Arnold-Chiari malformations (ACM) are structural defects in the cerebellum usually caused by defects in the brain and in the spinal cord. These defects develop during fetal development and are classified into four types. Type I (ACM-I) is the most common observed type in children, and the lower part of the cerebellum - but not the brain stem - extends into the foramen magnum. Normally, only the spinal cord passes through this access. It may be associated with syringomyelia, hydrocephalus, tethered cord syndrome and spinal curvature due to scoliosis and kyphosis.

Type I ACM may manifest with headaches, neck pain, and mild coordination problems, but most often it is asymptomatic and discovered incidentally on brain or cervical spine MRI scan.

In type II Arnold-Chiari malformation, or “classic” Chiari malformation, both the cerebellum and the brain stem extend into the foramen magnum. This is usually only seen in children who were born with spina bifida.

Type III is rare and is the most serious form of Chiari malformation, because involves the protrusion or herniation of the cerebellum and brain stem through the foramen magnum and into the spinal cord. This usually causes severe neurological defects.

Type IV involves an incomplete or undeveloped cerebellum and sometimes is associated with exposed parts of the skull and spinal cord.

According to Fernandez et al. (1) ACM-I may be the result of an evolutionary anthropological imprint, caused by evolving species population that eventually met each other and mingled in the last 1.7 million years. Due to genetic mutations or a maternal diet without some nutrients, the indented bony space at the base of the skull
is abnormally small. As a result, pressure is placed on the cerebellum with possible blocks to the flow of the cerebrospinal fluid. Most ACM-I occur during fetal development, but much less commonly, it could occur later in life and recently it has been documented on MRI scan the regression of this malformation in childhood or later in life (2, 3).

Some people with ACM-I may have no symptoms, some others may have dizziness, muscle weakness, numbness, vision problems, headaches and problems with balance and coordination.

Scientists once believed that ACM-I occurred in 1 in every 1,000 births, but the increased use of diagnostic imaging techniques such as CT scans and MRI suggests that this condition may be more common. It affects women more often than men. The interest of ACM-I in obstetrics is linked to the low number of patients reported in the literature and to the doubt existing in the management of labor/delivery and anesthesia. The aim of this paper is to report a pregnant patient with ACM-I with syringomyelia that delivered in our hospital and to make a review of the literature on obstetric management of this syndrome.

Case report

A 30-year-old woman was sent to our department for delivery at 40 weeks of gestational age. At age 26, she reported tension-type headache, neck pain, and poor balance as symptoms. After neurological examinations a MRI of the brain showed cerebellar tonsils protruding through the foramen magnum for 13 mm (Figure 1) and the MRI of the thoracic spine demonstrated a syrinx from C3 to D11 (Figure 2). Family history was negative. The course of pregnancy was uneventful with normal development of the fetus which was in vertex presentation. The preoperative anesthetic evaluation showed a Mallampati test degree II, interincisor distance to 6 cm, and normal neck flexion-extension degree. Preoperatively a neurosurgeon was consulted, who agreed to general anesthesia, because in patient with ACM-I and syringomyelia it should be chosen to avoid any spinal maneuver that could increase intracerebral pressure or reduce intraspinal pressure with deterioration of neurological symptoms; also in this case the patient was asymptomatic and a prophylactic caesarean section under general anesthesia was considered as the method of delivery less likely to aggravate the syrinx, although the association of cervical spine disease, such as syringomyelia and pregnancy increases the risk of difficult intubation.

This kind of patients is considered at risk to increase intracranial pressure and progression of the disease during labor and delivery; for this reason the anesthetic team and the obstetric team decided to perform a general anesthesia with endotracheal intubation to avoid the worsening of neurological symptoms due to the spinal manipulation.

