Case Report

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Radiology Section

Congenital Hypoplasia of Posterior Arch of Atlas: A Case Report

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ABSTRACT

Congenital defects of the posterior arch of the atlas ranging from simple clefts to partial or total aplasia are quite rare with incidence varying from 0.69% to 4%. Most of them are asymptomatic and detected incidentally. However, a complete workup with advanced radiological imaging is necessary to identify the extent of the malformation and any associated anomalies for proper patient counselling to prevent any future neurological complications.

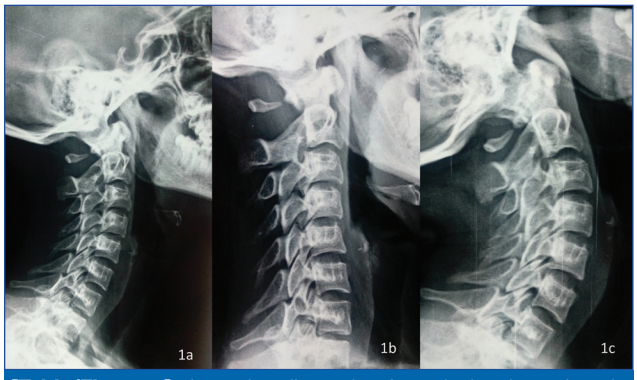
Here we discuss a rare form of congenital defect in posterior arch of atlas along with its clinical significance and a brief review of literature.

Keywords: Atlanto-axial instability, Currarino's classification, Posterior tubercle

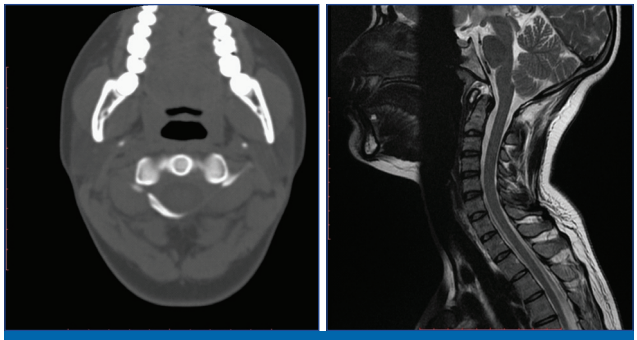
CASE REPORT

A 23-year-old young female, computer operator by profession presented with chronic mild occipital headache and upper neck pain with stiffness. General examination was unremarkable. Neurological examination including cranial nerves, motor and sensory system examination revealed no neurological deficits. All reflexes and visual acuity were normal. No clinical deterioration was found with neck flexion and extension.

Plain lateral radiograph of cervical spine [Table/Fig-1a] showed defect in posterior arch of atlas bilaterally. Radiographs in flexion and extension [Table/Fig-1b,1c] showed no evidence of atlantoaxial instability and no displacement or inward mobility of the posterior tubercle. The extent of the defect is better delineated in Computed Tomography (CT) [Table/Fig-2]. No evidence of spinal cord compression was shown on Magnetic Resonance Imaging [Table/Fig-3]. No other associated abnormalities, such as disc protrusion, craniovertebral junction malformation, Chiari malformation, syrinx or narrowing of the sagittal diameter at the level of C1 was noted. Patient was treated conservatively and advised regular follow-up. Surgical intervention was withheld because of lack of neurological symptoms and spinal cord compression. The main differential diagnosis is that of fracture or surgical excision. However, there was no history of trauma or surgery and radiologically it is differentiated from fracture as the margins are smooth and well corticated.



[Table/Fig-1a-c]: Lateral radiograph of cervical spine taken in neutral position; a) reveals bilateral defect (arrow) in posterior arch of atlas with posterior bony fragment; b) Lateral radiographs taken in flexion and; 1c) extension showing no displacement and no inward mobility of posterior tubercle during extension. No evidence of Atlantoaxial instability.



[Table/Fig-2]: Axial CT section at level of Atlas shows bilateral clefts more extensive on left side. (TYPE C).
[Table/Fig-3]: Mid sagittal T2 Weighted MRI image showing no evidence of spinal cord compression.

