SMGroup

SM Journal of Case Reports

Case Report

Arnold Chiari Malformation Type I without Syringomyelia in a 24-Year-Old Woman Admitted With Dull Pain in Her Upper Limbs: Case Report

Manuel Bassanini[1], Joyce Jiménez[2]* and Pablo Dueñas[2]

[1]*Department of Neurology, Clinica Kennedy Hospital, Guayaquil City, Ecuador*
[2]*Faculty of Medicine, Santiago de Guayaquil Catholic University, Guayaquil City, Ecuador*

Article Information

Received date: Jan 25, 2017
Accepted date: Feb 17, 2017
Published date: Feb 22, 2017

***Corresponding author**

Joyce Jiménez, Faculty of Medicine, Santiago de Guayaquil Catholic University, Guayaquil City, Ecuador, Tel: 220-0906-1802; E-mail: joyce.jimenez@cu.ucsg.edu.ec

Keywords Arnold Chiari malformation type I; Foramen magnum decompression; Surgical treatment

Abstract

Report of a case of a young patient with progressive onset of Arnold Chiari malformation type I without syringomyelia who debuted with a dull pain in her right upper limb and was treated with foramen magnum decompression showing a remarkably improvement on her symptoms.

Objective: To report this case along with a literature review.

Methods: A 24-year-old woman with a year history of dull pain in herrightarm was referred to the clinic. After physical and radiographical examinations, she was diagnosed with Arnold Chiari malformation type I without syringomyelia. A foramen magnum decompression by the removal of the outer layer of the duramater was performed.

Conclusions: A case of symptomatic Arnold Chiari malformation type I without syringomyelia in ayoung woman was successfully treated with foramen magnum decompression by the removal of the outer layer of the dura mater.

Introduction

Chiari malformations are a group of defects associated with congenital caudal displacement of the cerebellum and brainstem [1]. Hans Chiari, an Austrian pathologist, was the first to give a detailed description of hindbrain malformations [1,2,3-7]. There are four main types, but type 1, called Chiari I, is the most common [5,6,8,9]. It has also been described types 0, 1.5 and 5 [10-12]. Chiari Malformation Type I (CM-1) is defined as the amygdalin herniation of at least 5 mm below the foramen magnum level [7,13]. Some theories have been proposed about CM-1 etiology such us the molecular genetic theory, growth abnormality theories, the crowding theory and Gardner's hydrodynamic theory, but none of them explain all the clinical features [4,14-17].There may be a genetic component in Chiari malformations. Boyles et al found significant evidence for a gene associated with CM-1 on chromosomes 9 and 15 and has been linked with an autosomal dominant inheritance with reduced penetrance [18]. Patients with CM-1 often present symptoms such as headache, cervical pain, cranial nerve palsy, ocular disturbances, nausea, vertigo, gait disturbance, cord motor and sensory abnormalities [19,7,8] . Occasionally, C1M can be associated with occult spinal dysraphism [20].

Case Report

In October, 2016, a 24-year-old woman was admitted to the hospital because of upper limb and cervico-occipital pain.

She was in excellent health until a year earlier when she started to experience a dull pain that started in the right hand which she attributed to spending a lot of time working in front of the computer.

The pain progressed little by little taking the entire right upper limb and with less intensity on the left upper limb, she also referred occipito-lumbar pain. After this, she went to the traumatologist. At the clinic, she presented swollenness and tenderness of the right upper limb; epicondylitis was diagnosed and treated with anti-inflammatory medication (Celecoxib 200 mg + B complex + chlorazox 250 mg + paracetamol 300 mg. She didn't get better. She tried treatments like ocean therapy and acupuncture, but the results were not satisfactory.

The pain continued to intensify, exacerbated by physical activity, accompanied by headache that began in the occipital area and extended to the lumbar spine, dizziness and vomiting.

