Basilar Imagination : A Rare Cause Of "Top Of Basilar Artery" Syndrome.

Chauhan S R*, Shah C A**

*Professor, **Resident, Department Of Radiodiagnosis, GMC, Bhavnagar

Abstract: "Top of Basilar Artery" Syndrome as the presentation of CVJ anomalies is extremely rare. The association between skeletal CVJ anomalies and vertebro-basilar insufficiency (VBI) is recognised and angiographic abnormalities of the vertebro-basilar arteries and their branches have been reported. Atlanto-axial dislocation (AAD) is the commonest skeletal cranio-vertebral junction (CVJ) anomaly in India, followed by occipitalisation of atlas and basilar invagination. They usually present with a progressive neurological deficit (70 - 94% cases) implicating the high cervical cord, lower brainstem, and cranial nerves. We report one such case with the even more rare presentation of "Top of Basilar Artery" Syndrome as the initial presentation of basilar invagination.[Chauhan S NJIRM 2014; 5(3) :138-141]

Key Words: Basilar Imagination, Craniovertebral junction craniometry, Platybasia, Top of Basilar Artery Syndrome

Author For Correspondence: Shah C A,Resident, Department Of Radiodiagnosis, Government Medical College , Bhavnagr 364001; Email: dr.chirag_shah@yahoo.in

Introduction: The clinical and radiological profile of cranio-vertebral junctions is very well documented and atlanto-occipital dislocation (AAD) is the commonest type reported from India[1]. Vertebro-basilar territory infarction however, is one of the rarer presentations of CVJ anomalies. We report one such case with the even more rare presentation of "Top of Basilar Artery" Syndrome as the initial presentation of basilar imagination.

Case History: A 35 year old male presented with history of sudden onset of blindness, behavioural abnormalities and hallucinations. There was no history of headache, vomiting, cranial nerve involvement, speech difficulty, motor weakness, sensory loss, limb in coordination, or any bladder disturbance. There was no history suggestive of hypertension, diabetes, rheumatic heart disease, or any neck pain, manipulation, or trauma. On examination at the time of presentation, he was conscious and alert but withdrawn. On general examination patient had abnormal neck posturing with slight tilting of neck towards left side. Vitals were preserved and systemic examination was normal. On detailed cognitive examination, his attention was normal and comprehension and speech were intact. On cranial nerve examination, visual acuity, fundus examination and field of vision were normal. Other cranial nerves were intact. On motor examination, size, tone, power and reflexes were normal in all the four limbs and plantars were bilaterally flexor. There was no sensory loss. On cerebellar examination, he was found to have no intention tremor, finger nose incoordination, past-pointing, dysdiadochokinesia. In view of the history of a sudden onset of bilateral visual impairment, behavioural abnormalities and hallucinations, a possibility of a posterior circulation stroke affecting thalamus and bilateral occipital lobes was entertained. The presence of abnormal neck posturing raised the suspicion of possibility of an associated cranio-vertebral junction (CVJ) anomaly.

His investigations revealed a normal haemogram and ESR. Blood sugar, LFT, RFT, lipid profile, electrolytes were within the normal range. Tests for HIV, rheumatoid (RA) factor were negative. X-ray chest and ECG were normal. MRI of the brain and CV junction without administration of Gadolinium revealed acute infarcts involving bilateral thalamus and occipital lobes (Fig-1-4).

Figure 1: Axial T2 Weighted Image Showing Hyperintensity Involving Bilateral Thalamus And Occipital Lobes (Open Arrows)

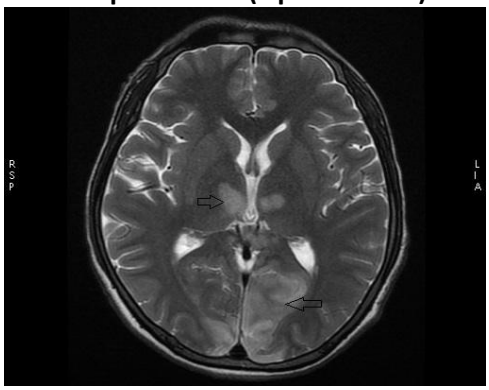


Figure 2: Axial T2 Weighted Image Showing Hyperintensity In Bilateral Parieto-Occipital Lobes (Open Arrows)

