Panigrahi MK, Chandramouli BA, Das BS, Jayakumar PN. Anomaly of arch of atlas--A rare cause of symptomatic canal stenosis in children. Pediatr Neurosurg. 1997;26(4):214-7; discussion 217-8.
8. Junior MGP, Dos Santos NADSQ, Ribeiro RT, Landeiro JA, Pessoa BL. Hypoplasia of C1's posterior arch: Is there an ideal anatomical classification? Surg Neurol Int. 2021;12:623.
9. Kwon JK, Kim MS, Lee GJ. The incidence and clinical implications of congenital defects of atlantal arch. J Korean Neurosurg Soc. 2009;46(6):522-7.
10. Geipel P. [Studies on the fissure formation of the atlas and epistropheus. IV]. Zentralbl Allg Pathol. 1955;94(1-2):19-84.
11. Richardson EG, Boone SC, Reid RL. Intermittent quadriparesis associated with a congenital anomaly of the posterior arch of the atlas. Case report. J Bone Joint Surg Am. 1975;57(6):853-4.