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Congenital Unilateral Atlanto-Occipital Rotatory Subluxation – Rare cause of C1 Neuralgia

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Abstract

Study Design: Case report

Objective: We report a rare case of congenital unilateral rotatory atlanto-occipital subluxation that presented with left C1 neuralgia.

Summary of Background Data: Secondary occipital neuralgia is commonly attributed to pathologies of the atlanto-axial joint and C2/C3 nerve involvement. Our case depicts a model of slow creeping atlanto-occipital subluxation due to a rare left C1 superior articular facet dysplasia with C1 foraminal stenosis presenting as C1 neuralgia. We discuss the eitology and patho-anatomy of this rare undescribed presentation.

Methods: A 42year old gentleman presented with deteriorating and intractable left occipital headache of 6months duration. The NDI(neck disability index) was 64%. Cervical MR/CT scan showed an unilateral C1 facet dysmorphism with a left sided C1 foramen bony compression. There was no central canal stenosis.

Results: Posterior left C1 arch excision and decompression of C1 foramina with occipital-cervical fusion relieved C1 neuralgia.

Conclusion: Our case depicts a model of slow creeping deformation due to left C1 superior articular facet dysplasia. An abnormal facet slope allowed the occipital condyle to migrate posteriorly and medially leading to crowding of the left C1 foramen. Although the etiology was congenital, the neck spasm was delayed till fourth decade. A secondary C1 foramen stenosis led to C1 occipital neuralgia that presented as an intractable headache.

Keywords: congenital; Cranio-vertebral junction; Foraminal compression; Facet subluxation; instability; cervical pain; facet dysplasia; facet dysmorphism; occipital neuralgia, occiput-cervical fusion

Level of Evidence: 5

Introduction:

We report a rare case of congenital unilateral rotatory atlanto-occipital subluxation that presented with left occipital neuralgia. To our knowledge this has not been reported in Literature. A posterior left C1 arch excision and decompression of C1 foramina along with occipital-cervical fusion relieved the C1 neuralgia.

Case History:

A 42year old gentleman presented with deteriorating and an intractable left occipital headache for 6months. There was no significant trauma. He was misdiagnosed as migraine and treated with medications and physiotherapy. Sleep was disturbed. There was a left torticollis. Attempted correction induced severe spasm. Slow flexion and extension movement was possible with terminal spasm. The NDI(neck disability index) was 64%. C2 dermatomal sensation(Greater occipital nerve) was normal. Cranial nerves examination was normal. There was no myelopathy. Markers for infection(CRP/ESR) and inflammatory spondylo-arthropathy(RA/HLA B27/Anti CCP) were normal.

Open-mouth and lateral radiographs showed normal soft tissue and bony relationships. MRI showed posterior subluxation of left occipital-cervical joint with left C1-foraminal stenosis. CT scan revealed a left C1 superior articular facet dysplasia(SAFD). Left facet sloped 49^0 posteriorly and 45^0 medially (25^0&15^0 respectively on right). The left occipital condyle showed mild deformation and had migrated postero-medially, causing C1-foraminal stenosis. Posterior C1 arch was bifid. Degenerative changes were observed around the occiput and dens. Reduced dens and left O-C1joint distance on coronal CT image implied rotatory subluxation. Atlanto-axial articulation was normal. Dynamic MRI ruled out sagittal instability(figure1A-F).

Posterior C1 arch excision with Occipito-C2 fixation was done (Figure 2). The patient experienced dramatic resolution of neuralgia. At 2 year follow-up, NDI was 6% with painless limitation of neck rotation & terminal flexion-extension. He resumed his normal activity.

Discussion:

Superior articular facets of atlas like a chalice, support the convex occipital condyles. This orientation permits sagittal movements (flexion23^0-extension24^0) while restricting rotations & lateral bending.[i] Impingement of dens against basion limits flexion while locking of facets limits extension.[ii] Our case depicts a model of slow creeping deformation due to a rare left C1 SAFD.

An abnormal facet slope allowed the occipital condyle to migrate posteriorly and medially. Surprisingly no neck-pain ensued. Altered weight bearing vectors, produced secondary ossification around tip of dens suggesting chronicity but the contralateral O-C1 & C1-2 articulation remained normal. C1 foramen stenosis due to displaced occipital condyle produced C1 neuralgia. As per authors' knowledge, unilateral C1 facet dysmorphism is unreported in literature.

