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Chiari I malformation presented post craniocervical trauma: A case report

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Abstract

Introduction: Chiari type I malformation, is a neuro-anatomical variation due to caudal herniation of the cerebellar tonsils through the foramen magnum and into the cervical spinal canal. It's a complication rarely presents following a cranio cervical trauma in young children. Symptoms presents as respiratory arrest, lower cranial nerve dysfunction and motor weakness. Supporting clinical findings on Neuro imaging studies disclosed Arnold Chiari I malformation.

Case Report: Reporting a case of 3-year old female child with history of cranio-cervical trauma, presenting with difficulty in deglutination and hoarseness of voice. Magnetic Resonance imaging of brain showed Chiari I malformation.

Conclusion: This case report emphasizes that Chiari type I malformation should be included in the differential diagnosis in children who presents with neurological manifestation following cranio-cervical injury. The neurological outcome depends on severity of herniation.

Keywords: Chiari I malformation, Cranio cervical trauma, Cranial nerve palsy, Cerebral tonsillar ectopia (CTE), Upright MRI, decompression surgery

1. Introduction

It was in 19th century, a pathologist named Hans Chiari first described congenital condition of cerebral tonsillar ectopia (CTE) since then it's named after him as Chiari malformation [1]. It is subdivided into 4 types. Chiari I malformation being the less severe form and may remain asymptomatic and diagnosis is usually and incidental finding on MRI.

Chiari Type I malformation is defined as caudal herniation of the cerebellar tonsils through the foramen magnum into spinal canal also called as cerebral tonsillar ectopia (CTE) [2]. Syringomyelia being the commonest complication following Chiari I malformation and may be associated with bony abnormalities at cranio cervical region [3].

Incidence of Chiari I malformation is 1 in 1000 births, with a slight female predominance with ratio of female to male is 1.3:1.

Chiari I malformation patients become symptomatic following cranio cervical trauma. Headache in occipital region being the most common presentation [4, 5]. Other complains includes motor weakness, altered sensation like paraesthesia, tingling, numbness, lower cranial nerve palsy. Brain stem and cerebellar dysfunction like respiratory arrest, cardiac arrest and abnormal gait, dysdiadochokinesia, nystagmus, slurred speech respectively can also present rarely [6].

The gold standard investigation for diagnosis of a Chiari I malformation is magnetic resonance imaging (MRI) characterised by cerebral tonsillar ectopia extending below foramen magnum [7, 8].

Approach to diagnostic criteria is though unfixed and considered significant when cerebral tonsils extend 5 mm or beyond below an arbitrary line drawn from anterior most point of foramen magnum (basion) to the posterior point of foramen magnum (opisthion) also known as cerebral tonsillar ectopia [9].

Mechanism of how Cranio cervical trauma contributes to Chiari I malformation or Cervical Tonsillar Ectopia is still obscured. At times there can be even downward displacement of cerebral tonsil. To differentiate between pre-existent (congenital) Chiari I malformation and Cranio cervical trauma triggered cerebral tonsillar ectopia, neuro imaging is done in standing position to rule out effect of gravity on cerebral tonsils known as Upright MRI [4].

Case report

A 3 year old healthy female child who is neurodevelopmentally normal presented in the Emergency department with a history of cranio cervical trauma while playing one day back with complains of difficulty in deglutination. CT Head was done which was normal. No history of loss of consciousness, seizures, or active bleeding. Birth history was normal. Parents gave a history of non-consanguineous marriage.

Then patient was admitted in PICU where she had 2 episodes of vomiting. Due to pooling of oral secretion and unable to deglutinate, prophylactically RT was inserted to avoid aspiration and continuous suctioning of oral secretion was done.

On examination Vitals parameters were within normal range. On CNS examination patient was conscious and oriented. Gag reflex was absent suggestive of Cranial nerve palsy. Sensory and Motor functions were normal. No sign of Brain stem or cerebral dysfunction.

Result of laboratory investigations included hemoglobin concentration of 9.1 g/L, hematocrit value of 28.9%, white blood cell count of 15500/mm^3 with 70% neutrophils, 26% lymphocytes, 1% eosinophil, 3% monocytes and platelet count was 3.98 lac/mm^3. Serum biochemical values, including C-reactive protein, electrolytes were in the normal range. Septic workup was negative.

I.V Fluids 0.45% DNS maintenance started. I.V. Antibiotics (Inj. Ceftriaxone and Inj. Amikacin) were started.

On day second Neurophysician opinion was taken suspecting Encephalitis with cranial nerve palsy, advised CSF study and MRI brain was done.

Inj. Methylprednisolone pulse therapy was given for 5 days.

CSF study showed total 4 cells/cu. mm, Lymphocytes 100%, RBC 0-1 cells/hpf, protein 46 mg/dl, glucose 102 mg/dl. CSF lactate was 5 mmol/L.

