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Curse Behind the Curtain- Arnold Chiari Syndrome Type 1 A Rare Case Report

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Abstract

Chiari malformations are developmental abnormalities involving hindbrain. It is a condition in which brain extends into spinal cord. A 21 year old male patient visited dental hospital for his routine dental evaluation. As an incidental finding it was identified and further by radiographic evaluation. Most of times it will be asympomatic and show its manifestations in late 30 s. patient should be monitored with regular intervals. Symptomatic findings like pain can be relived by using NSAIDS, Muscle relaxants etc.. If required surgical management can also be done. Early identification of lesion may prevent the secondary complications.

Keywords: Asympomatic, Rhombencephalon, Cranioventra, Pathological Herniations

Introduction

Chiari Malformations comprises group of abnormalities involving Rhombencephalon (Hind Brain) and contents of the CranioVentral junction. Chiari malformations are pathological herniations of hindbrain through the foramen magnum and into the cervical spinal cord. In 1883, John Cleland described a case of hindbrain malformation, which was found during autopsy. Hans Chiari, an Austrian pathologist, gave a detailed description of hindbrain malformations after performing post-mortem examination of forty cases in the year 1891 to 1896. Hence named as Chiari malformations.[1] Chiari stated in his descriptions that these malformations are congenital anomalies of the hindbrain characterized by downward elongation of the brain stem and cerebellum into the cervical portion of spinal cord.[2] Chiari classified the hindbrain malformations basing on their anatomical variations into type I, II and III and type IV malformation.[3] According to the theory given by Daniel and Strich, Chiari Malformations are mainly because of developmental arrest, especially in the progression of pontine flexure during 28th and 29th day of gestation.[4] The theory of overgrowth suggest that the overgrowth of neural plate before neurulation prevents fusion of neural folds. Barry et al reported two cases of human foetuses of 17 and 18 weeks of development with increased volume of cerebellum and brain stem having Chiari malformation.[5] and also observed that cerebellum

weighs less in patients with Chiari malformation than in normal people, at all ages.[6,7] According to hydrodynamic theory, imbalance between pulsating choroid plexus of forth and lateral ventricles result in Chiari malformation.[6] According to Jenning et al, Chiari malformation occurs because the normal zone of fusion at third and fourth somites is displaced caudally below the third to fifth somite pairs thus causing the displacement of the area of formation of cervicomedullary junction.[8] Chiari malformation is not as rare as would be expected from the small number of reported cases but with the increased use of Advanced imaging modalities like CT Scans and M.R.I's it is reporting to be much more common. The defect is almost always, but not invariably, associated with meningomyelocele or spina bifida occulta in lumbosacral region. Hydrocephalus is present in most cases. Other associated defects of development include creniolacunia, hydromyelia, sryngomelia, double cord, and basillar impression.[2]

Case Report

A 21 year male patient visited dept of oral medicine and radiology with a chief complaint of deposits on his teeth region. The patient medical and dental history was non contributory. The patient was moderately built and moderately nourished. The patient vital signs are within the normal range. Patient also reported frequent headache, intermittent cough, and back pain. Patient also reported aggressiveness of back pain while coughing. Patient also reported no adverse and parafunctional habits.

Patient was conscious, well oriented. Speech was normal with no dysarthria (Cerebellar or staccato speech). Other higher functions including memory were normal. Examination of cranial nerves including fundus was normal. No papillaedema. Motor system (no hypotonia), sensory system and reflexes were all normal. Cerebella/r examination reveals absence of hypotonia, Intention tremors. Ocular motor functions are normal. Finger nose test was also negative which shows that the cerebellum is normal. Other system examination including cardiovascular, respiratory and abdomen was normal

On intraoral examination, a diffuse swelling is seen on right buccal mucosa of size about 3cm x4cm seen 2cm away from corner of mouth extending into oropharynx involving pterygomandibular raphae anteroposteriorly, superioinferiorly upper vestibule into lower vestibular region, color of lesion is bluish red. On palpation lesion is non tender non compressible, firm in consistency with no evident pulsations. Diascopy was negative. Advised biochemical and serological tests including CBC, blood sugar and kidney function tests were normal. Test for human immunodeficiency virus was negative.

Based on clinical findings, provisional diagnosis given as vascular malformation and Differential Diagnosis as Hemangioma.