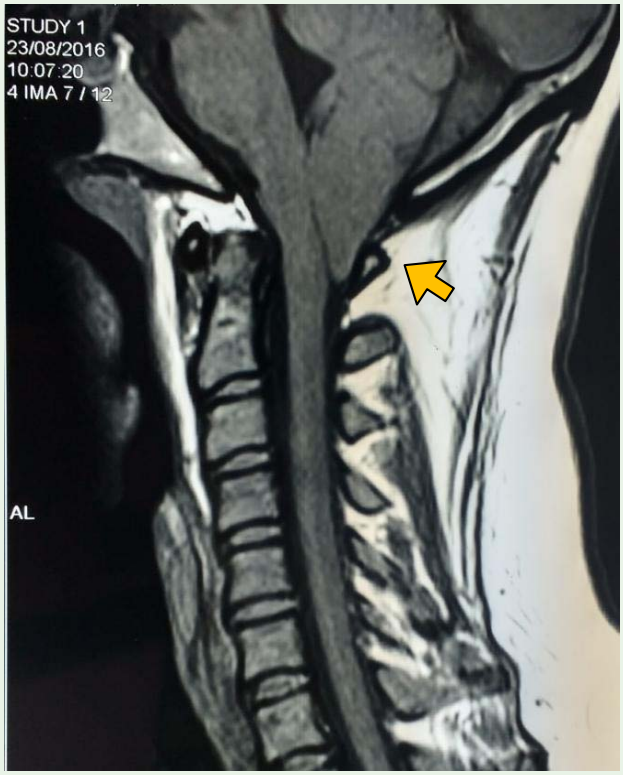

Figure 1: Sagittal T1-weighted MRI scan of the occipitocervical junction and the cervical spine. The compressed pointed cerebellar tonsils protrude through foramen magnum and fill the cisterna magna.

In August of this year she presented to the neurology consultation. On examination the patient appeared well. General physical examination on entry was negative. The lungs were clear, no murmur was heard and the examination of the abdomen was normal. In neurological examination the only thing that caught the attention was the presence of weakness 3/5 in the right upper limb and 4/5 in the left upper limb according to manual muscle testing. There was no presence of nystagmus or conjugate gaze. Hypoesthesia of right upper limb and tingling were noted. The tendon reflexes were preserved. Laboratory results were within normal limits. X-rays films of the cervical spine reported no abnormalities and the electrocardiogram was normal with a heart rate of 72 bpm.

It was decided to perform an MRI, in which a decrease of the cerebellar tonsils was observed through the foramen magnum (Figures1 and 2). She was diagnosed with Arnold Chiari Malformation type I without syringomyelia because of the characteristics in this images. Diprospan (betamethasone) was prescribed.

She presented relative improvement, but 3 weeks later, when exerting a physical effort, the pain was exacerbated in the left upper limb and in the periorbicular and occipito-lumbar area.

She was admitted to this hospital on October 19 of this year. In the general physical examination the patient was alert, oriented in time, space and person. She presented alteration of the coordination of voluntary movements in upper limbs when she performed the finger-to-nose maneuver. In the neurological exam she presented weakness 4/5 in the right upper limb and 4/5 in the upper left limb according to manual muscle testing. Gabapentin was given for pain, but did not improve. It was decided to perform an electromyography to rule out peripheral neuropathy, which showed no alterations.

Surgery is indicated and corresponding examinations are sent for preoperative assessment (blood count, creatinine, urea, prothrombin time, thromboplastin time, glucose, blood group and Rh factor), which are in normal ranges and showed no contraindications. It is assessed by cardiology with the Lee score which resulted in moderate risk II / IV, and suggesting antiembolic measures, adequate hydration and monitoring of vital signs.

Posterior fossa craniectomy is performed. The procedure consists of performing a suboccipitalcraniectomy associated with a laminectomy of all the cervical vertebrae necessary to decompress the tonsils and vermis in all their extension. The vertical incision of the occipital fascia was performed in the medial line with exposure of the posterior border of the foramen magnum to C1. Medial occipital craniectomy was performed with resection of the posterior border of the foramen magnum and the posterior arch of C1, durotomy with exposure and dissection of the thickened arachnoid to release adhesions. Therefore, athe effectiveness of decompression at the foramen magnum was immediately ascertained. Then a duraplasty with muscular fascia of the area and central suture of suspension to the superficial muscular planes.