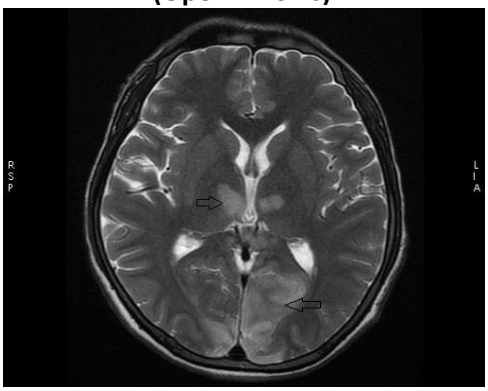


Figure 3 :DW Image Showing Restricted Diffusion In Bilateral Thalami And Occipital Lobes (Open Arrows)

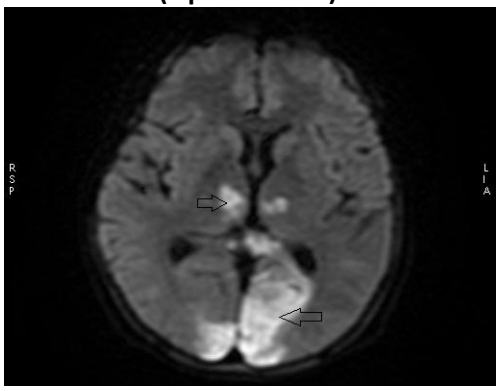
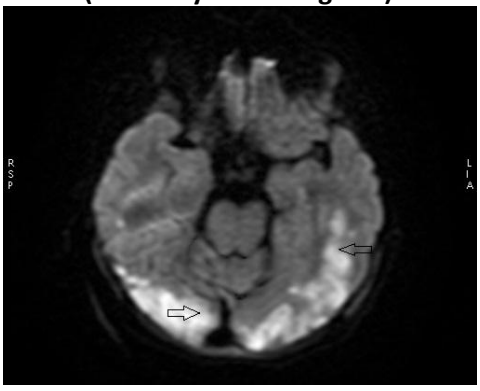


Figure 4: Saggital T1W Image Showing Welcher Basal Angle Measuring 162 Degrees (Normally: <140 Degrees)



Occipitalization of atlas with basilar invagination and platybasia was noted causing compression of lower brain stem and upper cervical spinal cord (Fig.8). Craniovertebral junction craniometry revealed abnormal measurements in the form of increased Welcher angle suggesting platybasia, decreased Cranio-Vertebral angle suggesting basilar invagination. The tip of odontoid was quite higher above the Chamberlain line suggesting basilar invagination (Fig. 5-7).

Discussion: Congenital cranio-vertebral junction (CVJ) anomalies encompass the developmental defects of the occipital bone surrounding the foramen magnum, atlas and axis vertebrae. During foetal development, the mesodermal somites form 4 occipital and 2 cervical sclerotomes that form the CVJ. Defects occurring in the 3rd and 4th week of embryogenesis can cause CVJ anomalies which may implicate either the skeletal or neural structures or both[1,2]. The bony anomalies may implicate the occiput (basilar invagination and platybasia) or the atlas (occipitalisation of the atlas and atlanto-axial dislocation) or axis (odontoid malformations) and other vertebrae (Klippel-Feil anomaly)[1,2]. Neural or soft tissue anomalies include Chiari malformation, syrinx, and Dandy-Walker syndrome)[1,2]. Presently, there is no epidemiological data available, and most of the information regarding skeletal CVJ anomalies is derived from

Figure 5 : DW Image Showing Restricted Diffusion In Bilateral Parieto-Occipital Lobes(Open Arrows)

