

Congenital defects of posterior arch of the atlas: a case report

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ABSTRACT

Our aim was to describe an unusual congenital anomaly of atlas and to emphasize its clinical significance.

A 43-year old woman presented with occipital headache that was worse with hyperextension of neck. Patient denied any neck pain and other neurological complain.

On examination, the only positive finding on physical examination was moderate pain at the upper cervical and occipital region throughout neck extension. The cranial nerve examination was unremarkable, and motor and sensory findings were normal. Plain cervical radiographs revealed bilateral defects in the posterior arch of atlas. Computed tomography scans in flexion and extension positions showed no displacement or inward mobility of the posterior tubercle during extension of the cervical spine. No evidence of compression of the spinal cord was detected on magnetic resonance imaging.

Congenital anomalies of the posterior arch of the atlas are very uncommon and usually asymptomatic. Neurological presentations depend on type of defect of posterior arch of atlas. It is crucial to recognize this defect to prevent major neurological deficits. *Neuroanatomy; 2007; 6: 72–74.*

Key words [atlas] [posterior arch] [congenital defect]

Introduction

Congenital anomalies of the posterior arch of the atlas (first cervical vertebrae) are very uncommon but well documented. This condition is usually asymptomatic and diagnosed incidentally. Nevertheless some patients present with some degree of transient or chronic neurological symptoms following minor neck or head trauma [1,2].

We present a case with congenital defect of posterior arch of atlas and review previous reports in literature.

Case Report

A 43-year old woman presented with occipital headache of 8 months' duration. She complained that the pain was worse with hyperextension of her neck. However, she denied any neck pain or other neurological complain.

On the physical examination, the only positive finding was moderate upper cervical and occipital pain throughout neck extension. Cranial nerve examination was unremarkable, and motor and sensory examinations of both upper and lower limbs were normal. Lhermitte sign and Romberg test were also negative. Reflexes were normal bilaterally, and her toes were down going.

Plain lateral radiographs (Figure 1) and computed tomography (CT) (Figure 2) of neck revealed bilateral defects in the posterior arch of atlas. Plain radiography (Figures 3A and 3B) and CT scans in flexion and extension showed no displacement and no inward mobility of the posterior tubercle during extension of the

neck. No evidence of compression of the spinal cord was shown on magnetic resonance imaging (MRI) (Figure 4). Surgical intervention was not advised because of lack of neurological symptoms and spinal cord compression.

Discussion

Normally atlas has three primary ossification centres during the embryonic period. The anterior tubercle formed from the anterior ossification centre, and two lateral centres form to lateral masses and the posterior arch. Lateral masses unite posteriorly, giving rise to the posterior arch at 3-5 years of age. Anterior arch usually unites with two lateral centres at 5-9 years of ages [3-5]. Defect of posterior arch of atlas is believed to occur due to failure of local chondrogenesis rather than ossification [4]. This has been agreed with the autopsy or intra-operative findings [6]. Currarino et al [2] classified this anomaly into five types, depending on the extent of absence of posterior arch and presence of posterior tubercle (Table 1). *Type A* anomaly is seen over the 90% of all posterior arch defects and present in 3% to 4% of individuals [2]. In their study, they estimated the *Type B-E* anomalies as 0.69% of all population. Currarino et al [2] also divided the posterior arch defects of atlas into five clinical groups: *Group I* - asymptomatic, their anomaly is found incidentally; *Group II* - neck pain or/and stiffness after trauma to neck or head; *Group III* - sudden neurological symptoms after neck or head trauma; *Group IV* - various neurological symptoms for



Figure 1. Lateral radiograph revealing aplasia of posterior arch of atlas with posterior bony fragment.

