IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 5 Ver. VI (May. 2015), PP 26-28
www.iosrjournals.org

Congenital Absence of Posterior Arch of Atlas with Atlantoaxial Subluxation-A Case Report

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Abstract: *Congenital absence of posterior arch of atlas with atlantoaxial subluxation is a very rare condition seen only in 4% of 1613 autopsies. In this we report a case of 12 year old boy presented with severe head ache over occipital area. There were no symptoms of neurological involvement. No muscle weakness of surrounding area was seen. After radiological investigation CT scan shows absent right lateral mass and posterior arch of atlas with rudimentary left posterior arch of C_1. He was diagnosed atlantoaxial dislocation and basilar invagination. This condition is asymptomatic and detected incidentally. Treatment for mild atlantoaxial subluxation is accomplished by wearing of cervical color. Most ideal treatment to correct subluxation is surgical correction by spinal fusion.*

I. Introduction

Congenital absence of posterior arch of atlas with atlantoaxial subluxation is a very rare condition. Giepel reported cleft of the posterior arch occurred in 4% of 1,613 autopsies[1]. Almost all cases are detected incidentally and misdiagnosed as a fracture or dislocation[2]. The spine assumed to be able to accommodate different regions of hyper mobility and fusion.

Structural defects of the posterior arch of atlas are rare, comprising abnormalities that may present as clefts with variable locations and size, ranging from small defects to more extensive defects such as complete agenesis. Anomalies like complete agenesis of posterior arch are asymptomatic. However they may be associated with atlantoaxial instability and neurological defects[3]. The severity of neurologic manifestations should serve as a marker of the degree of cervical involvement.

About 97% of posterior arch defects are type A. Congenital absence or hypoplasia the posterior arch of C_1 may also be associated with several genetic disorders such as Arnold- Chiari malformation, gonadal dysgenesis, Klippel- Feil syndrome, Turners and Downs syndromes[4,5].

In humans the junction between head and neck development corresponds to the boundary between the 4^{th} and 5^{th} somites[6] . Posterior arch of atlas arises from the dense area of sclerotome. The development of cervical spine particularly the upper cervical vertebra is closely related to the development of basiocciput. So anomalous development of cervical spine will affect both regions. Most defects of the atlas do not contribute to abnormal occipito cervical anomalies and are not associated with basilar invagination[7].

Atlantoaxial subluxation is a condition occurred due to non fusion of odontoid and body of C_2 causing impairment of rotation of the neck. The anterior facet of C1 is fixed on the facet of C_2. It may be associated with dislocation of the lateral mass of C_1 on C_2 . There are several ways in which a sublaxation can occur. 1) Antero-posterior subluxation 2) Rotatory subluxation.

II. Case Report:

A 12 year old boy presented with suboccipital head ache since 2 months and on and off in nature . Head ache is severe and intermittent, localized to occipital area .The episode lasting for 10-15 minutes. The child became semiconscious for 1 minute followed by redness of eyes. Neurological signs like parasthesia of both upper limbs and lower limbs with exaggerated deep tendon reflexes. He found with no other neurological symptoms. No muscle weakness of surrounding areas was seen. There was no history of spinal trauma.

He was sent for routine laboratory and radiological investigations. His CT spine report shows absent right lateral mass and right posterior arch of atlas with rudimentary left posterior arch of C_1.There is partial fusion of posterior element of C_2 and C_3. Finally he was diagnosed as craniovertebral junction anomaly with atlantoaxial dislocation and basilar invagination. He was sent for X-Ray C- spine including CV junction ,Computed tomography (CT) and Magnetic Resonance Imaging (MRI) scan to confirm the diagnosis. His CT and X-Ray of CV junction show partial occipitalization of atlas with fused right half and absent posterior arch.

Atlanto odontoid subluxation with basilar invagination. MRI shows there is a compression at the cervico medullary junction.

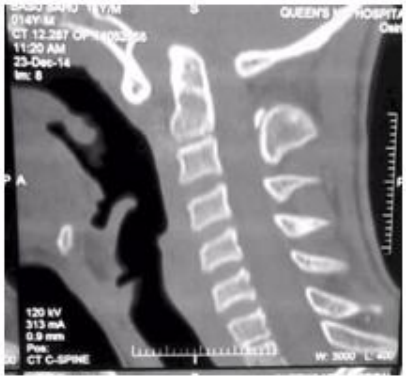

Fig.1- CT Scan sagital views showing posterior Arch absence and basilar invagination

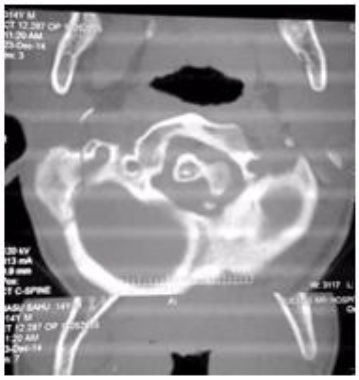

Fig.2- CT Transverse section showing absence of posterior arch of Atlas and fusion to the lateral mass of Atlas

CT angiogram report shows bilateral normal internal carotid arteries. Vertebro-basilar system is normal. No evidence of aneurysm or arterio venous malformation is seen

III. Discussion

Posterior arch of atlas begins its ossification during the seventh week of intrauterine life. The vertebral laminae arises from the buds in the chondrification centers and extends dorsally, being fused at the time of birth, except for some length of cartilage. Complete fusion of posterior arch is expected to occur between 3 and 5 years of age. In about 2% of population, additional centers may be present posteriorly in the midline forming the posterior tubercle of the atlas during the 2nd year of life[1-6].