The patient and her family were informed of the risks of general anesthesia. She received 500 ml pre-operatively of Hartman’s solution 0,9%. Three-lead ECG monitor, bracelet for non-invasive measurement of blood pressure, pulse oximeter and neuromuscular blockade monitoring by TOF were applied. General anesthesia was induced with propofol (2 mg/kg) IV, fentanyl 100 mcg IV and rocuronium bromide 50 mg IV. After we proceeded to tube placement with neck in the neutral position. There were no difficulties at intubation. The correct placement of the tube was confirmed by identifying breath sounds and end-tidal CO₂ (ETCO₂). Then anesthesia was maintained with sevoflurane 2.0% end-tidal, and fentanyl (3 mcg/kg). The patient was stable for all the time of surgery. A healthy baby with Apgar scores at 1 and 5 min of 9/9 was delivered one minute after skin incision. Intravenous short term antibiotic prophylaxis was administered. At the time of extubating an exaggerated response to rocuronium bromide was noticed. It was necessary to administer atropine/intrastig-
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Discussion

Chiari I malformation is typically present in young adults and is the most common cause of syringomyelia (4). The duration from onset of symptoms to diagnosis is usually around 3 to 7 years (5). The earliest symptoms are often headache, neck or arm pain or paraesthesia that runs down to upper extremities worsened by straightening or coughing. Primary symptoms are tussive headache, visual changes, syncope muscle weakness, sensory deficit, dysphagia, dysarthria and ataxia. Secondary symptoms and signs may be papilledema, hyperreflexia, hyporeflexia, positive Babinski’s and Romberg’s signs, nystagmus, seizures, tremor, speech delay, absent gag reflex, and hoarseness. Symptoms linked to acute myelodysplasia may be present. Drop attacks, and apneic spells with abnormal extensor posturing may be unpredictable potentially harmful paroxysmal events due to compression. Tetrapsis is a very uncommon complication (6).

Dissociated sensory loss over the neck and arms is characteristic when a syring is present, because the crossing spinothalamic fibers tract carrying pain and temperature sensation are most affected, whereas posterior column sensory fibers are spared (5). Hand and arms weakness develops in the lower limbs with further expansion of the cervical syring. Neurological findings depend on the structure involved and on the presence of the syring, which is often noted at the C4-C6 bone levels (4) and may extend into the brainstem and may lead to lower cranial, cerebellar and even respiratory symptoms (4, 5). Although most patients are asymptomatic, symptoms may take to rapid deterioration and, rarely, may be atypical such as recurrent syncope and seizures (7).

For these reasons neurological examination should be requested. The diagnostic method of choice is the T1 weighted sagittal imaging MRI scan, in which the tonsillar herniation is better seen (4). The syring most commonly occurs in the cervical spinal cord, but may occur in the thoracic or lumbar spinal cord (8). Worsening scoliosis is common particularly in childhood ACM-I malformation with syring but does not impute a set of symptoms, nor does it impute a prognosis or surgical treatment (8, 9), which depends on clinical conditions. Differential diagnosis for ACM-type I with syring associated includes spinal muscular atrophy multiple sclerosis, spinocerebellar atrophy, arterovenous malformation, and spinal cord or brain stem tumors.

The evaluation of the neurosurgeon is important because:
- clinical manifestation of ACM-I may be related to cerebrospinal fluid disturbance and direct compression of nervous tissue;
- posterior fossa reconstruction is mandatory in patients with progressive symptomatic/signs, such as hydrocephalus, but not in patients who are asymptomatic or those with stable and tolerable symptoms. The presence of cough headaches is a significant negative predictor of concomitant symptoms improvement;
- it is possible a sudden death in AMC-I with concurrent hydrocephalus; especially when hydrocephalus is active. This conditions should be treated promptly. Opioids should be avoided in these patients;
- syringomyelia is a progressive myelopathy with cystic degeneration into the spinal cord, which may cause severe neurological deficits and the patient should undergo to adequate clinical and radiological follow-up;
- the neurosurgeon has to be called if there is the presence of neurologic symptoms every time during or after delivery;
- delivery has to be performed in a hospital in which is available a neurosurgical unit, also to plan periodic check-up of the patient.

