

Research Article

Congenital posterior arch defects of the Atlas

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Abstract

Congenital anomalies of the posterior arch of the atlas vertebrae are very rare ranging from partial clefts to total aplasia. The congenital defects of the posterior arch of the atlas have been classified by currarino *et al* into five types (A to E), with type A being most common affecting 4% of general population and type B-E being found only in 0.69% of the general population. In the present study the incidence of congenital posterior arch defect type A is 2.75% and type B-E is 1.83%(type B;0.91% , type E; 0.91%). The clefts of the atlas may be mistaken for fracture in patients who have sustained cervical spine injuries. The clinical presentation of the patients with congenital defect of the posterior arch of atlas can be variable from asymptomatic to symptomatic, it is worthy to recognize and classify the exact type to prevent further neurological complications, diagnostic error and for proper management.

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

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. Malformations of the atlas are relatively rare and exhibit a wide range including aplasia, hypoplasia and various arch clefts^{2,3}. In seventh week of gestation, the lateral centers extend dorsally to form the posterior arch. Ossification of posterior arch usually proceeds perichondrally from two centers in the lateral masses, towards the midline and fusion occurs between 3 and 5 yrs of age. In rare cases, a separate ossification center forms in the midline and fuses secondarily with the lateral masses forming the posterior arch-a posterior midline ossified tubercle develops if this center fails to fuse with the hemiarches. At least two anomalies can develop during the ossification process - i) median clefts of posterior arch, ii) Varying degrees of posterior arch dysplasia^{4,5,6}. Defects of posterior arches are thought to occur due to failure of local chondrogenesis rather than subsequent of ossification^{7,8}.

We found the five atlas vertebrae which are having defects in posterior arch ranging from partial clefts to total agenesis-rare anomaly. Knowledge of congenital anomalies of atlas is essential for clinicians while treating patients with unstable cervical spine as new surgical techniques and instruments continue to evolve⁹. The clinical presentation of the patients with congenital defect of the posterior arch of atlas can be variable from asymptomatic to symptomatic, it is worthy to recognize and classify the exact type for better management¹⁰.

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 five atlas vertebrae with posterior arch defect ranging from partial cleft to total aplasia.

Fig.1- 109 atlas vertebrae

Fig.2- Congenital posterior arch defects of the Atlas; according to Currarino's classification showing Type A, Type B, and Type E¹⁰

3. Result and Discussion

Congenital defects of the posterior arch of the atlas ranging from partial to total aplasia are rare^{11,12}. Congenital defects of the posterior arch of the 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^{4,5}. Although the familial incidence is uncertain a report indicates an autosomal dominant inheritance describing the same disorder in an affected mother & daughter¹³ and an affected mother & son¹⁰. Plaut and Lawrence and Anderson first published an article about a developmental abnormality of atlas in 1937¹⁴. Giepel did not encounter any in 2,749 postmortem specimens of the atlas vertebrae. The true incidence is not known and only a few articles reported on this particular malformation. 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 aplasia¹⁵.

In 1990 Villas *et al* classified congenital malformations of the atlas into – Area 1 : Defect of formation of the posterior arch, Area 2 : Defect in union of the posterior arch with the superior articular facet, Area 3 : Defect of formation with hypoplasia or agenesis of the superior articular facet, Area 4 : Defect of formation of the anterior arch, Area 5 : Malformation of the atlanto-occipital junction, Area 6 : Malformation of the atlanto-axial junction¹⁶. 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 population¹⁰. In the present study the incidence of congenital posterior arch defect type A is 2.75% and type B-E is 1.83% (type B; 0.91% , type E; 0.91%). i.e., In 109 dried human adult atlas vertebrae. The posterior arch defects of the atlas are considered benign anatomical variations. The congenital defect being found incidentally, patients may present with asymptomatic to various symptoms including chronic neck pain, headache, Lhermitte's sign, transient quadriplegia, atlantoaxial instability^{11,12}.

When no neurological deficit found usually type A and type B, treatment is normally conservative. Patients with symptomatic usually suffer from type C/type D defects, with isolated posterior bony remnant may traumatize the dorsal spinal cord during extension of cervical spine and cause transient quadriplegia after minor trauma even inappropriate positioning of neck and head. Hence surgery is indicated when there is atlanto axial instability and/or spinal cord compromise¹⁷. Thus the type of the defect on the posterior arch of the atlas determination is essential to prevent further neurological complications^{17,18}. Patients with this anomaly especially in presence of type C/type D should be advised to avoid contact sports and seek medical treatment if they experience neurological symptoms¹⁸.

4. Conclusion

Congenital anomalies of the posterior arch of the atlas vertebrae are very rare ranging from partial clefts to total aplasia¹⁹. Congenital anomalies of posterior arch of the atlas once found, detailed evaluation of the patient should be done to avoid misdiagnosis as instability, fracture or osteolysis. Knowledge of the congenital anomalies 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 to provide proper management²⁰. According to Currarino *et al.* congenital defects of the posterior arch of the atlas are of five types (A to E), with type A being most common affecting 4% of general population and type B-E being found only in 0.69% of the general population. In the present study the incidence of congenital posterior arch defect type A is 2.75% and type B-E is 1.83%(type B;0.91% , type E; 0.91%). Because of the neurological presentation is associated with type of the defect, it is worthy to recognize and classify the exact type to prevent further neurological complications¹⁸.

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