



PLATYBASIA AND BASILAR INVAGINATION PRESENTING AS CEREBROVASCULAR ACCIDENT

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ABSTRACT The congenital and acquired deformities of the craniovertebral junction (basilar invagination, basilar impression, or platybasia) can present in the form of slowly progressive or acute neurologic deterioration in second decade . Basilar invagination, the commonest malformation of the craniocervical junction , is a well recognised cause of brainstem compression.

KEYWORDS : Platybasia , Basilar invagination.

1. Introduction

Basilar invagination is a developmental anomaly of the craniovertebral junction in which the odontoid abnormally prolapses into the foramen magnum[1]. It is often associated with other osseous anomalies of the craniovertebral junction, including atlanto-occipital assimilation, incomplete ring of C1, and hypoplasia of the basiocciput, occipital condyles, and atlas. Basilar invagination is also associated with neural axis abnormalities, including Chiari malformation, syringomyelia, syringobulbia, and hydrocephalus.

Platybasia is defined as an increase in the basal angle of the skull which is due to flattening of the base of the skull[2]. Most patients with basilar invagination have a normal basal angle and do not have associated platybasia[3,4].

We present a case of craniovertebral junction anomaly presenting in second decade as hemiparesis and bowel and bladder involvement.

2. Case report

A 14 year old girl, sixth standard student, presented with difficulty in lifting her right arm above the shoulder when she got up in morning 3 months back. She also noticed that she is not able to perceive bladder and bowel sensations and unknowingly she is making her bed wet. The weakness remained static for 2 months but gradually progressed to involve right lower limb over the next 1 month and then to left lower and upper limbs after 15 days.

The weakness was associated with inability to feel her clothes and hot and cold water over right upper and lower limbs since 3 months.

She also has history of fever during last 3 months which subsided on taking medications. There was no history of kochs or kochs contact, trauma.

On examination, pt. is lying comfortable in bed in supine position with normal vitals. No signs of meningeal irritation. higher functions were normal with no abnormality in speech, memory , intelligence. Cranial nerve examination showed no abnormality. There is hypertonia (clasp knife spasticity) in all 4 limbs. grade 4/5 in all 4 limbs. Deep tendon reflexes were brisk and bilateral plantar were extensor. Finger nose test was impaired on right side. Horizontal nystagmus was present on lateral gaze.

A provisional diagnosis of gradual onset chronic spastic quadriplegia with cerebellar involvement was made.

MRI brain and cervical spine was done suggestive of overcrowding of

brain stem and cerebellar tonsils (4 mm herniation) at foramen magnum with anteriorly split atlas causing indentation over brain stem suggestive of split atlas with platybasia. Short clivus with mildly retroflex dens. Lacunar infarct in head of caudate nucleus on left side.

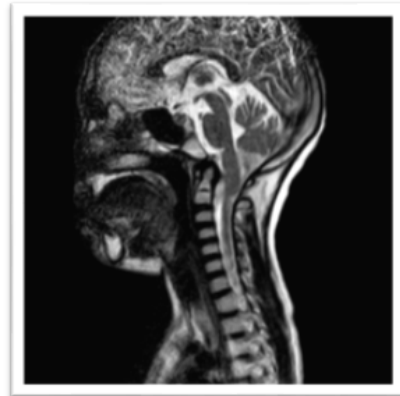


Figure 1 MRI CERVICAL SPINE showing short clivus with retroflex dens and overcrowding of cerebellar tonsils at foramen magnum

3. Discussion

According to Spillane et al. [5] one of the first accounts of basilar invagination was that of Anders Adolph Retzius (1796–1860) and Frederik Theodor Berg (1806–1887) in 1855. Platybasia is defined as a skull base with an abnormally obtuse angle between the plane of the clivus and the plane of the anterior fossa, which is greater than 142° which is due to flattening of the base of the skull. Not infrequently, platybasia is associated with basilar invagination or basilar impression. Platybasia itself is not a disease. Patients with minimal symptoms can be treated with non-operative modalities such as physical therapy, non-steroidal anti-inflammatory medication, or a cervical collar.

Surgical treatment is reserved for patients with symptoms refractory to non-operative management, neurological deficit, or severe spinal cord compression. Surgery usually involves the removal of bone that is causing the compression and stabilization with a fusion.

Although the term “platybasia” has also been used as synonymously of basilar impression and basilar invagination, it is not currently recognized as such. Platybasia is the term used for the condition in

which the base of the skull appears relatively flattened. This young patient presented with right sided hemiparesis progressing to involve left side has platybasia with basilar invagination.

Basilar invagination and basilar impression are a congenital or acquired craniocervical junction anomaly, that occurs when the superior part of the odontoid (part of the C2 vertebra) migrates upward. These terms basilar impression and basilar invagination are often used interchangeably because in both cases there is upwards migration of the upper cervical spine but they are not synonymous.

Basilar invagination is a developmental anomaly of the CVJ where the odontoid process is positioned abnormally upward and backward, prolapsing into the foramen magnum.

In our case, the patient presented with symptoms and signs of right sided hemiparesis with bladder and bowel involvement causing confusion in clinical diagnosis. Her NCCT brain showed no gross abnormality. But her MRI brain and cervical spine suggestive of overcrowding of brain stem and cerebellar tonsils (4 mm herniation) at foramen magnum with anteriorly split atlas causing indentation over brain stem suggestive of split atlas with platybasia. Short clivus with mildly retroflex dens. Lacunar infarct in head of caudate nucleus on left side.

She later underwent an operative procedure (C1 C2 lateral mass fixation) and observed a remarkable improvement in stiffness, power & tingling sensation in all 4 limbs. Thus we should always have a suspicion of cv junction anomaly in young individuals presenting with quadriparesis.

4. Conclusion

We are reporting a case of 14 year young girl with Platybasia who presented with sensorimotor and bowel bladder involvement improved after surgery.

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