The obstetric and anesthetic management of women with ACM-I remains problematic because of paucity of cases, and no uniform recommendations regarding mode of delivery (vaginal vs abdominal) and type of anesthesia (regional vs regional). Vaginal delivery has been performed successfully in some cases and usually is operative with vacuum extractor to avoid maternal voluntary expulsive efforts under epidural anesthesia, especially in the second stage of delivery (10). Spinal anesthesia has been safely adopted in most of the surgically decompressed cases of ACM-I (11).

Clinic

Anesthetic alerts in women with type I ACM are related to a possible increase of cerebro-spinal liquor (CSF) pressure associated with pregnancy and labor, or to a possible decrease of spinal pressure related to a loss of liquor after spinal anesthesia or accidental dural puncture during peridural performance, with an increase of differential pressure between CSF pressure above and below the foramen magnum. This increased differential pressure may lead to further descending of the tonsils through the fo-
ramen magnum with compression of the brainstem. Spinal neuroaxial anesthesia performed in mothers who have uncorrected type I ACM and syringomyelia may be complicated by neurological worsening, persistent headache and nystagmus. Signs and symptoms may develop up to 2 weeks after a dural puncture. On the other hand, there are reports of successful neuroaxial spinal anesthesia in women with Arnold-Chiari type I malformation. In some cases, neuroaxial spinal anesthesia has been performed after surgical correction of the malformation. General anesthesia, avoiding CSF pressure fluctuation due to neuraxial anesthesia, may be the best choice in this patient, however, each anesthetic plan has risks, and there isn’t an only strategy of management for all the patients in the absence of gold standard in the literature. General anesthesia was chosen in our patient with ACM-I and syringomyelia to avoid CSF pressure fluctuation.

Patients with syringomyelia may be also sensitive to non-depolarizing neuromuscular block due to muscle wasting, atrophy, and hyperkalemic response to succinylcholine administration (12). In our case we had block of the feet muscle an hour after the last administration of rocuronium dose. A correct dosing of non-depolarizing muscle relaxants and use of a neuromuscular twitch monitor (TOF in this case) as a guide may be an alternative. In addition, muscle relaxant reversal at the end of the case is recommended. It must be recognized that neostigmine could fail to reverse profound neuromuscular block in pregnant and post-partum patients. In this case we administered MIX with intrastigmine and atropine to resolve feet block and we assisted to a resolution of muscle blockage after 20 minutes with 4/4 TOF and 97% ratio.

Tarasnakar et al. (13) describes a pregnant woman who underwent to decompression of foramen magnum with removal of posterior arch of first and second cervical vertebrae and duroplasty with gelatin path, under general anesthesia; at term of pregnancy she underwent to elective cesarean section with spinal anesthesia. Only the pregnant with spinal anesthesia performed in mothers who have uncorrected type I, syringomyelia and cervical ciphosis.

It is important to evaluate the malformation with a competent neurological and radiological examination. The course of pregnancy is usually normal and symptoms may rarely appear. Some complications are related to local anesthesia, so it may be safe for the patient to plan delivery in a center with the availability of neurosurgery unit in case of emergency.

The case we presented has a syringomyelia extended between C3 and D11, and it represented a really rare case to treat; it was useful to evaluate the anesthetic risks related to the location and extent of syringomyelia and the natural history at follow-up. According to most AA, spinal anesthesia should be avoided due to inherent high intracranial pressure, but in scientific literature some cases of caesarean section with spinal anesthesia without any complication are described (15). More recently Kuczkoeski (17) described a newly diagnosed ACM-I (first trimester of pregnancy) and mildly symptomatic, who underwent to elective cesarean section at 37 week of gestation under single-dose spinal anesthesia without exacerbation of preexisting maternal symptoms or any other complications. However, these are case reports from which it is not possible to extract generalizations about patient selection and safety of spinal anesthesia. It should be noted that spinal anesthesia was used successfully for elective caesarean section after surgical decompression of the ACM-I (13, 18).

Declaration of interest

The Authors declare that there is no conflict of interest regarding the publication of this manuscript.

References


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