Presentation of Chiari I Malformation After Craniocervical Trauma: A Case Report

Kraniyoservikal Travma Sonrası Bulgu Veren Chiari I Malformasyonu: Olgu Sunumu

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Abstract: Reported is a unique case of Chiari I malformation with syringomyelia and medullar hemorrhage, which became symptomatic 4 days after a craniocervical trauma when the patient was doing neck exercises. The patient manifested respiratory arrest, tetraparesis, and persistent lower cranial nerve dysfunction. Magnetic resonance imaging studies revealed a Chiari I malformation with a large cervicothoracic syringomyelia defect and medullar hemorrhage. The patient underwent surgery but some symptoms persisted after the operation. Type I Chiari malformation should be included in the differential diagnosis of patients who present with extremity weakness (upper or lower), lower cranial nerve palsy, or respiratory arrest after trauma or neck exercises.

Key Words: Chiari I malformation, craniocervical trauma, medullar hemorrhage, syringomyelia

Özet: Kraniyoservikal travmadan 4 gün sonra, boyun egzersizleri uygulaması esnasında semptomatik hale gelen, Chiari I Malformasyonlu olgu sunulmaktadır. Egzersiz esnasında tetraparezi ve respiratuvar arrest gelişti. Radyolojik değerlendirilmesinde servikal syringomyeli ile birlikte olan Chiari I malformasyonu ve medüller kanama tespit edildi. Bu olgu; travma veya boyun egzersizleri sonrası ekstremite parezileri alt kraniyal sinir lezyonları ya da solunum sorunları gelişen olgularda Chiari Malformasyonlarının ayırıcı tanıda düşünülmesi gereğini ortaya koymaktadır.

Anahtar Sözcükler: Chiari I malformasyonu, kraniyoservikal travma, meduller kanama, syringomyeli

INTRODUCTION

Chiari described congenital anomalies of the craniocervical junction in 1891, dividing them into three groups (1). The association of syringomyelia with Chiari malformation is well known. The number of patients diagnosed with asymptomatic Chiari I malformation has risen with the advent of magnetic resonance imaging (MRI). In this article, we present a case of acute neurological deterioration and respiratory arrest in a patient with Chiari I malformation with syringomyelia and medullar hemorrhage. The symptoms developed 4 days after craniocervical trauma, when the patient was doing neck exercises. We discuss the case and review the literature. To our knowledge, this is the first report of a case of Chiari I malformation with medullar hemorrhage.

CASE REPORT

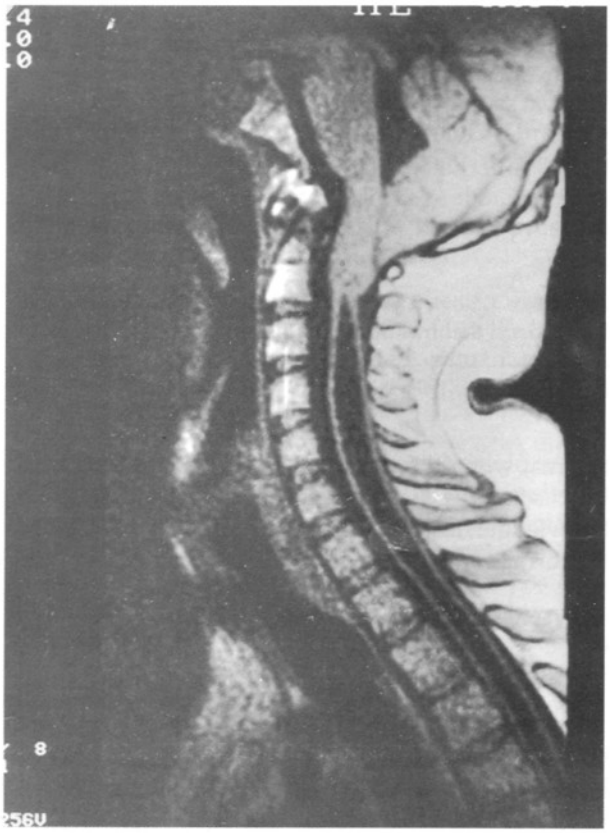
A previously healthy 20-year-old man was involved in a car accident, and was transported to the emergency room of a local hospital. His only complaint was neck pain. X-rays of the cervical spine and skull were normal. He was hospitalized for his neck pain, and began a physical therapy and rehabilitation

program. On the fourth day of the exercise program, the patient went into pulmonary arrest while he was doing neck exercises. He was intubated and transported to the intensive care unit (ICU) for mechanical ventilation. The patient was in serious condition. He was unconscious, had neither motor nor verbal responses, and his pupils were isocoric and myotic, with a very weak light reflex. There were no abnormal findings on repeat cervical radiographs. A tracheostomy was performed and mechanical ventilation was initiated. The patient was started on corticosteroids.

