

Aplasia of the Posterior Arch of the Atlas : A Proposal for a New Clinical Classification

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Abstract: Partial or complete aplasia of the posterior arch of the atlas is often detected incidentally when a patient has cervical X-rays. Patients with this anomaly are generally asymptomatic. However, some cases demonstrate neurological symptoms. It is important to predict cases of this anomaly that may be accompanied with neurological symptoms. Between April 2001 and March 2005, three patients were encountered who had an anomalous posterior arch of the atlas. The anomalies encountered in the three patients were midline cleft (two patients) and an absence of the posterior arch with persistent posterior tubercle (one patient). Based on our clinical experience and a review of the literature, we developed a new clinical classification system for this condition. From a clinical point of view, aplasia of the posterior arch of the atlas can be classified into three types. Type I is complete aplasia. Type II is partial aplasia with the isolated posterior fragment. Type III is divided into subtypes depending on the inward mobility of the isolated posterior fragment (Type III-a without and Type III-b with mobility). Type IV is partial aplasia without the isolated posterior fragment. Our new classification is simple and unambiguous, and it also provides a clear direction for developing appropriate diagnostic and treatment strategies.

Key words : Cervical spine, Atlas, Arch, Anomaly, Classification

Introduction

Patients with either partial or complete aplasia of the posterior arch of the atlas are generally asymptomatic. This anomaly is often detected incidentally when a patient has cervical X-rays. However, some cases with this anomaly demonstrate neurological symptoms.^{1)~4)} It is important to predict the cases demonstrating this anomaly which may be accompanied by neurological symptoms. Based on both our clinical experience and a review of the literature, we developed a new classification system of this condition for clinical use.

Materials and Methods

Between April 2001 and March 2005, 11,512 new patients were registered at the Department of Orthopaedic Surgery at Chikushi Hospital, Fukuoka University. The cervical X-rays examinations were performed for 4187 patients. Of these, three patients (0.071%) were encountered who had an anomalous posterior arch of the atlas. The diagnosis was made based on the findings of lateral cervical X-rays. Functional lateral cervical X-rays were taken in all patients. To demonstrate the anomaly in more detail, three-dimensional (3D) computed tomography (CT) and magnetic resonance imaging (MRI) of the cervical spine were performed. A neurological examination was per-

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formed at the initial and the last follow – up visit.

Results

The subjects included one male and two females with an average age of 30.7 years (range, 20–52 years). The cervical X – rays were obtained because of nonspecific neck pain or headache, tiredness in the upper extremities (Table 1). The anomalies encountered in the three patients were midline cleft (two patients) and an absence of the posterior arch with persistent posterior tubercle (one patient) (Table 1). No atlantoaxial instability or anterior displacement of the bony fragment during extension of the cervical spine was found on functional lateral cervical X – rays in all patients. An isolated posterior tubercle and a precise position of the defect were confirmed by CT and 3D–CT

(Figures 1, 2, Table 2) In all patients, MR images did not show a narrowing of the spinal canal nor any signal alternation of the spinal cord on T2–weighted images at the level from C1 to C7. In all patients, a neurological examination, including the mental status, gait and posture, cranial nerve reflexes, postural reactions, spinal reflexes and pain perception, was normal at the initial and the last follow – up visit. The symptoms of the patients subsided quickly after conservative treatments (Table 1).

Discussion

Partial or complete aplasia of the posterior arch of the atlas is an uncommon anomaly. In a study of 1,613 dissections by Geipel, a cleft of the posterior arch of the atlas was found in 4% of all

Table 1. Clinical features in the three patients

Case	Age	Sex	Symptom/ Reason for neck X – rays	Diagnosis	Treatment
1	20	F	Headache, neck pain, tiredness in the right upper extremity during work as security guard at an airport	Tension headache	NSAIDs, physical therapy, relaxation exercises, meditation, use a different pillow
2	20	F	Recurring pain on the outside of the right elbow during work as an office worker	Lateral epicondylitis of the humerus	NSAIDs, elbow orthosis, stretching and strengthening the forearm muscles
3	52	M	Limited ability to flex the head to the right side and to rotate the head	Cervical spondylosis	NSAIDs, physical therapy, cortisone injections, intermittent neck traction

