İLERİ DÜZEYDE SEREBELLAR HERNİASYONU OLAN CHİARİ TİP I MALFORMASYONLU GEBE KADINLARIN DOĞUM ŞEKLİ NE OLMALIDIR: OLGU SUNUMU

What Should be the Delivery Mode in Pregnant Women with Chiari Type I Malformation who have Severe Cerebellar Herniation : A Case Report

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ÖZET

Chiari tip I malformasyonu; serebellar herniasyon ile karakterize magnetik resonans görüntüleme ile tanı konulabilen konjenital bir anomalidir. En sık görülen semptomlar; baş, boyun ve extremite ağrısı, özellikle üst ekstremitelerde görülen hipoestezi ve güçsüzlüktür. Chiari tip I malformasyonlu gebe kadınlar için uygun doğum ve anestezi yöntemi hala tartışmalıdır. Biz bu makalede genel anestezi altında sezeryan ile doğurttuğumuz Chiari tip I malformasyonlu, ileri derecede serebellar herniasyonu olan bir olguyu sunduk. Multidisipliner bir yaklaşımla bu hastalar için uygun doğum ve anestezi yöntemini tartışmayı amaçladık.

Anahtar kelimeler: *Chiari tip I malformasyonu; Gebelik; Doğum*

ABSTRACT

Chiari type I malformation which is characterized by cerebellar herniation is a congenital anomaly that can be diagnosed by magnetic resonance imaging. Most common symptoms are headache, neck and extremity pain, hypoesthesia and weakness especially in the upper extremities. The suitable mode of delivery and anesthesia for the pregnant patients with Chiari type I malformation is controversial. In this case report, we present a case of 37 weeks of gestation of a pregnant woman with Chiari type I malformation who had severe herniation of cerebellum and gave the birth by cesarean section with general anesthesia. We aimed to discuss about the suitable mode of delivery and anesthesia for each patient with Chiari type I malformation with multidisciplinary approach.

Key words: *Chiari type I malformation; Pregnancy; Delivery*

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INTRODUCTION

Chiari malformations were first described by Cleland in 1883 and classified by Hans Chiari in 1891, into four groups. Type I malformation is characterized by abnormaly shaped cerebellar tonsils which are displaced below the level of the foramen magnum with abnormal cerebrospinal fluid flow (1). Incidence of Chiari type I malformation ranges between 0.56% and 0.77% on magnetic resonance imaging (MRI) studies (2). It may manifest with headaches, neck pain, and mild coordination problems, but most often it is asymptomatic. Women with Chiari type I malformation are of particular concern because of the potential risk of increased intracranial pressure during pregnancy and delivery. There is limited information in the literature about the obstetric management in pregnant patients with Chiari I malformation. Some authors declare that these patients can safely deliver vaginally (3), on the other hand some authors believe that it would be more appropriate to prefer cesarean delivery because of the morbidity of potential of the vaginal delivery (4).

Also there is no consensus about the mode of anesthesia for the labor of these patients. Although some authors report in the opposite way (5), most of the authors declare that spinal anesthesia should be avoided in these patients, and epidural anesthesia should be made by an experienced anesthesiologist in order to avoid dural puncture which may initiate or aggravate the neurological symptoms of this condition (6).
In this case report we report the succesful use of general anesthesia for elective cesarean section in a woman with known Chiari type I anomaly. We discuss the anesthetic and the obstetric management of this case.

CASE REPORT

A 35-year-old woman, gravida 1 para 0 with clinically asymptomatic Chiari type I malformation was admitted to our delivery room because of the onset of her labor pains at 37th gestational week. Her neurological condition had been diagnosed after an MRI scan 4 years ago because of her shortness of neck and kyphoscolosis.
The sonographic examination revealed that the fetal biometry was compatible with her gestational week, and the amniotic fluid index was in normal limits. The nonstress test was reactive with irregular uterine contractions. Because of her known diagnosis of Chiari type I malformation, we decided to consult the patient to neurology, neurosurgery and anesthesiology in order to decide the appropriate mode of delivery and anesthesia. Neurological examination was considered to be in normal limits except for limited lateral gaze bilaterally. Magnetic resonance imaging which was done before the pregnancy reported that there was developmental fusion of the C2-C3 vertebral corpuses, kyphoscoliosis, dilatation of foramen magnum and almost 28,22 mm herniation of cerebellar tonsils through the foramen magnum. There was also cervical costal formation beginning at the 4th cervical vertebral level (Figure1).

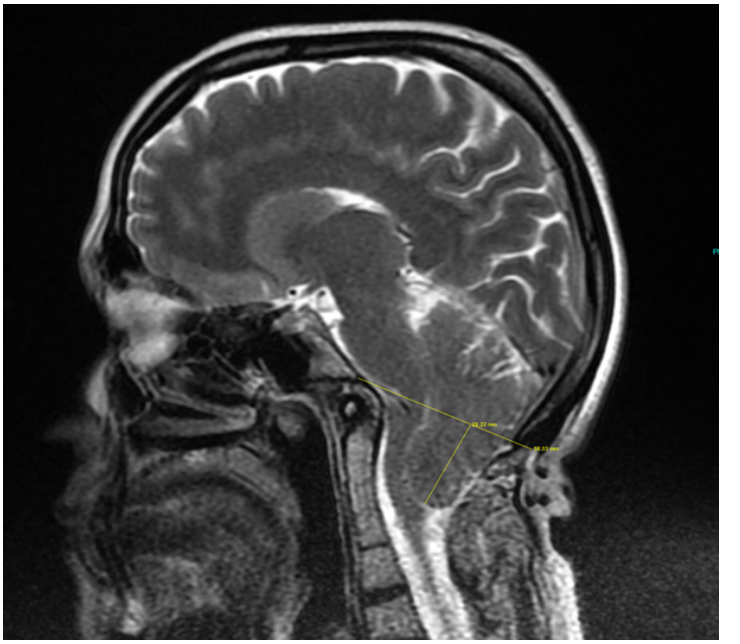

Figure 1. Sagital magnetic resonance image of Chiari type I malformation [28.2 mm tonsillar herniation from the cerebellum].