On his second day in the ICU, the patient was able to respond to commands. His pupils were isocoric and reactive to light, he had neither a gag reflex nor palate elevation, and he was moving all extremities at two-fifth's strength. Because the patient's spontaneous respiration was insufficient, mechanical ventilation was continued in synchronized intermittent mandatory ventilation mode.

By the third day, the patient's spontaneous breathing was adequate and mechanical ventilation was discontinued. MRI of his cervical and thoracic spine showed a type I. Chiari malformation with a hyperintense lesion in the medulla, which was interpreted as subacute hemorrhage. A large syringomyelia defect was also observed, extending from C2 to the lower thoracic segments. There was no connection between the fourth ventricle and the syrinx (Figures la and 1b). Surgery was scheduled.

Decompression of the foramen magnum region was achieved via suboccipital craniectomy, laminectomy of Cl and C2, and dural grafting. Postoperatively, the patient was able to breath spontaneously. His neurological examination showed some improvement in motor function. It was necessary to leave the tracheostomy cannula in place because of the need for frequent aspiration of the patient's secretions. Aspiration pneumonia developed, and the patient was treated with intravenous antibiotics. Pulmonary arrest occurred again, and mechanical ventilation was restarted. The day after he arrested for the second time, the patient began to breathe spontaneously and arterial blood gases were within normal limits. At that time, he was taken off mechanical ventilation, but was still unable to swallow. A gastrostomy tube was placed, and nutrition was administered via this route. A postoperative MRI taken 20 days after the operation revealed narrowing of the



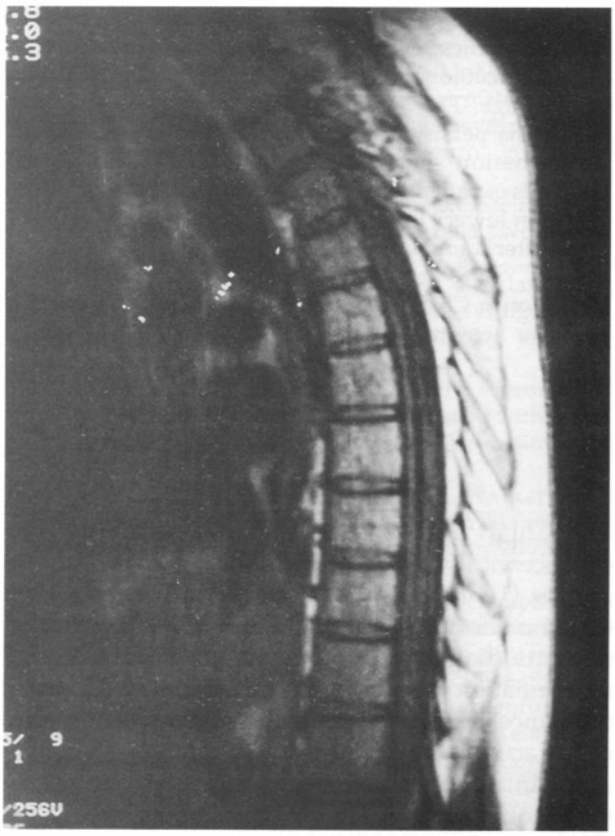


Figure 1: a,b,

cervical syrinx, an artificial cerebellomedullary cistern, and opening of the fourth ventricular outlet (Figure 2). The patient was transferred to a rehabilitation center.

DISCUSSION

Type I Chiari malformation has manifested in a variety of ways in otherwise healthy adults and adolescents (6,9,16,19). Saez et al. treated 60 adult patients with Chiari malformation and found that a foramen magnum compression syndrome, including headache, ataxia, dysphagia, and motor weakness, was the most common presentation. Paroxysmal intracranial hypertension, bulbar palsy, and central cord syndrome were also common (19).

In Chiari I malformation, the symptoms are generally present for several years before the diagnosis is made (6,9,16,19). Sudden onset of symptoms after trauma is unusual for this type of malformation, and there are few reports of this kind of scenario. Tomazek et al. reported the case of a 3-year-old child who died 48 hours after craniocervical trauma, and in whom type I Chiari malformation was diagnosed at autopsy (24). Vleck and Ito reported the case of a 2.5-year-old boy with acute paraparesis after a fall. The patient underwent surgery after being diagnosed with Chiari type I malformation (28). Scully et al. described a 17-year-old girl who developed hemihypesthesia, nystagmus, dysarthria, and tongue deviation 1 week after chiropractic manipulations. MRI revealed type I Chiari malformation and syringomyelia (20). Mampalam et al. described sudden development of symptoms in a patient with type I Chiari malformation who had sustained a closed head injury (9). Olivera and Dinh reported the case of a 28-year-old woman with Chiari I malformation and traumatic syringomyelia, which developed after closed head injury. The syringomyelia resolved spontaneously (14). Fish et al. reported two patients with Chiari I malformation who presented in respiratory arrest (2). Pidcock et al. described a patient with syringomyelia as a late-onset complication of head injury in the presence of an underlying Chiari I malformation (17). Nomura et al. reported a case of apnea associated with Chiari malformation in which MRI revealed medullar hemorrhage (12).