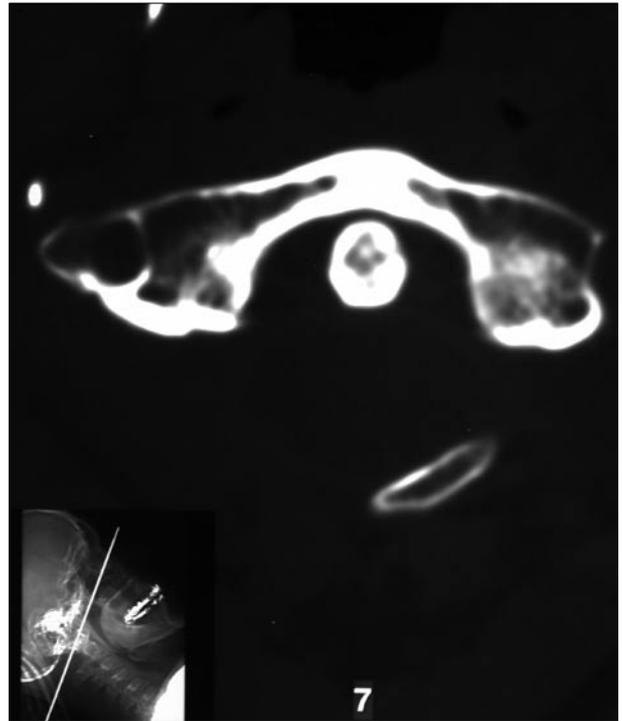


Figure 2. Axial CT at the level of C1 showing absent posterior arch of atlas.



Figure 3. Lateral radiographs of cervical spine taken in flexion (**A**) and extension (**B**) showing no displacement and no inward mobility of posterior tubercle during extension.

some time before the diagnosis of the anomaly; *Group V* - chronic symptoms referable to the neck.

The clinical presentation of congenital aplasia of the posterior arch of atlas can be variable. According to Currarino's review, almost one third of these patients are asymptomatic. However, the neurological presentation in this anomaly described clearly in a few articles

in literature, including sensory symptoms such as paresthesia in all four limbs [6], in both upper limbs only [7], ipsilateral upper and lower limbs [8] or motor deficits such as episodic weakness of all four limbs [7].

The presence of a posterior tubercle is important for determination of clinical aspect. The posterior tubercle can cause transient quadriplegia by impinging on the



Figure 4. Midsagittal T1-weighted MRI revealing no compression of the spinal cord.

spinal cord during neck extension or following minor trauma to neck or head [3,7].

The most important reasons to present this case was that it was *Type C* according to Currarino's classification; neurological symptoms such as numbness or weakness at any limbs or transient quadriplegia did not develop but patient complained headache which is an uncommon complaint for this anomaly. Patient denied she has never developed neurological symptoms even after trauma or inappropriate positioning. We examined her meticulously

Table 1. Classification of congenital anomalies of the posterior arch of the atlas according to Currarino [2].

Type	Definition
A	Failure of posterior midline fusion of the two hemi-arches
B	Unilateral cleft
C	Bilateral cleft
D	Absence of the posterior arch with persistent posterior tubercle
E	Absence of the entire arch including posterior tubercle

in neck extension but could not find any neurological sign.

Sharma et al [7] were first to document the inward mobility of posterior fragment during extension of neck. Their findings supported the hypothesis that an isolated posterior bony fragment is the potential cause of neurological morbidity. The cumulative effects of trauma may cause myelopathy, additional trauma or inappropriate posture related to neck may cause compression on cervical part of spinal cord too.

It is important to determine the type of the defect on the posterior arch of atlas to understand the clinical significance and to prevent further neurological complication. *Type A* and *B* do not cause neurological symptoms but *Type C-D* are likely to cause transient quadriplegia after minor trauma even inappropriate positioning of neck and head. The other considerable matter is that a patient with this anomaly should be warned about avoiding contact sports and other strenuous athletic sports, especially in the presence of *Type C-D* anomaly and experience of neurological symptoms.

Congenital defects of posterior arch of atlas are rare and most of them are found incidentally. This anomaly is thought to develop due to a failure of chondrogenesis. These defects were classified into five types, depending on the presence of posterior tubercle. Because of the neurological presentation is associated with the type of the defect, it is worthy to recognize and classify the exact type.

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