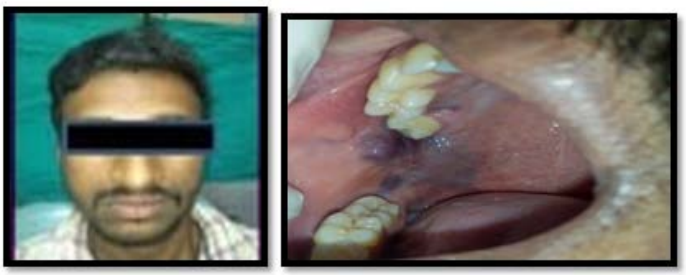

Fig. 1: Intra Oral Photograph

For further evalution Ultrasonography was taken which showed a multiple oval hypo echoic areas. For further confirmation MRI was advised. It showed a Multilobulated T2 Hyper intense transpatial lesion is noted in the lateral wall of oral cavity extending to retro maxillary lesion and premaxillary lesion approximately measuring 4.2cm x1.6cm.peg like cerebellar tonsil herniation measuring 5.0mm below the foramen magnum is noted with associated syrix from C2-D1.

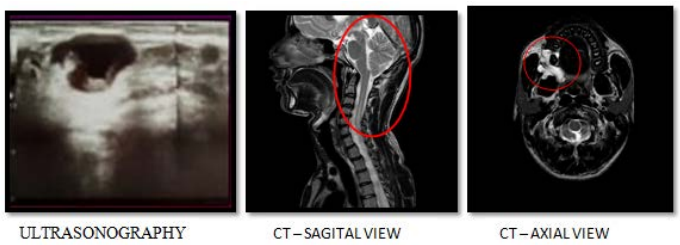

Figure 2

Correlating all clinical radiographic and laboratory investigations, case was diagnosed as ARNORLD CHIARI SYNDROM TYPE-I. Patient was shifted to Neurological evaluation. As the patient was asymptomatic completely, patient was advised continuous monitoring and evaluation.

Discussion

Chiari Malformation includes a large spectrum of anomalies of hindbrain formation which appear at different stages of development of the central nervous system.[1] It is a serious neurological disorder where the bottom part of brain, the cerebellum, descends out of skull and crowds the spinal cord putting pressure on both brain and spinal cord causing various symptoms. The symptoms are mainly because of structural defects in cerebellum, characterized by downward displacement of one or both cerebellar tonsils through foramen magnum. This occurs mainly because of failure of pontine flexure to form normally from 28-29th day of gestation which leads to elongation of brainstem. Normally growth of the cerebellum during the third month of intra uterine life causes the caudal vermis and choroid plexus to come under the tonsils. However if there is failure of this process, the vermis and choroid plexus remain in extra ventricular position. These structural deformities lead to blockage in the flow of cerebrospinal fluid, producing an embryological hydrocephalus. The secondary hydrocephalus causes further herniation of cerebellar tonsils into spinal canal and drags tentorium along with it, thus reducing the dimensions of posterior cranial fossa.[9]

These are series of hindbrain anomalies. Majority are congenital rarely can be acquired. The cases reported are very rare which is about in 1:1000 individuals.[10] Women are more common when compared to men. [11]

The patho physiology might occur in 2 ways.

- Direct compression of neurological structures against the surrounding foramen magnum and spinal canal.[12] In normal adults, posterior fossa comprise 27%of total intracranial space, while in adults of chiari malformation it is only 21% [13] In patients with the Chiari type I malformation, the bones of the skull base often are underdeveloped, which results in a reduced volume of the posterior fossa, the volume of which is inadequate to contain the entire cerebellum, thus cerebellar tonsils are displaced into the cervical canal
- Syringomyelia or syringobulbia development

✓ The obstruction of cerebrospinal fluid (CSF) outflow eventually results in syrinx development

✓ Fluid-filled cavities (syrinx) develop within the spinal cord or brainstem, resulting in neurologic symptoms as the cavity expands.[12]

Based on their clinical and radiographic manifestations, they are classified as

Type I

Downward displacement of cerebellar tonsils through foramen magnum is most often diagnosed in adulthood.[17] Most commonly observed Chiari malformation which is associated with tonsillar herniation through foramen magnum. It is often associated with syringomyelia but not hydrocephalus. Radiologically, Type I is described as tonsillar decent of 5 mm below foramen magnum. Type I Chiari malformation may be asymptomatic or present with mixture of cerebellar and pyramidal tract signs associated with dysfunctioning of lower cranial nerves.[18]

Type II

Commonly called Arnold-Chiari malformation characterized by descent of cerebellar tonsils, the inferior vermis and portion of cerebellar hemispheres into spinal canal along with elongation and displacement of brain stem and fourth ventricle. It is diagnosed in childhood and almost always associated with meningomyelocele and spina bifida.[19]

Type III

Herniation of cerebellum and brain stem through foramen magnum into the spinal cord. This is rare, but most serious form and causes severe neuro deficit like delayed milestones, seizures, ataxia, spasticity and other features common to Type I and II. [20]

Type IV

This involves undeveloped cerebellum sometimes associated with exposed part of skull or spinal cord. This is a rare type. [21]

Arnold chiari malformation type I is the most commomnly seen variant.It is a congenital variant often detected in adults and older children of age group 20s lates 30's incidentally. [14] Patient may always report occipital headache, lower cranial nerve palsy, sensorimotor abnormalities, Weakness and ataxia. As symptoms are not specific, it mostly may cause delay in diagnosis.[16] In this case, patient presented with headache weakness back pain with no other secondary symptoms.

