

Research Article

Total aplasia of posterior arch of the Atlas

U. Solomon Krupanidhi^{*1}, Jayaprakash B.R², Mallikarjun M.³ and Raveendra Patil⁴

¹Associate Professor, Department of Anatomy, Vijayanagar Institute of Medical Sciences, Bellary, Karnataka, India.

²Post graduate student, Department of Anatomy, Vijayanagar Institute of Medical Sciences, Bellary, Karnataka, India.

³Professor and Head of the department, Department of Anatomy, Vijayanagar Institute of Medical Sciences, Bellary, Karnataka, India.

⁴Assistant Professor, Department of Anatomy, Vijayanagar Institute of Medical Sciences, Bellary, Karnataka, India.

***Correspondence Info:**

Dr. U. Solomon Krupanidhi
Associate Professor,
Department of Anatomy,
Vijayanagar Institute of Medical Sciences, Bellary, Karnataka, India.
E-mail: krupanidhidrsolomon@gmail.com

Abstract

Congenital anomaly-total aplasia of the posterior arch of the atlas vertebra is very rare, a developmental failure of chondrogenesis rather than the disturbance of ossification. Reported prevalence is 0.95-4%. In the present study the incidence of total aplasia of posterior arch of the atlas is 0.91% i.e., 1 in 109 dried human adult atlas vertebrae. The atlas which does not possess the posterior arch comes under the type E category of congenital posterior arch defect classification proposed by currarino *et al.* Knowledge of congenital anomaly of atlas is important for clinicians as new surgical techniques and instruments continue to evolve for the treatment of unstable cervical spine and also to avoid misdiagnosis among trauma victims and to provide information on its proper management.

Keywords: Atlas, Posterior Arch, Aplasia, Anomaly, Congenital

1. Introduction

The atlas presents an anterior arch, a posterior arch and lateral masses. The superior surface of the posterior arch of the atlas having a groove for vertebral artery, venous plexus immediately behind and suboccipital nerve¹. The onset of ossification of the posterior arch of the atlas occurs during seventh week of intrauterine life proceeding perichondrally from two centers located in the lateral masses. Complete fusion of posterior arch is expected to occur between 3 and 5 yrs of age. About 2% of fetuses presents fourth ossification center during development leads to i) median clefts of posterior arch, ii) Varying degrees of posterior arch dysplasia^{2,3}. Defect of posterior arch of atlas is believed to occur due to failure of local chondrogenesis rather than disturbance of ossification⁴. This has been agreed with the autopsy or intra operative findings⁵.

We found an atlas with total aplasia of posterior arch which is a very rare anomaly even although well documented congenital anomaly. Knowledge of congenital anomaly of atlas is essential for clinicians while treating patients with unstable cervical spine as new surgical techniques and instruments continue to evolve⁶.

2. Materials and Methods

A total of 109 dried Human Adult Atlas vertebrae of either sex were carefully studied from the collection in the Department of Anatomy, Vijayanagar institute of medical sciences, Bellary, Karnataka, India. Among these 109 atlas vertebrae we found only one atlas with total aplasia of posterior arch.

Fig. Total aplasia of posterior arch

3. Discussion

So many theories have been proposed to explain congenital defects of the posterior arch of the atlas⁷. However, the exact mechanism remains obscure. Congenital aplasia of posterior arch of atlas may be associated with several disorders including Down's syndrome, Turner syndrome, Gonadal dysgenesis, Arnold chiari malformation, Klippel-Feil syndrome, leading to atlantoaxial instability^{2,3}. Hereditary factors may contribute to these anomalies, but the frequency is not known. Montatenau *et al*⁸ reported an affected mother & daughter and Currarino *et al*⁷ an affected mother & son, suggesting an autosomal dominant inheritance. The anomalies of upper cervical vertebrae occur more frequently in individuals with cleft lip, cleft palate and both⁹. Total or partial aplasia of posterior arch of atlas are rare². Plaut and Lawrence and Anderson first published an article about a developmental abnormality of atlas in 1937¹⁰. Giepel did not encounter any in 2,749 postmortem specimens of the atlas vertebrae¹¹. The true incidence is not known and only a few articles reported on this particular malformation^{7,8}.

