



Chiari Malformation Type 1 Associated with Syringomyelia Which Clinical Symptoms Occuring during Tooth Extraction

CASE REPORT

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ABSTRACT

Chiari Malformation type 1 (CMI) is a congenital anomaly of the hindbrain, in which cerebellar tonsils herniate into the spinal canal. A 47-year-old male patient, who never had an additional disease up to that time sat on a dentist's chair for approximately half an hour with his neck extended for a tooth extraction. While getting up from the chair, the patient developed clinical symptoms. CMI may be undetected for years. Clinical symptoms may have appeared because of keeping the neck in the flexion or extended position for a very long duration of time.

Keywords: Chiari malformation, Chiari type 1, Syringomyelia

INTRODUCTION

Chiari malformation type1 (CMI) is a congenital disorder consisting of caudal herniation of the cerebellar tonsils exceeding 5 mm below the foramen magnum (1-5). Usually, dental ligaments, cervical nerve roots, IV ventricle, and brain stem are in normal positions (1, 2, 6). CMI is usually associated with syringomyelia. The onset of clinical symptoms generally occurs in the second or third decade. The diagnosis of CMI is made using neuro-imaging technique, and the preferred technique is magnetic resonance imaging (MRI) (1, 5). Clinical symptoms may either develop in a slow process through the clinical years or acutely because of trauma (2, 7, 8). If the patient shows any clinical symptoms, surgical treatment is recommended. Despite the fact that surgical treatment and procedures are still debatable, the recommended method is craniocervical decompression (6, 9-11).

CASE REPORT

A 47-year-old male patient who never had an additional disease up to that time sat on a dentist's chair with his head extended for a tooth extraction for almost half an hour. While getting up from the chair, the patient began to feel numb in the left part of his body, and this numbness increasingly spread to the right part of his body. Symptoms such as difficulty in swallowing, double vision, and ataxia began to appear. In the physical examination, it was found that in all the dermatomes in C3 and underneath, there was 1/5 of power loss in the right hand finger adduction; moreover, swallowing and pharyngeal reflex disappeared, DTR slightly increased, and double vision was determined while looking in the lateral direction.

The patient underwent cranial and cervical MRI scan. The patient showed tonsillar herniation up to C1, syrinx from obex to C7, and brain stem oedema (Figure 1a, b). The patient did not show hydrocephaly. The patient was diagnosed with CMI and syringomyelia.

The patient underwent craniocervical decompression surgery. Midline incision was performed on the skin. The posterior wall of the foramen magnum was opened for an approximately 2.5-cm-diameter craniectomy on the occipital bone. C1 posterior arch was removed till the length of 1.5 to 2 cm. The hard extradural ligaments at craniocervical junction and the outer layer of dura were removed. It was observed that the inner layer of dura was protruding. The movements of the tonsils were seen through the thinning dura, and BOS flow was observed using USG.

In the postoperative 3rd month control, the symptoms shown by the patient disappeared, and it was found that control MRI craniocervical decompression was adequate and that there was a significant decline in syringomyelia (Figure 2a, b).

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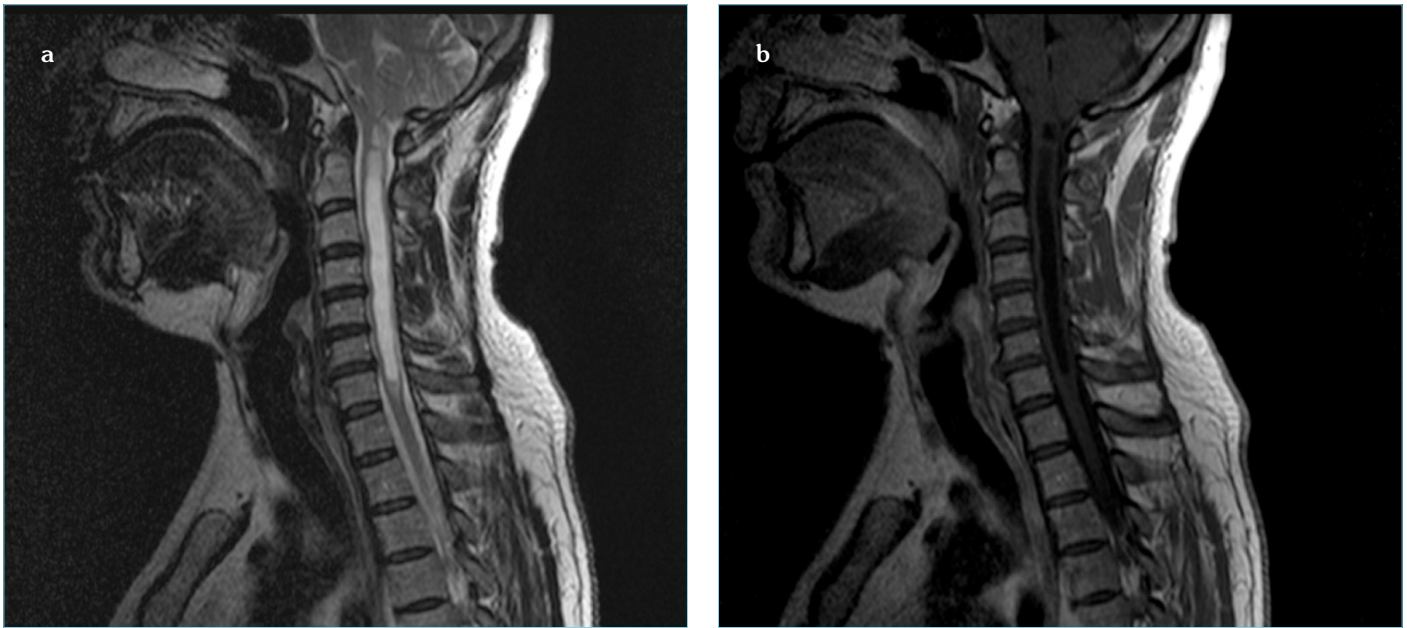