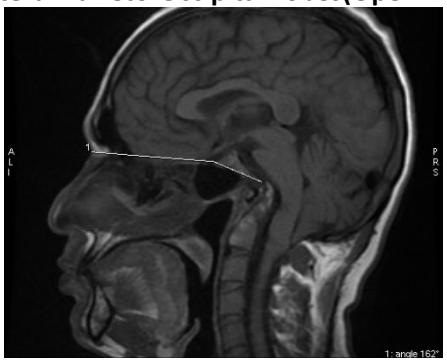
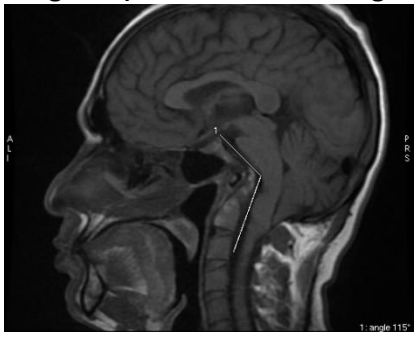


Figure 6 : Sagittal T1W image demonstrating cranio-vertebral or clivus-canal angle measuring 115 degrees (Normal: 150-180 degrees)



retrospective reviews of hospital records. CVJ anomalies are more common in India as compared to the West. Basilar invagination is more common in the West, whereas atlanto-axial dislocation (AAD) is the commonest in India, followed by occipitalisation of atlas, and basilar invagination. AAD, occipitalisation of atlas, and fusion of C2 and C3 vertebrae are the commonest anomalies occurring in combination in India[1]. The CVJ anomalies occur since birth, but the mean age of manifestation is around 25 years. Males are most commonly affected and a predisposing factor like trivial neck trauma may be identified in nearly 50% of the cases. Our patient was also a male and first became symptomatic at 35 years of age but there was no history of preceding neck trauma or manipulation. Skeletal CVJ anomalies commonly present with aprogressive neurological deficit (70-94% cases) implicating the high cervical cord, lower brainstem, and cranial nerves[1]. The clinical features include spasticquadriparesis (100% in some series), but hemiparesis and Brown-Séquard syndrome can also occur[1]. Sensory loss is seen in around 33% cases, and loss of proprioception is more prominent in fingers than toes[1,2]. Lower cranial nerve involvement is seen in 12%, and cerebellar signs in 10% cases[1,2]. Bilateral hand wasting is a common (25% cases) false localising sign[1,2]. Urinary involvement is uncommon. Mirror movements and pseudoathetosis are infrequent but useful signs in the diagnosis of CVJ disorders. Transient deficits are seen in around 20% patients and may include motor weakness, sensory disturbances, visual loss, dysarthria or inability to speak, and urinary disturbances. These may last from a few minutes to hours and are possibly due to platelet microembolisation in the vertebro-basilar artery territory[1,2]. Our patient however presented as a case of "Top of Basilar Artery" syndrome with sudden onset of bilateral visual impairment, behavioural abnormalities and hallucinations. "Top of Basilar Artery" syndrome is usually caused due to embolism followed by atherosclerosis involving the distal basilar artery. The postulated pathology behind "Top of Basilar Artery" syndrome in a case of basilar invagination is due to thrombosis of the distal basilar artery secondary to chronic repeated microtrauma of the artery. MRI is the most accurate to evaluate the CVJ anomalies with the help of various craniometric measurements[3].

Figure 7 : Sagittal T1W Image Demonstrating Chamberlain Line And The Distance Of Tip Of Odontoid Process And Anterior Arch Of C1 From It. (Normal: Max. Distance That The Tip Of Odontoid Can Be Above This Line Is 1+/- 3 Mm. Anterior Arch Of C1 Should Be Below This Li)

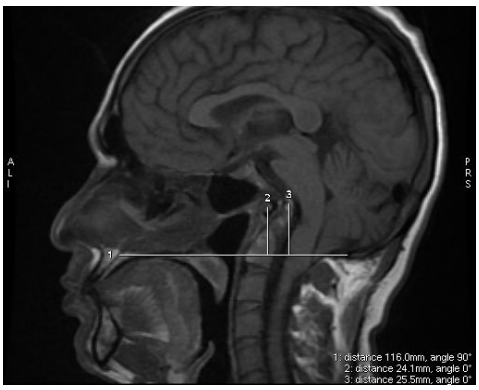
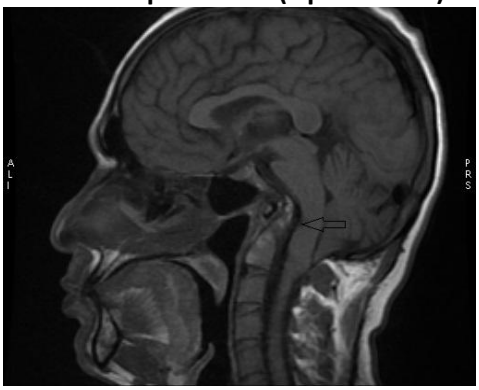