DISCUSSION

The C1 vertebra, also called "Atlas" is an atypical vertebra, which differs from other vertebra both in morphology and in embryological development. Atlas develops from three ossification centres, one for the anterior arch and two for the lateral masses. Lateral masses then extend towards the midline where they fuse to form the posterior arch [1]. Sometimes an additional ossification centre forms in the midline, which forms the posterior tubercle of the atlas. Clefts or aplasia of the posterior arch may result from either failure in fusion of ossification centres or absence of fourth ossification centre. Previously it was thought to be a primary defect in ossification, however is has been proved to be a defect in chondrification [2]. This has been proved intraoperatively and at autopsy [3].

Though many theories [2,4] have tried to explain the aetiology of congenital anomalies of atlas the exact cause remains obscure. Hereditary factors may have an important contribution. Motateanu M et al., [5] have reported a case of affected mother and daughter and Currarino G et al., [2] have reported a case of affected mother and son. However, most congenital defects in posterior arch of the atlas occur sporadically. Even though most of these occur in isolation they may be associated with syndromes affecting craniovertebral junction including Down's syndrome, Arnold chiari malformation, Klippel-Feil syndrome. These patients are more prone for atlantoaxial instability [6,7].

Incidence of posterior arch of atlas varies according to different anatomical and radiologic studies [2,4,7]. According to study by Currarino G et al., the overall incidence was only 0.69% with Type A being the most common variant comprising 97% of all posterior arch defects [2]. Senoglu M et al., [8] studied 1354 cases (CT-scans of 1104 patients, 166 dried atlas and 84 autopsies). The overall incidence of posterior arch anomalies in their study was 3.32%. Again Type A was the commonest and types C and D were not observed in their study. Guenkel S et al., [9] studied 1069 CT-scans and the overall incidence of posterior arch anomalies was 3.6% with Type A accounting for 3.2%. There were no Type D and E defects observed in their study.

Von Torklus D and Gehle W were the first to describe and classify congenital defects of posterior arch of the atlas [10]. Subsequently many classification system were proposed. The most widely accepted and currently followed morphological classification was given by Currarino G et al., [2] [Table/Fig-4].

Currarino G et al., also divided the posterior arch defects of atlas into five clinical groups based on clinical symptoms and findings. Group I - Asymptomatic and detected incidentally; Group II - Neck pain or/and stiffness; Group III - Acute neurological symptoms following neck or head trauma; Group IV - Chronic neurological symptoms before the diagnosis of the anomaly; Group V - Chronic symptoms referable to the neck [2].

Type	Description
A	Failure of posterior midline fusion of the two hemi-arches. Commonly appear as a fissure or a small gap in the midline.
B	Unilateral cleft. A defect is present on one side, which ranges from a small cleft to complete absence of one half of the arch
C	Bilateral clefts with preservation of the most dorsal part of the arch
D	Absence of the posterior arch with an isolated persistent posterior tubercle
E	Complete absence (aplasia) of the entire arch including posterior tubercle

[Table/Fig-4]: Currarino's morphologic classification for congenital defects of posterior arch of Atlas [2].

Most of these patients are asymptomatic. Various neurological presentations have been described clearly in few articles in literature [2,11-13] including intermittent tingling or shock like sensations, sensory symptoms such as paresthesia, motor symptoms like hemiparesis or quadriparesis. These symptoms may also be episodic in nature.

Patients with congenital defects of atlas must be evaluated for atlantoaxial stability with either plain lateral radiographs or CT in flexion and extension of neck. The bony gaps described in these anomalies are usually bridged by connective tissue rather than cartilage and this has been proved both by surgically and in autopsy specimens [3]. These dense fibrous bands and membranes provide good general stability of the cervical spine and prevent atlantoaxial subluxation/dislocation.