Magnetic Resonance Imaging (MRI) of Brain with contrast study is demonstrated in figure below:

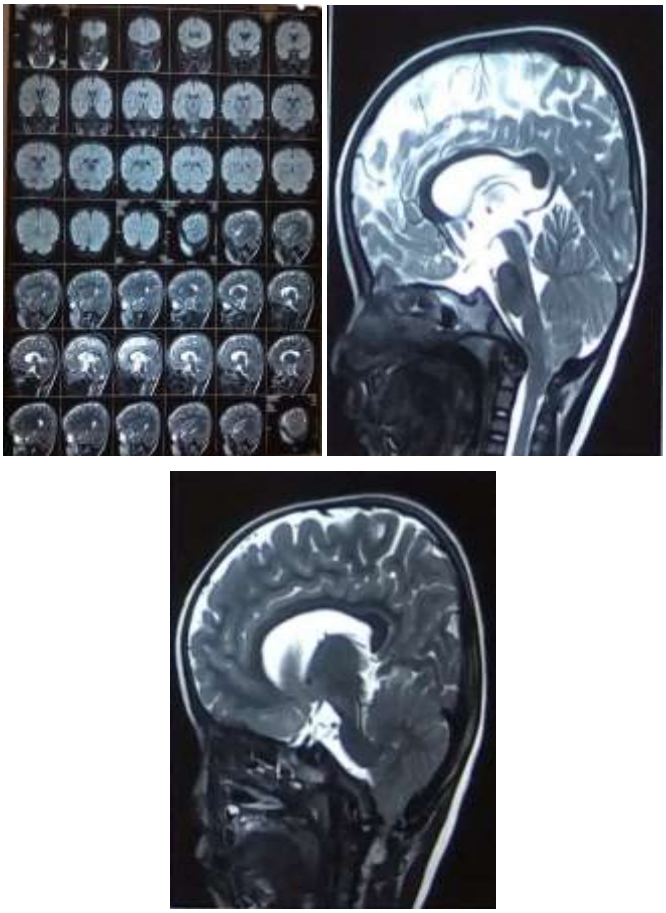

Fig 1: A Figure B Figure C

(Figure A, B, C): MRI scan of the brain and cervical spine revealed: FLAIR coronal images and T_2 sagittal images showing Tonsillar herniation on sagittal segment as hyperintense signal in lower part of medulla and upper part of cervical pons. These findings were suggestive of Chiari type I malformation.

On Day 3rd on basis of MRI report Neurosurgeon opinion was taken, advised Decompression surgery. Decompression surgery of foramen magnum was done by craniotomy and cervical laminectomy. Surgery was well tolerated by the patient.

On post operative Day 4th weak Gag Reflex appeared. RT feeding was started.

On subsequent days improvement noted in deglutination and was then able to accept solid food better than liquid. Then patient was transferred to rehabilitation Centre with normal vital parameters with proper parental counselling.

Discussion

Chiari I malformation also known as cerebral tonsillar ectopia defined as herniation of lower cerebral tonsils exceeding 5 mm through foramen magnum. Chiari Type I malformation can present in multiple ways and its incidence increases post cranio- cervical trauma in otherwise asymptomatic children [4, 5, 10].

Among symptomatic patients, headache in occipital region is the most common symptom followed by weakness, altered consciousness, sensory deficit, motor weakness, cranial nerve palsy and brain stem and cerebral dysfunction. Syringomyelia and raised intra cranial tension is the most common complication [3].

Neuro surgical procedure are the definitive management to treat Chiari I malformation, which includes decompression surgery and cervical laminectomy. Sub occipital craniotomy and duraplasty have also been proven to be effective in associated complications.

Various previous studies about Chiari I malformation being reported are as below:

Tomazek *et al*. reported a case of asymptomatic Chiari I malformation diagnosed on an autopsy, of a 3-year-old child who passed away after cranio cervical trauma [11].

Saez *et al*. in his study of 60 patients with Chiari I malformation found most common symptom presented includes occipital headache, cerebral dysfunction, lower cranial nerve palsy and motor weakness [12].

Vleck and Ito reported a case of a 2.5 year child with acute motor weakness post head injury, MRI revealed Chiari I malformation, treated with decompression surgery [13].

Pidcock *et al*. described syringomyelia as a common complication in a Chiari I malformation patient post head injury [14].

Our patient who was previously healthy, became symptomatic following cranio cervical trauma presented with lower cranial nerve palsy. The sudden appearance of the symptoms occurred due to increased herniation of cerebral tonsils post trauma, diagnosed on upright MRI as Chiari I malformation and treated with decompression surgery.

Thus a detail history and neurological examination should be done in any patient who comes to ED room with neurological manifestation following cranio cervical trauma and thoroughly investigation with neuro imaging like MRI and Chiari I malformation should be considered as differential diagnosis in all such cases.

Conclusion

Chiari I malformation is herniation of cerebral tonsils, becomes symptomatic following cranio-cervical trauma.

Presented is a peculiar case of Chiari I malformation with post traumatic lower cranial nerve palsy and symptoms resolved

following decompression surgery.

Through this case report, we would like to emphasize on the association of symptomatic Chiari I malformation following cranio-cervical trauma and if diagnosed early symptoms may resolve with decompression surgery with good neurodevelopmental outcome.

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