About 1/3rd of cases with Type I malformation are asymptomatic, but commonly show their symptoms in third decade of life (25 - 30 years).[22] Many times patient with ACM Type 1 presents with complex clinical features. These highly variable clinical manifestations are due to compression of neural structures at cranio-cervical junction or obstruction of CSF flow. ACM Type I malformation is accompanied by syringomyelia in 25-75% of cases.[23,24] The commonest symptom is occipital headache which worsens with straining or coughing, but neck pain, ataxia, dysarthria, dysphagia, dissociated anesthesia have been reported. Oculomotor nerve palsy produces visual disturbances.[25,26] Syncopal episodes are described, but are rare Raised intracranial pressure causing papilloedema due to tonsillar herniation is reported in 2% of cases.[27] This patient was not showing features of raised intracranial pressure or papilloedema.

All these findings are non-specific, so differential diagnosis of ACM Type 1 from other diseases is difficult. The diagnosis of ACM Type I am usually late; till it is correctly diagnosed it can be mistaken as multiple sclerosis, other degenerative conditions. [28]

To make differential diagnosis, clinical examination in depth with skeptical approach is needed. Early and accurate diagnosis will help to refer these patients for better treatment before they develop further neurological complications.

All these findings are non-specific, so differential diagnosis of ACM Type 1 from other diseases is difficult. The diagnosis of ACM Type I is usually late; till it is correctly diagnosed it can be mistaken as multiple sclerosis, other degenerative conditions like Cerebrovascular accident due to vascular involvement in posterior cerebral circulation. Posterior cranial fossa tumour, eg. Cerebellopontine angle tumour. [28]

Diagnostic Criteria of Arnold-Chiari Malformation

Criteria to diagnose Arnold-Chiari malformation (ACM) differ according to study. In normal adults more than 3 mm descent of cerebellar tonsils through foramen magnum is rare; descent more than 5mm produces symptoms; so diagnostic criteria of ACM is descent of cerebellar tonsils more than 5mm below foramen magnum.[29,30] Ultrasonogrphy, neurological examination and imaging study of brain with computerized tomography (CT scan) and magnetic resonance imaging

(MRI) along with proper clinical history helps in revealing Arnold chiari syndrome.[21]

Management

The treatment of Arnold Chiari syndrome Type –I will differ depending upon condition of patient. Usually asymptomatic patients may not require treatment but they are continuously monitored. Symptomatic treatment cases can be treated both by medical and surgical management depending upon severity index. Usually symptomatic patients with ACM Type I malformation are recommended surgical treatment with craniocervical decompression, but not for asymptomatic individuals. Symptoms are resolved after surgical intervention, but there are not many studies to prove this hypothesis.[31]

Medical Management

Patients with Chiari malformation and who have no symptoms can be managed medically. Headaches and low neck pain can be treated with muscle relaxants, NSAIDs, and temporary use of a cervical collar. However, studies show that while a headache and nausea may improve, in many symptomatic patients there will be no improvement in gait with medical management. Close to 90% of patients with Chiari type I may remain asymptomatic even if they have syringomyelia.[32, 33]

Surgery is recommended for patients complaining of the classic pattern of symptoms and confirmed tonsillar herniation.

Better surgical results are seen when surgery is performed within 2 years of symptoms onset.

Surgical Techniques inluudes

Posterior fossa decompression: suboccipital craniotomy with or without the following:

- C1 and C2 laminectomy with or without [34]
- Dural opening/patching with or without
- Tonsilar cauterization

In general, patients who are asymptomatic without syringomyelia and who had an incidental diagnosis following an MRI study should not undergo surgery. This group can be managed with

Conclusions

CM constitutes an interesting clinical entity, where the embryological background still requires further studies. Recent imaging studies have increased the diagnostic ability of these anomalies enabling clinicians to adapt a conservative approach both in surgical and nonsurgical methods of management. The spectrum of clinical presentation in children differs significantly from that of adults or adolescents. While early surgical correction is recommended for symptomatic cases, incidental and asymptomatic malformations are best treated by watchful expectancy.

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