Figure 1. a, b. Preoperative T1 and T2 sagittal MRI shows tonsillar herniation up to C1 syrinx from obex to C7 and brain stem oedema

MRI: magnetic resonance imaging



Figure 2. a, b. Postoperative T1 and T2 sagittal MRI after 3 months show significant decline in syringomyelia

MRI: magnetic resonance imaging

DISCUSSION

CMI is defined as a cerebellar tonsil herniation of at least 5 mm through the foramen magnum into the cervical spinal canal (1-4). Syringomyelia is most commonly known as a complication because of CMI. Some cases show hydrocephaly (5). In addition, vertebral column and skull-based anomalies are also shown (kyphosis, scoliosis, basilar invagination, etc.) by the CMI cases (5). However, other brain anomalies are not seen (5, 6). CMI is detected in younger or middle ages (2, 6). The symptoms of CMI are generally suboccipital

head ache, diplopia, blurred vision, ocular disorders like photophobia, ataxia, dizziness, tinnitus, hearing disorders, muscle weakness (particularly in the upper extremity), paresthesia, hypo-hyperesthesia, and analgesia (1, 5).

Magnetic resonance imaging is the best diagnostic imaging technique for CMI and syringomyelia (1, 9). Computed tomography (CT), Brain-Stem Auditory Evoked Potential (BAEP), and Somatosensory Evoked Potential (SEP) are tests that help to diagnose, determine the surgery, and choose the treatment method (5).

Tonsillar herniation continuously compresses the adjacent cervical cord with severely edematous changes in the cord or brain stem (12). In general, patients with such serious long-term cervical cord compression would be continuously suffering from a series of symptoms and could not live a healthy life. However, our patient lived for years and incredibly without any clinical presentation (12, 13).

The generally acknowledged mechanism of sudden onset of the clinical symptoms of CMI in such situations is that the head trauma-induced neck hyperextension or other positional shift causes sudden pressure against the displaced tonsil by the bony edge of the foramen magnum (7, 12). Neck hyperextension or flexion at the time of trauma begins and ends in a very short time. However, in our case, no history of trauma or neck injury was reported. The neck stayed in the extended position on the dentist's chair for about 30 min and just after that the symptoms began to appear.

We are of the opinion that even though there was not any traumatic injury of the neck, tonsillitis due to extended neck position for a long duration of time led to a pressure on the brain stem, which caused edema in the brain stem and clinical symptoms appeared in the patient.

Patients with neurologic symptoms must be operated (6, 9-11). In spite of the fact that there are some discussions on the surgical procedure, craniocervical decompression method is the most frequently used one (6, 9-11).

The patient was operated in prone position and craniocervical decompression was performed. Hard extradural ligaments at craniocervical junction were cleaned. It was checked using USG, and CSF flow was shown. Duraplasty was not performed. It was observed that in the postoperative 3rd month control, the symptoms of the patient disappeared. Control MRI craniocervical decompression was sufficient and there was a significant decrease in syringomyelia (Figure 2a, b).

CONCLUSION

Chiari malformation type 1 symptoms generally appear after trauma or acute physical exercise. The neck movement in these situations is severe and short-term. However, this case has shown us that keep-

ing the neck in flexed or extended position for a long duration of time may lead to the appearance of the symptoms.

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