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Case Report

A Case of Arnold Chiari Malformation Type 1 Admitted with Hypoesthesia

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Abstract

Arnold Chiari malformation type I is a developmental pathology characterized by herniation of the cerebellar tonsils towards to the spinal canal through the foramen magnum. The herniation of the cerebellar tonsils can also compress or stretch anatomical structures in the brainstem, leading to various symptoms. This condition is rarely seen, due to common and non-specific complaints, it may be confused with other diseases, and patients may get misdiagnosis. In this article, a patient who applied to hospital with the complaint of hypoesthesia and diagnosed as Arnold Chiari malformation type 1 is reported.

Introduction

Arnold Chiari Malformation (ACM) type 1 is a clinical condition in which cerebellar tonsils extends to the upper cervical spinal canal through the foramen magnum. This form of ACM is considered to be adult type [1]. Older children or adults usually constitute patient population and patients are often asymptomatic [2]. Patients with ACM may present subtle and varying symptoms such as headache, extremity pain or weakness, vocal cord paralysis, vertigo, hypoesthesia. Non-specific symptoms may cause delay in correct diagnosis [3]. Symptoms seen in ACM type 1 are well described in previous studies. In this case, a patient with ACM type 1 who applied with hypoesthesia is presented.

Case

33 years old female patient was applied to hospital with sensory loss of right upper extremity. Patient mentioned this condition evolved in the last 10 years. It was learned from medical history that patient was experiencing headaches and loss of balance from time to time. The patient reported that she had received analgesic treatment for her complaints, with diagnosis of cluster headache. But patient also reported that she had not seen any benefit from treatment and had not applied to hospital until she got worsen. On physical examination, vital signs were within normal range, for her age, her weight was found to be normal. In neurological examination, positive rowsing test and hypoesthesia of right upper extremity was noticed, patient was not able to walk on a straight line and not able to make sequential movements. In examination of the eye muscles, bilateral difficulty in elevating bulbous oculi was found. Fundus examination revealed that there is no papilledema. Patient's complete blood count and biochemical tests were normal. Ferritin, folate and vitamin B12 levels were within normal limits. The patient underwent electroencephalography test. However, no pathological finding was observed in this test. Cranial and cervical Magnetic Resonance Imaging (MRI) was performed due to findings of neurological examination. In MRI, cerebellar tonsils were found to be protruded (herniated) caudally through the foramen magnum and mild brain stem compression was observed (Figure 1).

In addition, extensive dilatation of the central canal (hydromyelia) and bifid spinous process of 7th cervical vertebra was noticed, vertebral canal was found to be larger than normal (dural ectasia) (Figure 2).

Patient was diagnosed as ACM type 1 due to existing complaints, physical examination and cervical magnetic resonance imaging findings. The patient was referred to neurosurgery for decompression. The patient refused to have brain surgery due to complications might be seen after the procedure.

Discussion

With or without Syringomyelia, due to unspecific clinical findings, definitive diagnosis of symptomatic ACM is often made lately. In the period of time, until correct diagnosis, patients usually get diagnosis of multiple sclerosis, muscular dystrophy or other degenerative diseases [4]. Previously, current case was diagnosed as cluster headache and given analgesic treatment.

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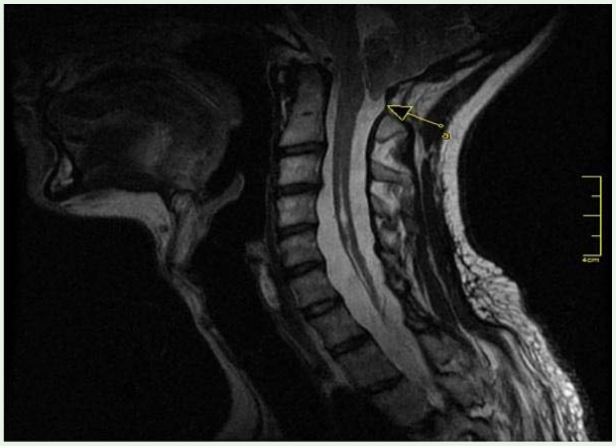

Figure 1: Cervical Magnetic Resonance Imaging. A: Cerebellar tonsils are herniated through the foramen magnum and the caudal brainstem compression is seen.

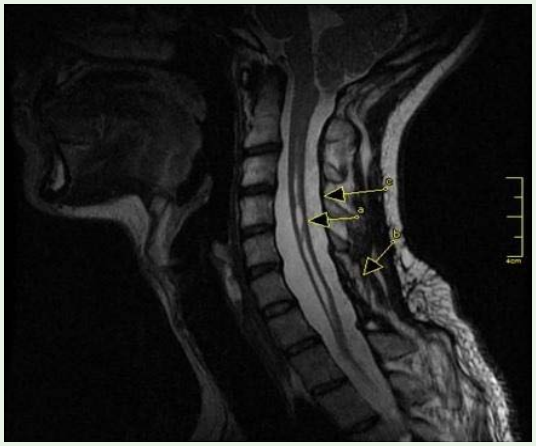

Figure 2: Cervical Magnetic Resonance Imaging. A: Spinal cord is thinner (atrophic) than normal and central dilatation is compatible with syringohydromyelia. B: Bifid spinous process of C7 vertebrae. C: Spinal canal is wider than normal (dural ectasia).

Four types of ACM were described previously. ACM type I was shown to be a developmental pathology characterized by herniation of cerebellar tonsils to vertebral canal through the foramen magnum [5]. Type I malformation, the most common variant, is often seen in adults and onset of symptoms is seen at range of 25-30 years [6]. Thirty percent of the cases are asymptomatic. Symptoms and signs are seen in a wide spectrum, typically progresses insidiously and slowly, severe neurological deficits can be seen in progression [7]. In this case, a patient who admitted for loss of sensation in upper extremity is presented but also eye movements found to be affected in physical examination. ACM type 1 is often present a complex clinical presentation. Symptoms are usually seen during early adulthood and occur gradually. Due to dysfunction of cetrebellum, brain stem and spinal cord, a highly variable manifestation is seen. Somehow, occasional exacerbations develop in some of the patients. Due to Valsalva maneuver, temporary increase in pressure at the posterior of the brain may lead to headaches. Symptoms and associated clinical findings of spinal cord syndrome depend on location and length of the syringohydromyelia and therefore may vary from patient to patient. The most common symptom is headache felt in the back of the head, but ataxia, dysarthria, dysphagia, neck pain, sensory differences have also been reported. Visual symptoms occur due to ocular motor nerve palsy [1,8-10]. Incidence of sensory loss in ACM type 1 has been reported to be 72 of 364 patients in a study [11].

Increased intracranial pressure caused by tonsillar herniation leads to papilledema. But this situation has been reported in only 2% of cases [11]. Findings of increased intracranial pressure and papilledema were not seen in this case.

Coexistence of ACM type 1 and syringohydromyelia have been reported to be 25-75% [4,12,13]. Cranium and spine abnormalities are seen in 30 to 50% of ACM type 1 patients. These abnormalities include basilar impression, atlanto-occipital fusion, atlantoaxial assimilation, Klippel-Feil deformity, cervical spina bifida occulta, and scoliosis [6].

Conclusion

ACM type 1 is usually presented with many different symptoms when diagnosed in adults. Symptoms of this malformation such as headache, loss of sensation and neck pain are common complaints. Due to common and unspecific symptoms, ACM Type 1 may be confused easily with other disorders. For this reason detailed examination and skeptical approach is important in differential diagnosis. We think that detailed examination of patients who presented with ongoing or increasing unspecific symptoms over the years, may contribute us to make accurate diagnosis.

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