NSAIDs : nonsteroidal anti – inflammatory drugs

Table 2. Radiological findings in the three patients

Case	Plain X – ray	Atlantoaxial instability	Inward mobility of the posterior fragment	CT	MRI
1	Absence of the posterior arch of the atlas except for the posterior tubercle (Fig. 1a)	(-)	(-)	Partial aplasia of the posterior arch of the atlas with an isolated posterior tubercle (Fig. 1b)	spinal cord compression (-) signal alteration (-)
2	ADI:2 mm, SAC:12 mm S/O congenital stenosis at the C1 level(Fig. 2a)	(-)	N/A	Posterior arch of the atlas became gradually thin Spinal canal remains wide at the C1 level (Fig. 2b)	spinal cord compression (-) signal alteration (-)
3	Double bulbous ends of the hemiarches Absence of a spino – laminar line (Fig. 3)	(-)	N/A	A bony defect is present in the midline posteriorly	spinal cord compression (-) signal alteration (-)

ADI : atlantodental interval SAC : space available for the cord S/O : suspected of N/A : not applicable

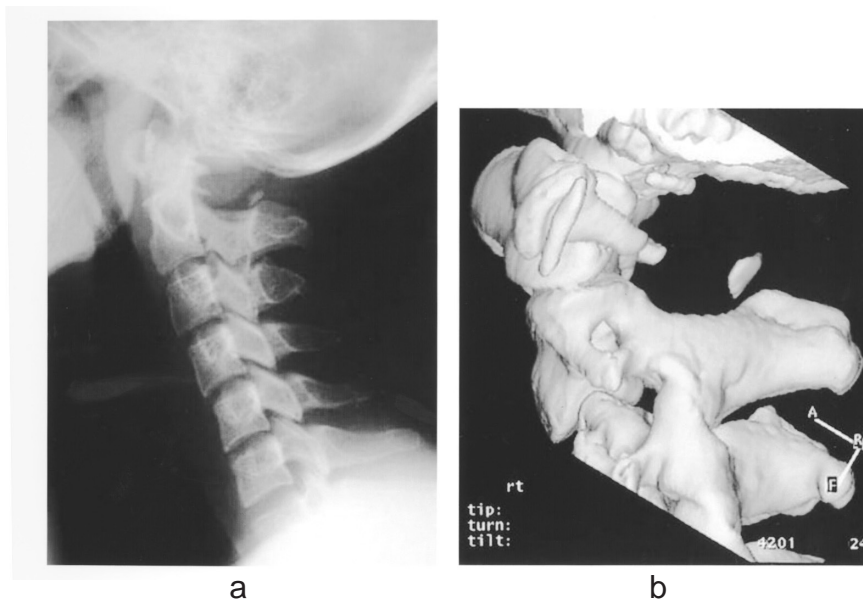


Figure 1. a Case 1 : A lateral radiograph of the cervical spine, showing the absence of the posterior arch of the atlas except for the posterior tubercle.
 b Case 1 : A lateral view of a three-dimensional computed tomography of the atlas, showing aplasia of the posterior arch of the atlas with an isolated posterior tubercle.

