Total aplasia of posterior arch of the Atlas

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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 in109 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 nerve. The onset of ossification of the posterior arch of the atlas occurs during seventh week of intrauterine life proceeding perichondrically 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. Defect of posterior arch of atlas is believed to occur due to failure of local chondrogenesis rather than disturbance of ossification. This has been agreed with the autopsy or intra operative findings.

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 evolve.

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.
3. Discussion

So many theories have been proposed to explain congenital defects of the posterior arch of the atlas\textsuperscript{7}. 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\textsuperscript{2,3}. Hereditary factors may contribute to these anomalies, but the frequency is not known. Montatenau et al\textsuperscript{8} reported an affected mother & daughter and Currarino et al\textsuperscript{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 both\textsuperscript{9}. Total or partial aplasia of posterior arch of atlas are rare\textsuperscript{2}. Plaut and Lawrence and Anderson first published an article about a developmental abnormality of atlas in 1937\textsuperscript{10}. Giepel did not encounter any in 2,749 postmortem specimens of the atlas vertebrae\textsuperscript{11}. The true incidence is not known and only a few articles reported on this particular malformation\textsuperscript{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)\textsuperscript{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. Hemiaplasia, F.Unilateral posterior arch partial aplasia\textsuperscript{13}. 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 population\textsuperscript{7}.

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\textsuperscript{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 management\textsuperscript{14}, for e.g., during posterior fusion of cervical vertebra in case of atlantoccipital instability\textsuperscript{15}.
References