In Senoglu's study, the overall incidence (CT scan +dried specimens +fresh specimens) of posterior atlantal arch in 1354 evaluated cases is 2.95% (40 cases)¹². The congenital defects of posterior arch of the atlas was first summarized by Von Torkulus and Gehle into 6 forms- A.Total aplasia, B.Aplasia with persistent posterior tubercle, C. Aplasia with paramedian unilateral posterior arch remnant, D. Aplasia with paramedian bilateral posterior arch remnant and rachischisis, E. Hemiplasia, F.Unilateral posterior arch partial aplasia¹³. Subsequently, Currarino *et al* modified and then developed a new classification system for congenital defects of posterior arch atlas, which is now widely used; according to it- Type A:Failure of posterior midline fusion with a small gap remaining, Type B:Unilateral cleft, Type C:Bilateral defects with preservation of the most dorsal part of the arch, Type D:Complete absence of the posterior arch with a persistent isolated tubercle. Type E:Complete absence (total aplasia) of the entire posterior arch. Type A most common type affecting 4% of general population, other type(B-E) are found to affect 0.69% of general population⁷.

In the present study the incidence of total aplasia of posterior arch of the atlas is 0.91% i.e., 1 in109 dried human adult atlas vertebrae. The present atlas belongs to type E category of congenital posterior arch defect classification proposed by currarino *et al*⁷. Patients with total aplasia of posterior arch of the atlas should be advised to avoid contact sports and seek medical treatment if they experience neurological symptoms.

4. Conclusion

Congenital anomalies of posterior arch of the atlas are very uncommon but well documented. Total aplasia of posterior arch of atlas are very rare. The incidence of total aplasia of posterior arch of the atlas in our study is 0.91% i.e., 1 in109 dried human adult atlas vertebrae. Knowledge of the congenital anomaly of the atlas is essential for clinicians mainly neurosurgeons and orthosurgeons while treating patients especially the trauma victims in order to avoid excessive investigations, diagnostic errors and providing useful approaches to further management¹⁴, for e.g., during posterior fusion of cervical vertebra in case of atlantooccipital instability¹⁵.

References

1. Johnson D, Ellis H, Collins P, Standring S, editor. Gray's anatomy :the anatomical basis of clinical practice,38th ed.90 Tottenham court road London W1P9HE:Elsevier Churchill Livingstone;2005.p.518-519.
2. Dalinka MK, Rosenbaum AE, Van Houten F. Congenital absence of posterior arch of atlas. *Radiology* 1972;103:581-583.
3. Torriani M, Lourenco JL. Agenesis of the Posterior arch of the atlas. *Rev Hosp clin Fac med Sao Paulo* 2002;57:73-76.
4. Logan WW, Stuard ID. Absent posterior arch of the atlas. *Am. J. Roentgenol. Radium. Ther. Nucl. Med.* 1973;118:431-434.
5. 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;57:853-54.
6. Senegul G, Kodiglu HH. Morphometric anatomy of atlas and axis vertebra. *Turkish Neurosurgery.* 2006; 16(2):69-76
7. Currarino G, Rollius N,Diehl J T: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;15:249-54.
8. Montateanu M, Gudinchet F, Sarraj H, Schnyder P.case report 665.Congenital absence of posterior arch of atlas. *Skeletal Radiol* 1991;20:231-232.
9. Ugar DA, Semb G. The prevalence of anomalies of the upper cervical vertebrae in subjects with cleft lip, cleft palate, or both. *Cleft palate Craniofac J* 2001;38:498-503.
10. Hakan SABUNCUOGLU, Selcuk OZDOGAN, Demet KARADAG, Erdener TIMURKAYANAK. Congenital Hypoplasia of the posterior arch of the atlas:case report and Extensive review of the literature. *Turkish Neurosurgery* 2011;21:97-103.
11. Geipel p. Zur Kenntnis der Spaltbildungen das Atlas und Epistropheus.Teil IV. *Zentralbl Allg pathol* 1955;94:19-84.
12. Senoglu M,Safavi-Abbasi S,Theodore N,Bambakidis NC,Crawford NR,Sonntag VK.The frequency and clinical significance of congenital defects of the posterior and anterior arch of the atlas. *J Neurosurg spine.*2007;7:399-402.
13. Von Torklus D,Gehle W. The upper cervical spine.New York:grune & Stratton;1972.
14. Gehweiler JA, Daffner RH, Roberts L. Malformations of the atlas vertebra simulating the Jefferson fracture. *AJR* 1983;140:1083-1086.
15. Hierholzer H, Isalberti M, Hosten N, Stroszczyński C, Gandini G, Felix R: A rare,complex developmental anomaly of the atlas:Embryological and radiological considerations. *Neuroradiology* 1991; 41:901-903