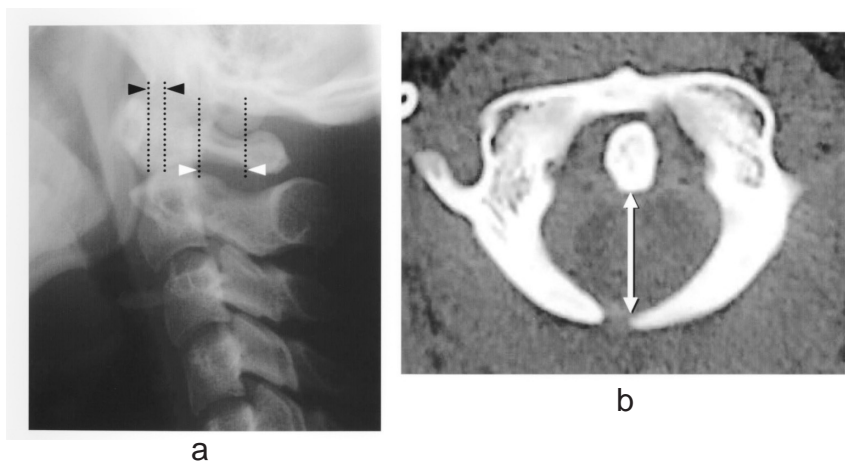


Figure 2. a Case 2 : A lateral radiograph of the cervical spine, showing hypoplasia of the atlas ; the anterior atlantodens interval was 2 mm (black arrowheads), whereas the space available for the cord was 12 mm (white arrowheads).
 b Case 2 : An axial view of computed tomography at the C1 level. The posterior arch of the atlas had gradually become thin ; as a result, the space available for the cord remained wide (white arrow).

cases.⁵⁾ It appears to be more common in women and an autosomal dominant inheritance has been suggested.⁶⁾ Autopsies and surgical explorations have shown that the bony gap in the posterior arch tends to be bridged by loose^{3,7)} or dense⁵⁾ connective tissue rather than cartilage. Furthermore, based on the shape and developmental pattern

of the structures of the cartilaginous human craniovertebral junction, some authors have suggested that certain pathologies most likely originated during the chondrification phase of development.⁸⁾⁻¹⁰⁾ These observations support the notion that these anomalies result from a defective development of the cartilage rather than a distur-



Figure 3. Case 3 : A posterior median cleft of the arch of the atlas could be recognized in the double bulbous ends of the hemi-arches and the absence of a spino-laminar line in a lateral view of X-rays which indicates the absence of the posterior wall of the spinal canal. A posterior cleft also is shown particularly clear on an oblique view of X-rays.

bance of ossification by itself.

The diagnosis of this anomaly can be made either incidentally on plain films in cases of trauma to the neck,¹⁾²⁾¹¹⁾ nonspecific neck or shoulder pain,¹⁾²⁾⁴⁾¹²⁾¹³⁾ an evaluation of the tonsil size or epiglottitis,¹⁾ and in an examination of edema in the upper extremities.¹³⁾ In some patients with partial aplasia of the posterior arch of the atlas with an isolated posterior tubercle, the isolated posterior fragment moves anteriorly into the spinal canal during extension of the cervical spine.⁴⁾¹²⁾¹⁴⁾ This inward mobility of the isolated posterior fragment may compress the cord during the extension of the cervical spine. Therefore, the clinical manifestations, neck pain or neurological deficits, may be caused by this malformation.¹⁾⁻⁴⁾¹¹⁾⁻¹⁵⁾ The Torklus-Gehle classification¹⁶⁾ and the Currarino classification¹ for aplasia of the posterior arch of the atlas have all been accepted

worldwide. The existence of an isolated posterior tubercle was noted in these previous classifications based on the morphological criteria, but its clinical significance was not described. These morphological classifications were not suitable for evaluating the clinical features because of its complexity and a lack of information about the movement of an isolated posterior tubercle. From a clinical point of view, the existence of an isolated posterior tubercle and its movement during cervical motion thus seem to be most important factors for predicting the development of neurological symptoms. Therefore, a new classification should be based not only on the morphological findings but also on functional factors.

