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Case Report

Patient with Spastic Ataxic Quadriparesis-Case of Arnold Chiari Type-1 Malformation

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Abstract:

A 22 years old patient presented with complaints of weakness in all four limbs and ataxia for 2 months. He had also history of slurring of speech and inclining to right side during walking. His gait was wide based and spastic ataxic. Patient was spastic quadriparetic with subtle right sided cerebellar sign and dissociated sensory loss in both upper limb and right Hemithorax. MRI of the brain and cervical spine revealed tonsillar herniation > 12 mm in cervical canal with large syrinx from cervical cord to thoracic region. Finally, the case was diagnosed as a Chiari Type-1 Malformation. Early diagnosis and intervention by Neurosurgeon actually halted the further syrinx extension and reduce neurological deficit with improvement of daily life activities.

Introduction

Chiari-1 Malformation as defined by tonsillar ectopia located below the foramina magnum, is being increasingly identified as a result of advance of MR imaging.[1] The malformation appears to be complex in its presentation, cause and natural history.[2]

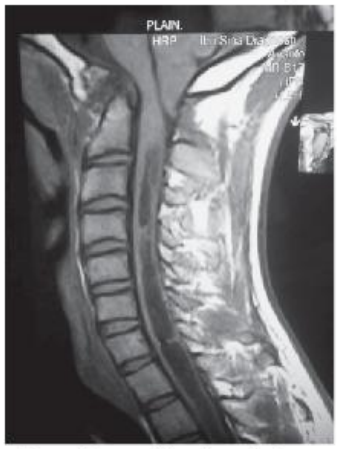

Figure-I: Showed tonsillar herniation and large syringohydromyelia.

This and associated anomalies were first described by Chiari (1981, 1896).[3] Arnold name is often attached to the syndrome but recently this malformation divided into Type-1 to IV- i.e. Chiari malformation, Types-I-IV, refer to spectrum of congenital hind brain abnormalities affecting the structural relationship between the cerebellum, brainstem, upper cervical cord and the bony canal base.[4,5] Numerous factors contribute to the development of such malformation. Currently there are over 20 different practiced operation directed at improving the abberrant CSF dynamics thought to be a major concept of malformation[2,6,7] but the pathogenesis is best explained by a molecular genetic analysis, Ectopic expression of a segmentation of gene in the rhombomeres explain not only the Chiari malformation but also the brainstem anomalies and defective basioccipital and supra occipital bone formation that result in a too small posterior fossa.[8]

Mostly Chiari Type-I shows the caudal displacement of Cerebellar tonsils below plane of foramina magnum (+ syringomyelia 20-75%).[9] Prompt clinical suspicion and early MR evaluation and neurosurgical intervention improve CSF dynamic and modify the disease process and disability.

Case report

Twenty-two years aged, normotensive non-diabetic, right handed person presented on 10th May-2015 with history of weakness in all four limbs (right sided weakness more than the left), difficulty in walking, imbalance and neck pain for two months. He also complained of intermittent stiffening of both lower limbs during walking and apnoea during sleep with occasional neck deviation (torticollis). He had also history of recurrent fall and inclining to right side during walking. His speech was slurred and tremor in both upper limbs. He did not give any history of fever, vomiting, head or spine injury or any alternation of