At 1 month postoperative, the sensory disturbances in her upper limbs and the occipito-lumbar pain improved, vomiting and dizziness sensation disappeared. The intensity of headache was remarkably reduced. No postoperative complications occurred.

Control appointment is recommended every 30 days.

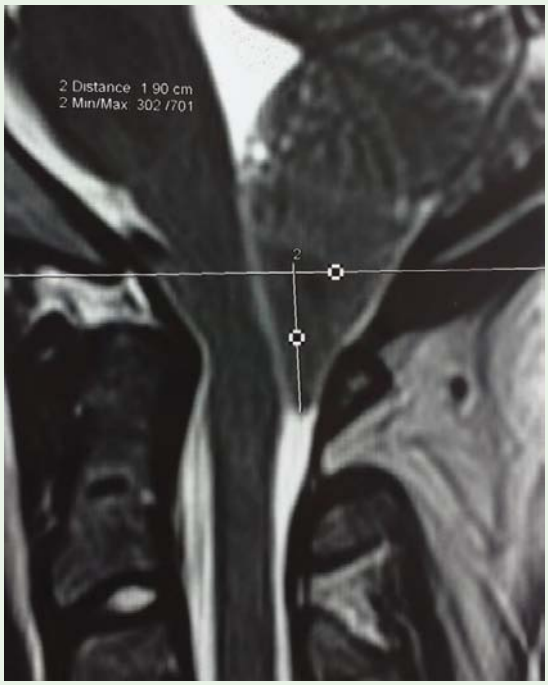

Figure 2: Sagittal T2-weighted MRI scan of the occipitocervical junction and cervical spine. It reveals a compressed, slender, peg-like herniated cerebellar tonsil of 1.90 cm, closely applied to the dorsal surface of the cervical cord.

Discussion and Conclusions

Chiari type I malformation is a condition described by downward displacement of the cerebellar tonsils of at least 5 mm through the foramen magnum into the spinal canal. In the general population, its prevalence is 0.6 – 0.9 % [21]. The etiology and pathogenesis of C1M remains a subject of debate. There's been proposed multiple theories, but none explains all the clinical features [4,14-17]. The molecular genetics theory mentions that C1M results from primary defects in the genetic programming of hindbrain segmentation and of growth of associated bone and cranial structures [16]. A growth abnormality theory stands that collision between caudally-directed cranial growth and rostrally-directed cervical growth is the main abnormality in C1M [15]. The crowding theory postulates that restricted growth of the posterior fossa causes compression of the neural tissue, which is then squeezed through the foramen magnum [14]. C1M can be due to either neuroectodermal or mesodermal anomalies. When isolated, it is thought to be of mesodermal origin. However, C1M that is associated with neurological disorders is thought to be of neuroectodermal origin [17].

It is possible that children born with this disorder may have inherited the gene. Boyles et al, supported that this condition could be linked with abnormalities in chromosomes 9 and 15 [18]. Mostrecently, Poretti and her colleagues [9] proposed that C1M is due to a deformation of the hindbrain, which is in contrast, not malformed. They named this condition Chiari type 1 Deformation because the downward displacement of the cerebellar tonsils may develop postnatally, increases or regress in a particular patient. Symptoms from CM I usually start in the 2nd or 3rd decade of life. Due to asymptomatic or unspecific clinical features, diagnosis of C1M is often made lately [2]. In the period of time, until C1M diagnosis, patients usually get diagnosis of demyelinating diseases, tumor and vascular disorders [2,22]. Previously, our current case was diagnosed as epicondylitis and given anti-inflammatory and analgesic treatment.

The unusual feature of this case is the insidious onset of a dull pain in her right upper extremity that the patient confused with working in front of the computer for hours. The most common presenting symptom in C1M is headache. The headache may be due to meningeal irritation and induced often with a Valsalva-inducedcomponent [19,23,9]. The pain is usually either occipital or nuchal in location. The pain is typically paroxysmal, but it may be dull and persistent [24]. Other manifestations are neck pain, vertigo, syncope, sleep apnea, drop attacks, cranial nerve palsy, ocular disturbances, nausea, gait disturbance, cord motor and sensory abnormalities [2,6]. The variety of presentation seen in C1M is due to dysfunction of cerebellum, brain stem and spinal cord. The presence of C1M and syringomyeliahas beenreported to be 25-75% [2,25].