Although C1 nerve has no cutaneous distribution, the dorsal ramus supplies sensory innervation to deep somatic tissues in sub-occipital region including short muscle of occiput triangle and the ventral ramus gives innervation to sternocleidomastoid trapezius and OC joint. The sinu-vertebral nerve of C1 innervates medial C1-C2 joint, dura and vertebral artery in conjuction with the sinovertebral nerves of the C2 and C3.[iii] Hollinshead described the Cruveilheir plexus as neural interconnection in upper cervical nerves (sensory branches of C1, C2 & C3).[iv] C1 root producing occipital headache is unexplained. Although the etiology was congenital, neck spasm was delayed till fourth decade Authors hypothesize, C1 radiculopathy occurred through this plexus and presented as occipital neuralgia. The commoner causes of occipital neuralgia are trauma, myositis, atlas fracture, C1-C2 lateral mass arthritis and tumour effecting C2/C3 nerve roots.[v]

Altered left occiput-C1 joint morphology inhibited anatomical reduction. Posterior C1 arch excision relieved C1root compression. Posterior fixation was extended up to C2 because of left hypoplastic C1 lateral mass.

Conclusion:

The case depicts a unilateral SAFD of C1 with posterior rotatory subluxation causing secondary C1 foramen stenosis. A rare C1 occipital neuralgia ensued presenting as intractable headache. Diagnosis was delayed due to lack of suspicion and subtle radiological signs

Left C1 foramina decompression with prophylactic fixation of occiput-C2 resolved left occiput C1 neuralgia.

[i] Alejandro J, Justin K, Kayla E, Zachary A, Brian J, Nader S. Anatomy and biomechanics of the craniovertebral Junction. Neurosurg Focus 38 (4):E2, 2015

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[iii] Bogdug.N. Cervicogenic headache: Anatomical basis and pathophysiological mechanisms. Current pain and headache reports, 2001, 5: 382-386.

[iv] Choi II, Jeon Sang Ryong. Neuralgias of Head: Occipital Neuralgia. J Korean Med Science 2016; 31; 479-488.

[v] Samer Narouze. Occipital Neuralgia Diagnosis and Treatment: The role of Ultrasound. Headache 2016; 801-807.

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Figure I : A) Left Occiput-C1 rotatory subluxation**. B)** MRI T2 Axial image, left Occiput-C1 foramina narrowing due to subluxation of left occiput- C1 facet. **C)** Flexion-extension MRI, no dynamic instability. **D)** Rightt side normal Occiput-C1 joint **E)** Posterior subluxation of Left Occiput condyle over C1, abnormal joint variance. **F)** CT scan Coronal view, Reduced interval between dens and left O-C1 joint on left side as compared to right side, abnormal joint variance.

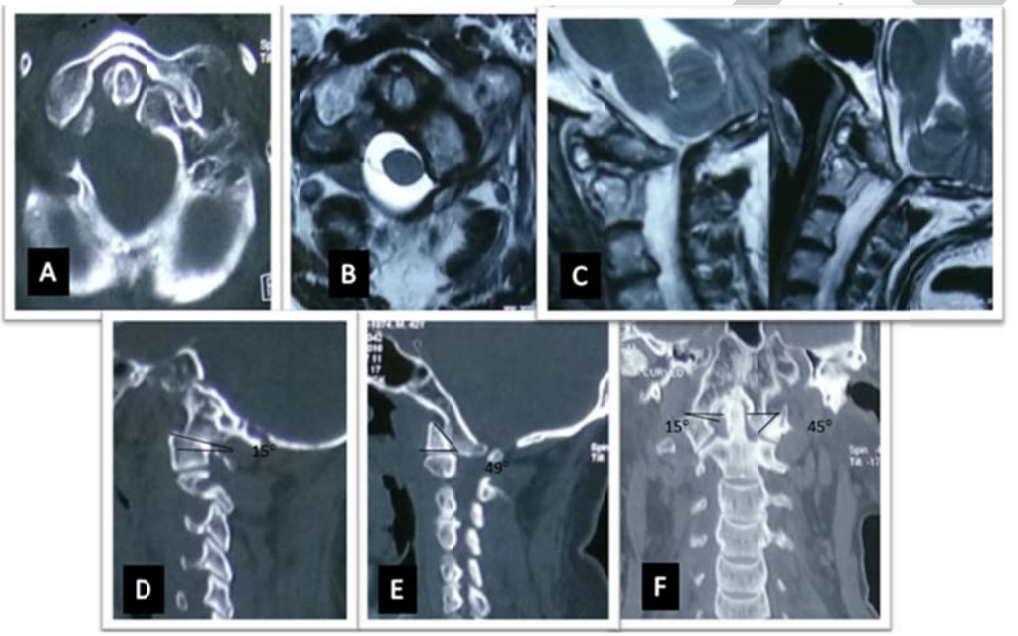

Figure 2: Post operative Radiograph and CT Scan, left posterior C1 arch excision for foraminal decompression.