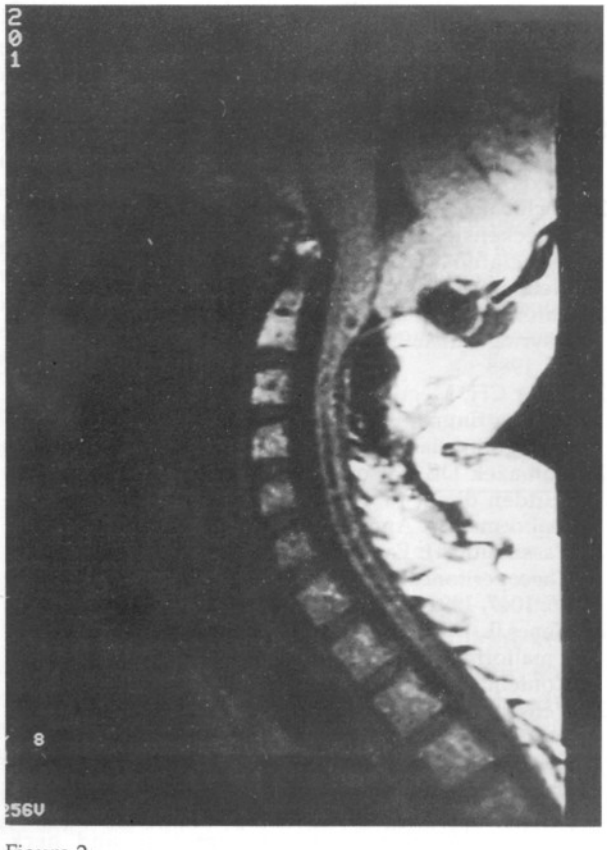


Figure 2:

Our patient became symptomatic 4 days after the trauma, while he was doing neck exercises. Pulmonary arrest occurred, the patient manifested lower cranial nerve dysfunction, and he was quadriparetic. It is possible that Chiari type I malformation and syringomyelia may have made him sensitive to hyperextension of the atlantooccipital junction (9,20). Although we do not have a baseline neurological examination of the patient, his family reported that he was asymptomatic before the trauma. The sudden appearance of symptoms may have been caused by rapid enlargement or formation of a cervicothoracic syringomyelia defect due to aggravation of a preexisting asymptomatic Chiari I malformation. It is difficult to pinpoint exactly when the medullar hemorrhage may have occurred. We presume that craniocervical trauma may have caused hemorrhage in the compressed medulla. Medullar hemorrhage may also have exacerbated the symptoms and caused the recurrent episodes of respiratory arrest. Our patient's symptoms persisted after the operation due to syringomyelia (13) and medullar hemorrhage.

Several different neurosurgical procedures have been used to treat Chiari I malformation with syringomyelia, including suboccipital craniectomy and duraplasty, alone or combined with any of the following: maintenance of an intact arachnoid; exposure or opening of the fourth ventricle; plugging of the obex with muscle; placement of a drainage tube from the lower portion of the Sylvian aqueduct, or from the fourth ventricle, to the spinal subarachnoid space;

ventriculoperitoneal shunting; terminal ventriculostomy; shunting from the syrinx to the subarachnoid space, pleura or peritoneum; intermittent percutaneous aspiration of the syrinx; and lumboperitoneal shunting (3-8,l0,ll,15,16,l8, 19,21-23,25-27).

The use of MRI has shown that treatment which eliminates the syrinx may not improve the clinical symptoms, presumably because of irreversible cord injury that may have occurred prior to treatment. Many patients respond best to simple bone decompression with duraplasty, leaving the arachnoid intact (6-8,10,13,21).

In conclusion we have presented a unique case of Chiari I malformation with posttraumatic syringomyelia and intramedullary hemorrhage. Chiari I malformation must be included in the differential diagnosis for patients who present with tetraparesis or paraparesis, lower cranial nerve dysfunction, recurrent respiratory arrest, or cerebellar signs after closed head injury or chiropractic manipulation.

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