The diagnostic procedure was a magnetic resonance imaging study with axial and sagittal images of the head and spinal cord. The presence of an ectopic location if the cerebellar tonsils and a normal position of the fourth ventricle in the absence of any supratentorial or posterior fossa abnormality are consistent with the diagnosis of C1M. Chiari type I malformation is based upon MRI studies of the posterior fossa because it is possible that the brain stem changes will not be revealed by the CT scans [18,7].

For C1M, the goals of surgery are to relieve the compression at cervicomedullary junction, to re-establish adequate CSF circulation and to achieve reduction in the syrinx size [6,26,13]. There have been reported a general improvement of symptoms and signs in 86.10% of patients using this surgical intervention [27]. Thanks to the successful surgical treatment using foramen magnum decompression, the patient presented a remarkable improvement on her clinical features.

Chiari type I malformation has been associated with Pierre Robin sequence, [28, 29] neurofibromatosis type 1, [30] Noonan syndrome [31,32] and Klsippel-Feil syndrome [7,24,33].

References

1. Adams RD, Schatzki R, Beecher Scoville W. The Arnold-Chiari Malformation - Diagnosis, Demonstration by Intraspinal Lipiodol and Successful Surgical Treatment. New Engl J Med. 225: 125-131.
2. Aytekin A, Parlak A, Develi S, Ekinci S, Parlak N. A Case of Arnold Chiari Malformation Type 1 Admitted with Hypoesthesia. SM J Case Rep. 2015; 1: 1001.
3. Di Genova C, Charitos S, Ba G, Viganò CA. Case Report: Atypical Psychotic Onset of Type I Arnold-Chiari Malformation. F1000Research. 2015; 4: 195.
4. Gardner, WJ. Hydrodynamic Mechanism of Syringomyelia: Its Relationship to Myelocele. J Neurol Neurosurg Psychiatry. 1965; 28: 247-259.
5. Hassan A, Yaseen S, Rashid M, Afza R, Kaur M, et al. Arnold-Chiari Malformation: Anatomical Variations and Latest Embryological Perspective. Review of Literature. International Journal of Contemporary Medical Research. 2016; 3: 1489-1491.
6. Hernández AV, Betancourt GM, García RM, Guriérrez JC, Guamuro RIG. Descompresión de la Fosa Posterior En Pacientes Con Malformación de Chiari I– Siringomielia. Revista Cubana de Neurología y Neurocirugía. 2014; 4: 43-48.
7. Khan AA, Bhatti SN, Khan G, Ahmed E, Aurangzeb A, et al. Clinical and Radiological Findings in Arnold Chiari Malformation. J Ayub Med Coll Abbottabad. 2010; 22: 75-78.
8. Osborn AG, Salzman KL, Jhaveri MD. Diagnostic Imaging: Brain. 2016; (3rd Edn.) Elsevier, Philadelphia, PA: Amirsys, USA.
9. Poretti A, Jallo GI, Garzon-Muvdi T, Ashmawy R, Huisman TA, Raybaud C. Chiari Type 1 Deformity in Children: Pathogenetic, Clinical, Neuroimaging, and Management Aspects. Neuropediatrics. 2016; 47: 293–307.
10. Kim IK, Wang KC, Kim IO, Cho BK. Chiari 1.5 malformation : an advanced form of Chiari I malformation. J Korean Neurosurg Soc. 2010; 48: 375-9.
11. Tubbs RS, Muhleman M, Loukas M, Oakes WJ. A new form of herniation: the Chiari V malformation. Childs Nerv Syst. 2012; 28: 305-307.
12. Tubbs RS, Elton S, Grabb P, Dockery SE, Bartolucci AA, et al. Analysis of the Posterior Fossa in Children with the Chiari 0 Malformation. Neurosurgery. 2001; 48: 1050-1055.
13. Salle F, Spagnuolo E, Aramburu I, Cabrera V, Jaume A, et al. Tratamiento Quirúrgico En Malformación de Chiari I En la Edad Pediátrica: Estudio Y Resultados En Serie de 57 Casos. Revista Argentina de Neurocirugia. 2013; 27: 51–58.
14. Marin-Padilla M, Marin-Padilla TM. Morphogenesis of Experimentally Induced Arnold-Chiari Malformation. Journal of the Neurological Sciences. 1981; 50: 29-55.
15. Matsuoka T, Ahlberg PE, Kessaris N, Iannarelli P, Dennehy U, et al. Neural crest origins of the neck and shoulder. Nature. 2005; 436: 347-355.
16. Sarnat HB. Disorders of segmentation of the neural tube: Chiari malformations. Handb Clin Neurol. 2008; 87:89-103.
17. Schijman E. History, anatomic forms, and pathogenesis of Chiari I malformations. Childs Nerv Syst. 2004; 20: 323-328.