Figure 8 : Sagittal T1W Image Demonstrating Basilar Invagination And Platybasia With Compression Of Lower Brain Stem And Upper Cervical Spinal Cord (Open Arrow)



The association between vertebro-basilar insufficiency(VBI) and skeletal CVJ anomalies is well-recognised but may be under-estimated, as according to some studies only 30% of cases with VBI undergo X-rays of the cervical spine, and only 11% are evaluated using proper flexion and extension views of the CVJ per se [4,5]. However, "Top of Basilar Artery" syndrome as the presentation of CV junction anomalies, as was observed in our case, is extremely rare. The clinical rarity of posterior circulation infarcts in CVJ anomalies has been attributed to dual supply through the two vertebral arteries and the

adequacy of the circulation from the circle of Willis[4].

Atlanto-axial dislocation[1,4,6,7,8], is the commonest CVJ anomaly implicated in causing stroke or VBI, followed by odontoid aplasia[9], basilar impression, occipitalisation of the atlas, Klippel-Feil anomaly[1], and anomalous osseous process of the occipital bone projecting to the posterior arch of the atlas[10]. Our patient also had basilar invagination with platybasia with atlanto-axial dislocation and occipitalization of atlas.

In conclusion, physicians should be aware of the uncommon or rare presentations of CVJ anomalies. All young patients presenting with features of VBI or posterior circulation stroke should be screened for CVJ anomalies.

References:

1. Wadia NH, Bhatt MH, Desai MM et al. Myelopathy of congenital atlantoaxial dislocation. In: Chopra JS, editor. Advances in Neurology. Amsterdam: Elsevier Science Publishers, B.V. (Biomedical division) 1990; 2: 455-64.
2. Shukla R, Nag D. Craniovertebral anomalies: The Indian scene. Neurosciences Today 1997; 1: 51-8.
3. Craniovertebral junction: normal anatomy, craniometry, and congenital anomalies. Wendy R K. Radiographics 1994; 14 (2)
4. Agrawal D, Gowda NK, Bal CS et al. Have cranio-vertebral junction anomalies been overlooked as a cause of vertebro-basilar insufficiency. Spine (Phila Pa 1976) 2006; 31 (7): 846-50.
5. Lorenstan KJ, Schrospshire LC, Ahn HS. Congenital odontoid aplasia and posterior circulation stroke in childhood. Ann Neurol 1988; 23: 410-13.
6. Ando S, Matsui Y, Fujii J et al. Cerebellar infarctions secondary to cranio-cervical anomalies: a case report. Nippon Geka Hokan 1994; 63 (4): 148-54.
7. Singer WD, Haller JS, Wolpert SM. Occlusive vertebrobasilar artery disease associated with cervical spine anomaly. Am J Dis Child1975; 129: 492-5.
8. Shim SC, Yoo DH, Lee JK et al. Multiple cerebellar infarction due to vertebral artery obstruction and bulbar symptoms associated with vertical subluxation and atlanto-occipital subluxation in ankylosing spondylitis. J Rheumatol 1998; 25 (12): 2464-8.
9. Phillips PC, Lorentsen KJ, Shropshire LC, Ahn HS. Congenital odontoid aplasia and posterior circulation stroke in childhood. Ann Neurol 1988; 23 (4): 410-13.
10. Tominaga T, Takahashi T, Shimizu H, Yoshimoto T. Rotational vertebral artery occlusion from occipital bone anomaly: a rare cause of embolic stroke. Case report. J Neurosurg 2002; 97 (6): 1456-9.
11. Khwaja G A, Batla A, Gupta M et al. Posterior circulation stroke as the initial manifestation of CVJ anomaly with Atlanto-Axial Dislocation. Journal, Indian Academy of Clinical Medicine 2011; 12 (1)

Conflict of interest: None
Funding: None