The most important factor in deciding neurological outcome and patient management is the presence or absence of posterior fragment or tubercle. In his study, Richardson EG et al., [12] attributed the symptoms of intermittent quadriplegia with compression of the cord by the inward mobility of the isolated posterior bony fragment during extension of the cervical spine. Inappropriate posture related to neck or repeated micro trauma may result in myelopathy, or compression on cervical part of spinal cord. So, Types C and D which have a free-floating posterior tubercle at the apex of the arch is a potential risk factor for neurological morbidity. Patients must be advised against participation in strenuous athletic sports and other contact sports, however normal activity should be encouraged.

Types A and B usually are asymptomatic and are less prone for neurological complications; hence, asymptomatic patients are to be followed up and treatment is usually conservative. Surgery is usually indicated when there is spinal cord compression or

atlantoaxial instability [13]. In patients with deficient posterior arch and associated atlanto- axial instability posterior fusion involving the occipital bone and the lower cervical segments is the recommended procedure. Surgical excision of the posterior tubercle is required when it is impinging on the cord [14].

CONCLUSION

Congenital anomalies of the posterior arch of the atlas vertebra though very rare are not always inconsequential normal variants. Exact knowledge of the type of anomaly and its implications is essential for radiologists and neurosurgeons in order to avoid diagnostic errors and to provide proper patient counselling and management.

CONSENT

Informed consent was obtained from the patient for publication of this case report, including accompanying images.

REFERENCES

[1] Junewick JJ, Chin MS, Meesa IR, Ghori S, Boynton SJ, Luttenton CR. Ossification patterns of the atlas vertebra. AJR. 2011;197:1229-34.

[2] Currarino G, Rollins N, Diehl JT. Congenital defects of the posterior arch of the atlas: a report of seven cases including an affected mother and son. Am J Neuroradiol. 1994;15:249-54.

[3] Krupanidhi S, Jayaprakash BR, Mallikarjun M and Phaniraj S. Congenital posterior arch defects of the atlas. IJBR. 2013;4(12):713-15.

[4] Sabuncuoglu H, Ozdogan S, Karadag D, Aynak ET. Congenital hypoplasia of the posterior arch of the atlas: case report and extensive review of the literature. Turkish Neurosurgery. 2011;21(1):97-103.

[5] Motateanu M, Gudinchet F, Sarraj H, Schnyder P. Case report 665. Congenital absence of posterior arch of atlas. Skeletal Radio.1991;20:231-32.

[6] Menezes AH. Hypoplasia of C-1 in children with Down syndrome. J Neurosurg. 2007;107:455-56.

[7] Pasku D, Katonis P, Karantanas A, Hadjipavlou A. Congenital posterior atlas defect associated with anterior rachischisis and early cervical degenerative disc disease: A case study and review of the literature. Acta Orthop Belg. 2007;73(2):282-85.

[8] Senoglu M, Safavi-Abbasi S, Theodore N, Bambakidis NC, Crawford NR SV. The frequency and clinical significance of congenital defects of the posterior and anterior arch of the atlas. J Neurosurg Spine. 2007;7:399-402.

[9] Guenkel S, Schlaepfer S, Gordic S, Wanner GA, Simmen HP, Werner CM. Incidence and variants of posterior arch defects of the atlas vertebra. Radiol Res Pract. 2013; Article ID 957280, 3 pages.

[10] Von Torklus D, Gehle W. The upper cervical spine. Regional anatomy, pathology and traumatology in: A systematic radiological atlas and textbook. New York: Grune and Stratton, 1972:41.

[11] Klimo P Jr, Blumenthal DT, Couldwell WT. Congenital partial aplasia of the posterior arch of the atlas causing myelopathy: case report and review of the literature. Spine 2003;28:E224-28.

[12] 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.

[13] Sharma A, Gaikwad SB, Deol PS, Mishra NK, Kale SS. Partial aplasia of the posterior arch of the atlas with an isolated posterior arch remnant: findings in three cases. Am J Neuroradiol. 2000;21:1167-71.

[14] Sridhar K, Jose B. " Floating" posterior tubercle of atlas as a cause of cord contusion: Case report and review of the literature. Spinal Surgey. 1(1):60-62.

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