consciousness or convulsion. He did not give any history of the bladder and bowel involvement and no family history of such neurological problem. Personal history revealed no history of addiction or history of exposure. On examination, he looked ill and assume abnormal posture. Pulse was 88/min, BP-100/65 mmHg, Temperature was normal, and others general parameters were normal. Various skeleton abnormalities such as high arch palate, short neck and scoliosis were also seen. On systemic examination, all the systems seem to be normal except nervous system which revealed: -Patient was conscious and oriented, slurred-speech syllable by syllable, gait was spastic ataxic with wide stance and swaying, that increased by eye closure. All the cranial nerves were intact. Muscle power was reduced in all four limbs, grade 4 in Upper limb and grade-III in lower limbs. All DTR were increased in all 4 limbs, coordination was defective in both lower limbs and right upper limb. Romberg test was positive, and vibration was impaired in both lower limbs. Planter extensor in right side and Hoff-man sign positive in right upper limb. Bilateral horizontal nystagmus was seen in both eye and occasional downbeat nystagmus. Sensory examination revealed loss of pain and touch sensation in both upper limb and right half of the trunk. Fundoscopy revealed no abnormalities. Relevant investigations showed, T-11180, N-63%, HB-13.1 gm/dl, PBF-Non-specific morphology, RBS-9.2 mm/L, S. creatinine-0.90 mg/DL, S. electrolytes-No abnormalities, VDRL-Non-reactive, Vit-B_{12} assay-524 pg/ng, X-Ray-Skull and Cervical spine, Chest PA view revealed no abnormalities, ECG was normal. MRI of the brain revealed post contrast, T1W1 showed elongated pointed, low lying cerebellar tonsil which displaced inferiorly through the foramina magnum into upper cervical spinal canal. Tonsillar herniation in the spinal canal is more than 12 mm below the foramina magnum and MRI of cervical spine showed that enlargement of cervical cord with a hypointense central fluid filling cavity (syringohydromyelia) extending from the level of C_2 down to thoracic region. Above MRI features were suggestive of Chiari Malformation. We immediately started symptomatic treatment by pain killer, baclofen, gabapentin and referred the patient to Neurosurgeon for PF decompression and syringotomy with duroplasty. Operations were done and patient improved his gait and further follow-up revealed no progression of disease.

Discussion

Although Chiari malformation is still listed as a rare disease by the office of rare disease of the National Institute of Health, this categorization is based on outdated data from before the MRI era. With routine use of MRI, Chiari Malformation is discovered with increasing frequency. For Chiari-I, Prevalence rate of 0.1--.5% with a slight female predominance are suggested by recent study.[10] Based on analysis of familial aggregation, a genetic basis of chain has been suggested.[11] Recent study suggested linkage to chromosome 9 and 15.[12] In type 1 Chairi malformation (without maningocele or another sign of dyraphism), neurologic symptom may not develop until adolescent and adult life.[3] Our case presented after the age of 22 years. The symptoms are increased intracranial pressure mainly headache, Progressive Cerebellar ataxia, Progressive spastic quadriparesis, downbeating nystagmas, Syringomyelia.[3] In figure-1 this case showed the tonsillar herniation and large syrinx extending from cervical to thoracic region.

In another study[13] analyzed for 17 case of Chiari malformation, younger than 20 years of age. The initial symptoms were skeleton abnormalities (71%), Such as scoliosis (11 patient), Pes cavas (1 Patient), Pain or Numbness (24%), and motor weakness (6%). Frequently seen signs on admission were sensory difficulty (100%), Scoliosis (85%), muscle weakness (6.4%) Muscle atrophy (35%) and Lower cranial nerve (35%) The characteristic neurological findings were unilateral sensory with motor weakness (65%) with decreased or absent DTR on the same side.

Our case presented mainly difficulty in walking and gait abnormalities as spastic quadiparesis and cerebellar sign on both upper limbs. Dissociated sensory loss in both upper limb and right hemithorax. Some cases of syringomyelia are associated with trauma and tumour. Our case did not show any evidence of trauma or tumour. This patient also showed the sleep apnoea and neck deformity that may occur if lower brain stem is involved, which leads to difficulty in feeding and respiratory distress.[9] The most common differential diagnosis is Multiple Sclerosis, Lower brainstem stroke, foramina magnum tumour, SCD, basillar impression and related conditions were excluded by relevant clinical examinations and investigations. MRI of the spine and brain is the most useful and most widely used imaging study for diagnosis of Chiari malformation- in addition to detecting the anatomy of cervicocranial junction, it provides useful information about associated abnormalities such as syringomyelia and hydrocephalus.[14] Contrast enhanced MRI should be obtained to search for abnormal enhancement from an associated spinal Cord tumour.[15] Our case typically

showed the feature of Chiari Type-I and syringomyelia without spinal cord tumour. Preoperatively those with little or no neurological deficit, symptomatic primarily with pain, can expect an excellent outcome. A systemic review of decompressive surgery in adult for Chiarai malformation with syringomyelia revealed that syrinx may persist after surgery at an average rate of 6.7% (range 0-22%).[16]

Conclusion

It is generally accepted that if the causative pathology is identified and treated, the long-term improvement and resolution of syrinx is good. Prognosis is also largely depended on the prognosis of the primary pathologic that result in the syrinx. So, it is concluded that early clinical suspicion and diagnosis with neurosurgical intervention overall improve the disease course and reduced disability.