The neurosurgeon recommended avoiding vaginal delivery because of the diagnosis of Chiari type 1 malformation. So we decided to perform cesarean section. The preoperative anesthetic evaluation showed limited neck movement. The patient was informed of anticipated difficult airway entubation. The trial of epidural anesthesia had been unsuccessful and general anesthesia was performed. Tracheal intubation was considered to be easy. A female newborn was delivered with a weight of 2620 grams and a height of 46 centimeters, with an Apgar score of 8 in the first minute and 9 in the fifth minutes of delivery. She had diffuse nevus formations and bullous lesions throughout her entire body which were diagnosed as 'giant melanocytic nevi'. The baby was hospitalized in the neonatal care unit for follow up. The mother was discharged on the second postoperative day without any complications.

DISCUSSION

The Chiari malformations are a heterogenous group of congenital disorders that are defined by anatomic anomalies of the craniocervical junction, with downward displacement of the cerebellar structures. Type 1 is the most common type of Chiari malformations and usually diagnosed at adult age Associated abnormalities may include syringomyelia and kyphoscoliosis. Nearly one third of the patients are clinically asymptomatic and do not need any therapy (2). These patients are usually diagnosed incidentally when an imaging study is performed for another indication. On the other hand, it may be observed progressive neurologic deterioration and finally neurologic deficits in the latter two thirds of the patients with Chiari type 1 malformation. In this group, decompression with suboccipital craniectomy can be a surgical therapy option.

In Chiari type 1 malformation, the length of the cerebellar tonsil herniation below the foramen magnum can vary between 3 to 29 milimeters. Although the clinical severity of the symptoms does not correlate with the degree of herniation, patients with 12 mm herniation or more are usually symptomatic (1). We determined 28.2 mm herniation in our patient but she was neurologically asymptomatic except for limited lateral gaze bilaterally. She was diagnosed incidentally while investigating about kyphoscoliosis.

The suitable mode of delivery for the pregnant patients with Chiari type I malformation remains controversial. The alterations in the abdominal and thoracic pressures secondary to uterine contractions lead to an elevation of the cerebrospinal fluid pressure. It is unclear whether this elevation causes any harmful effects on maternal and fetal outcomes. Generally, cesarean section is prefered even if there is no obstetrical indication, because of the risk of cerebrospinal fluid pressure elevation (7). On the other hand there is no established evidence supporting that vaginal delivery should be contraindicated in these patients. Chantigian and et al. reported that 24 vaginal deliveries in 12 patients with Chiari type 1 malformation ended up without any complications (8). Newhouse and et al. have also reported that a pregnant woman with Chiari malformation 1 and also sickle cell anemia successfully delivered vaginally by vacuum extraction under epidural anesthesia (3). Nevertheless vaginal birth is not prefered for women with Chiari type 1 malformation, because of the risk of the detoriation of the herniation, especially at the 2nd stage of labor while straining. This thought is supported by the fact that headache, feeling of dizziness and other neurologic symptoms are exacerbated by Valsalva maneuvers (such as couhing, laughing, sneezing and etc) in these patients. In our patient there were no symptoms of elevated intracranial pressure, but the herniated part of the cerebellar structures was extremely wide (28.2mm). For this reason in order to avoid further herniation, we decided to perform cesarean section with the advise of neurosurgeon. The patient was informed and the consent form is signed by the patient. The suitable mode of anesthesia for the pregnant patients with Chiari type I malformation remains also controversial. Although there has been several case reports declairing that spinal anesthesia could be successfully performed in these patients (2), most authors recommend that spinal anesthesia should be avoided in these cases (9).

Hullander and et al reported that a 31 years old pregnant woman presented with paroxysmal headaches after spinal anesthesia for cesarean section. On the postoperative 6th day, because of the exacerbation of her headaches and development of visual disturbances, epidural blood patch had to be performed to this patient (10). Because of the high risk of difficult endotracheal intubation, epidural anesthesia can be an appropriate option for these patients. On the other hand, it should be taken into account that epidural anesthesia can cause intracranial pressure elevation and if incidentally dural puncture occurs, neurologic symptoms could be triggered (8). Due to the limitation of the neck movements and skeletal deformities, we preferred epidural anesthesia for our patient. After an unsuccessful attempt for epidural anesthesia, the operation is continued with general anesthesia. No surgical or anesthesiological complications have occured intra- or postoperatively.

As a result, no consensus have been reached until now about the appropriate mode of delivery and anesthesia in pregnant women with Chiari type 1 malformation. According to the available data in the literature, any interventional procedure which can cause an elevation of intracranial pressure could initiate neurologic symptoms or worsen the neurologic condition of the patient. For this reason it is acceptable that cesarean section is much more safer when compared to vaginal delivery. The advantages and disadvantages for all modes of delivery and anesthesia should be discussed with the patient and should be individualized with a multidiciplinary approach including the obstetrician, anesthesiologist and the neurosurgeon.

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