At least two different anomalies can develop during the ossification process.
1) Median clefts of the posterior arch of C_1
2) Varying degrees of posterior arch dysplasia[1,5].
These findings are further classified as
Type A - Median cleft of posterior arch of C_1
Type B- Unilateral cleft defect
Type C- Bilateral cleft defect
Type D- Absence of posterior tubercle
Type E-Total agenesis of posterior arch

Type A clefts occur in about 4% of the population and represents 97% of posterior arch defects, These disturbances have been attributed to anomalies in the cartilaginous formation of posterior arch rather than disturbances of ossification[3,5].Posterior arch absence has been reported as congenital anomaly. Absence of posterior arch is associated with chromosomal disorders such as Downs and Turners syndromes, Arnold- Chiari malformations and gonadal dysgenesis.

Total agenesis of the posterior arch of atlas is rare. This condition is detected incidentally and sometimes symptomatised by neck pain . This defect can be associated with atlantoaxial instability and neurological deficits. Neurological examination should be conducted carefully. In this examination upper motor neuron signs including hyperreflexia, clonus and extensor plantar reflexes may be indicative symptomatic atlantoaxial instability. Somato sensory evoked response may reveal information regarding neurological involvement. In individuals with rotator displacement torticollis may be the presenting symptom.
Atlantoaxial subluxation can occur in several ways.
I .Anteroposterior subluxation.
II .Rotatory subluxation, characterized in four different types
Type 1-The atlas is rotated on the odontoid with no anterior displacement
Type 2-The atlas is rotated on one lateral articular process with 3 to 5 mm of anterior displacement.
Type 3-Comprises a rotation of the atlas on both articular process with anterior displacement greater than 5 mm.
Type 4 -Posterior displacement of the atlas

In this case report atlantoaxial distance is increased to 4.5 mm with marked compromise of the foramen magnum with spinal cord compression. Rest of the vertebral column is normal. CT cervical spine report shows absent lateral mass and right posterior arch with rudimentary left posterior arch of C_1. There is basilar invagination with 4 mm tip of odontoid process projecting above the foramen magnum. In this case we found atlantoaxial dislocation with 5 mm distance between posterior margin of atlas and anterior margin of odontoid process.

CT images of CV junction in axial, sagital and coronal sections finding show partial occipitalisation of atlas with a fused right half and absent posterior arch. MRI shows there was atlanto odontoid subluxation with basilar invagination of odontoid causing narrowing of the foramen magnum and compression on the cervicomedullary junction. The atlanto odontoid interval measures 6 mms and retro odontoid space measures 8 mm in AP diameter. No evidence of any cord edema or myelomalasia. He was posted for neurosurgery.

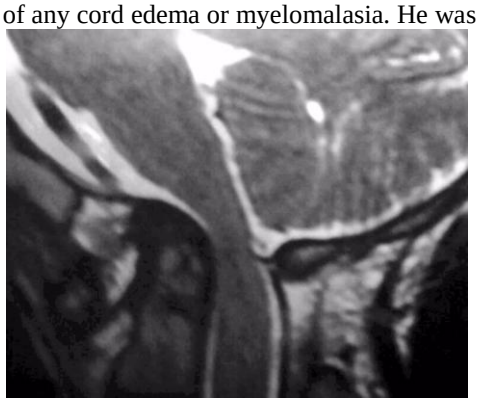

Fig.3- MRI image shows Cervicomedullary junction compression

Atlantoaxial subluxation risk seen in the 26-38% children aged 2-3 years. Compensatory hypertrophy of anterior arch. CT provides excellent imaging contrast between ossified and non ossified portions of the posterior arch of the atlas. CTs are useful particularly in minor conditions. MRI scan is useful in neurological symptoms particularly of spinal cord evaluation. In mild atlanto axial subluxation the goal may to provide neck stability, which could be accomplished by wearing a soft or hard cervical collar. Moderate instability may require traction or stabilizing techniques to correct free floating neck. Most ideal treatment to correct atlantoaxial subluxation is surgical correction. Spinal fusion can prevent misalignment though it may also reduce the range of movement.

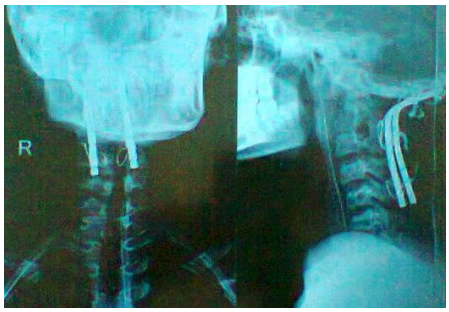

Fig.4- Radiological images of spinal fusion

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