Our classification for aplasia of the posterior arch of the atlas can be achieved by X-rays and CT scans (Figure 4). Functional lateral cervical X-rays provide better information about atlantoaxial

instability and/or inward mobility of the isolated posterior fragment. Aplasia of the posterior arch of the atlas can be classified into three basic types; Type I to III, with Type II having two subtypes. Type I is complete aplasia. Type II is partial aplasia with the isolated posterior fragment. Type III is divided into two subtypes depending on the inward mobility of the isolated posterior fragment (Type II-a without and Type II-b with mobility). Type III is partial aplasia without the isolated posterior fragment. According to our new classification, case 1 corresponded to Type II-a, while cases 2 and 3 corresponded to Type III. If atlantoaxial instability or inward mobility of the isolated posterior fragment (Type II-b) is suspected based on functional lateral cervical X-rays, then MRI is required. MRI is also required to evaluate the spinal cord in Type III, because the filamentous fibrous band replacing the absent bony arch may act on the unprotected cord in a manner similar to that of a guillotine in traumatized cases. Weisz reported a case of trauma to the cervical spine with partial aplasia of the posterior arch of the atlas with an isolated posterior tubercle.¹⁵⁾ This case corresponds to our Type II-a, because of the lack

of the inward mobility of the isolated posterior fragment. The injury was severe enough to produce a vertebral body compression fracture, but it did not affect the stable occipito-atlanto-axial mechanism or the posterior fragment position, thus resulting in no neurological deficit.¹⁵⁾ All reported patients with neurological symptoms, who had to undergo operative treatment, thus correspond to our Type II-b.^{3,17)-19)} If atlantoaxial instability is not present, then good results have only been reported after an excision of an isolated posterior tubercle.

Our new classification for aplasia of the posterior arch of the atlas is simple, unambiguous and functional while also providing a clear direction for developing appropriate diagnostic and treatment strategies.

One limitation of the present study is the lack of experience with Type III and Type II-b cases. However, we believe that our review of the literature was extensive enough to allow us to develop a new classification system.

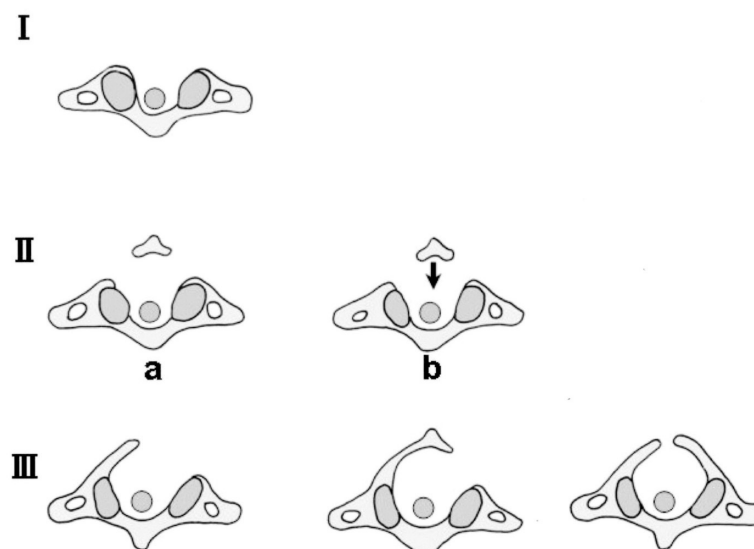


Figure 4. A new clinical classification for aplasia of the posterior arch of the atlas. Type I: complete aplasia. Type II: partial aplasia with an isolated posterior fragment. Type II-a; without inward mobility of the isolated posterior fragment. Type II-b; with inward mobility of the isolated posterior fragment. Type III is partial aplasia without an isolated posterior fragment.

Conclusion

Aplasia of the posterior arch of the atlas is often detected incidentally when a patient has cervical X-rays. Patients with this anomaly are generally asymptomatic. However, in some cases, clinical manifestations may be caused by this malformation. It is therefore clinically important, to predict cases demonstrating this anomaly that may be accompanied by neurological symptoms. We believe that our new classification described in this report may thus be a useful tool for predicting the clinical manifestations while also providing a clear direction for developing appropriate diagnostic and treatment strategies in the clinical management of aplasia of the posterior arch of the atlas.

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