References

1. Barkovich AJ, Lippold, FJ, Sharmann JL, et.at. Significance of Cerebellar Tonsillar Position on MR AJNR 1976; 7:795-799.
2. Milhorat TH, Chum W, Trinnidad EM, et. al. Chiari I malformation defind clinical, radiographic, and genetic feature in 364 symptomatic patients Neuro Surgery 1999; 44:1005-1017.
3. Adam RD, Victor M, Rooper AH. Developmental disease of nervous system. In; Principle of Neurology 7th edn, MC grawwhill Book Company, New York 2001; 1064-65.
4. Greenberg MS. Chiari malformation Hand book of Neuro Surgery, New york; Theme; 2006; 6:103-109.
5. Abd-El Barr MM, Strong CI, Groff MW. Chiari malformation: Diagnosis, Treatment and Failures J neuro surg Sci 2014 Dec 58 (4); 215-21.
6. Heiss JD, Patronas N, Devroom HL, et al. Elucidation the Pathophysiology of Syningomyelia J Neuro Surg 1999; 91: 553-562.
7. Dyste GN, Meneres AH, Vangilder JC. Symptomatic Chiari malformation, An analysis of Presentation, management and long-term outcome J Neuro Surg 1989; 71:159-168.
8. Bradley WG. Developmental disorder of the Nervous system. In: Bradley WG, Fenicle GM, JANKOVIC J (eds) Neurology in clinical practice, 4th ed. Butter-worth-Heinmenn, A imprint of Elsevier, Philadelphia. 2004, 399, 591.
9. Huges E, Cros JH. Disorder of Central Nervous System development. In: Fowler TJ, Scadding JW (eds), Clinical Neurology 3rd ed, Arnold 2003, 431-32.
10. Speer MG, Enterline DS, Mehltreter L, Manmock P, Joseph J, Dickson M et.al. Chairi Type I malformation with or without sysngomyelia prevalance and genetics. J Genet Focus. 2003, 12: 297-311.
11. Speer MC, George Tm, Enter line DS, Frankling A, Wolperrt CM, Milhorat TH, A genetic hypothesis for Chiaria I malformation with or without syringo myelia. Neuro Surg Focus. 2000 March 15, 8(3): E 12.
12. Boyless Al, Enter line DS, Hammock PH, Sigel DG, Slifer SH, Mehltertter L et.al. Phenotypic defination for Chiari type I malformation Coupled with high density SNP genome screen shows significant evidence for linkage to region chromosome 9 and 15. Am J. Med Genet A. 2006 Dec 15; 140(24): 2776-85.
13. Toyohiko I, Youshiuohu I, Minoru A, Hiroshi A. Hydrosyringomyelia associated with a Chiari I malformation in Children and Adolescents. Neuro Surg 1990; 26(4): 591-97.
14. MC vige JW, Leonmdo J. Neuro Imaging and Clinical manfestation of Chiari type I (CMI), Curr Pain Headache Rep. 2015 June, 19(6): 18.
15. Hoser SL. Diseases of the spinal cord. In: Brawdnwald E, Faucias Kasper DL, Hauser SL, Longo DL, Jameson JL (eds)., Harison Principle of Internal Medicine. 15th Ed. MC Graw-Hill Book Company, New York, 2001; 2432.
16. Schuster JM, Zhang F, Norvell DC, Hermsyeeyn JT. Persistent recurent syringomyelia after Chiari decompression-Natural History-management Strategy: A Systemic review. Evid based spine care J. 2013 (Oct) 4(2): 116-125.