18. Boyles AL, Enterline DS, Hammock PH, Siegel DG, Slifer SH, et al. Phenotypic definition of Chiari type I malformation coupled with high-density SNP genome screen shows significant evidence for linkage to regions on chromosomes 9 and 15. Am J Med Genet A. 2006; 140: 2776-2785.

19. Cobeñas R, Aguilar M, Aranguren J, Gallo JC, Espil G, et al. Headache… and something else? Neuroimaging in the study of headaches. Revista Argentina de Radiología 2016; 80: 192-203.

20. Caldarelli M, Di Rocco C. Diagnosis of Chiari I malformation and related syringomyelia: radiological and neurophysiological studies. Childs Nerv Syst. 2004; 20: 332-335.

21. Victorio MC, Khoury CK. Headache and Chiari I Malformation in Children and Adolescents. Semin Pediatr Neurol. 2016; 23: 35-39.

22. Russell GE, Bruce W, Rosa T. Arnold-Chiari Malformation. Optometry and Vision Science. 1992; 69: 242-247.

23. Miranda SP, Kristopher KT, Silberstein HJ. Acute Presentation of Chiari I Malformation with Hemiparesis in a Pediatric Patient. World Neurosurg. 2016; 85: 366.

24. Steinbok P. Clinical features of Chiari I malformations. Childs Nerv Syst. 2004; 20: 329-331.

25. Schneider B, Pravardhan B, Sara S. Arnold-Chiari 1 Malformation Type 1 with Syringohydromyelia Presenting as Acute Tetraparesis: A Case Report. J Spinal Cord Med. 2013; 36: 161-165.

26. Munshi I, Frim D, Stine-Reyes R, Weir BK, Hekmatpanah J, et al. Effects of Posterior Fossa Decompression with and Without Duraplasty on Chiari Malformation-Associated Hydromyelia. Neurosurgery. 2000; 46: 1384-1390.

27. Zhao JL, Li MH, Wang C, Meng W. A Systematic Review of Chiari I Malformation: Techniques and Outcomes. World Neurosurg. 2016; 88: 7-14.

28. Lee J, Hida K, Seki T, Kitamura J, Iwasaki Y. Pierre-Robin Syndrome Associated with Chiari Type I Malformation. Childs Nerv Syst. 2004; 20: 5-7.

29. Tubbs RS, Oakes WJ. Pierre-Robin syndrome associated with Chiari I malformation. Childs Nerv Syst. 2003; 19: 380-383.

30. Tubbs RS, Rutledge SL, Kosentka A, Bartolucci AA, Oakes WJ. Chiari I Malformation and Neurofibromatosis Type 1.Pediatr Neurol. 2004; 30: 278-280.

31. Holder-Espinasse, M, Winter RM. Type 1 Arnold-Chiari Malformation and Noonan Syndrome. A New Diagnostic Feature? Clin Dysmorphol. 2003; 12: 275.

32. Peiris A, Ball MJ. Chiari (type 1) malformation and syringomyelia in a patient with Noonan's syndrome. J Neurol Neurosurg Psychiatry. 1982; 45: 753-754.

33. Bresnan MJ, Shoukimas GM, Hedley-Whyte ET. Case 3-1987 - A 17-Year-Old Girl with Numbness of the Right Leg and the Recent Onset of Vertigo and Right-Sided Weakness. New Engl J Med 1987; 316: 150-157.