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Case Report

Congenital Spastic Paraplegia with Neobladder, Pregnancy and Cesarean Section - A Case Report

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Abstract

Spinal cord injury (SCI) in women requires a multidisciplinary approach to monitor their health. Although rare, pregnancies among women with SCI are on. Hereditary spastic paraplegias represent a set of rare neurodegenerative diseases on a genetic basis and are characterized by progressive spasticity and hyperreflexia of the lower limbs. Hypertonic urinary disorders, deep sensitivity of the lower limbs and, occasionally, joint proprioception are also often associated. The most frequent complications during pregnancy are urinary tract infections/pyelonephritis; venous thrombosis/pulmonary embolism, preterm rupture of membranes, and preterm/obstructed labor.

ABBREVIATIONS

SCI: Spinal Cord Injury; MAP: Medically Assisted Procreation; MR: Magnetic Resonance; AH: Autonomic Hyperreflexia; IUGR: Intrauterine Growth Restriction; PI-UA: Pulsatility Index – Umbilical Artery

INTRODUCTION

Spinal cord injury (SCI) in women requires a multidisciplinary approach to monitor their health and for the necessary care related to pregnancy and childbirth [1,2]. SCI does not necessarily decrease a woman's desire for pregnancy [3,4]. Although rare, pregnancies among women with SCI are on the rise and generally have favorable outcomes. However, various factors should be considered during pregnancy and childbirth: urological complications, the risk of thromboembolism and of autonomic dysfunction. The most frequent complications during pregnancy are urinary tract infections/pyelonephritis, venous thrombosis/pulmonary embolism, preterm rupture of membranes, and preterm/obstructed labor [5]. Hereditary spastic paraplegias represent a set of rare neurodegenerative diseases on a genetic basis and are characterized by progressive spasticity and hyperreflexia of the lower limbs [6]. From a clinical point of view, there are two forms: 'pure' and 'complicated'. Pure forms are characterized by spasticity and weakness in the lower limbs which progress slowly. Hypertonic urinary disorders, deep sensitivity of the lower limbs and, occasionally, joint proprioception are also often associated. Complicated forms also present neurological and non-neurological diseases. Spastic paraplegias can be inherited as an autosomal dominant, autosomal recessive or X-linked recessive trait, and there are multiple dominant and recessive variants. Treatment is symptomatic (muscle-relaxant drugs, functional rehabilitation). In some studies in order to reduce spasticity has been described therapy by intramuscular injection of botulinum toxin into the muscles of adults and children affected by spasticity [7,8].

CASE PRESENTATION

We present the case of a 38-year-old woman with congenital spastic paraplegia of uncertain etiology. The patient was born from elective cesarean section for previous maternal cesarean section, with neonatal weight of 2,600 Kg. The mother reports the physiological course of pregnancy and denies taking drugs. From birth there was a strong motor impairment of the lower limbs which appeared to be hypotonic. No special checks were carried out until the age of 14, when an MRI was performed which showed a medullary cord reduced in thickness at the level of D12-L1 with marked reduction of the surrounding liquor spaces. The patient also showed a severe neurogenic bladder dysfunction (neuropathic bladder with detrusor hyperreflexia and detrusor-sphincter dysynergy) with complete urinary incontinence. Continent urinary diversion was scheduled. The enlargement cystoplasty, bladder neck closure and cutaneous appendectomy according to Mitrofanoff were performed at the age of 17. In 2007 and again in 2009, due to the formation of bladder stones (vesical calculi); the patient underwent

endoscopic crushing and removal surgery. All stone fragments were removed. The patient regularly performed bladder self-catheterization. A genetic investigation has never been carried out in the diagnostic process, so it cannot be excluded the case of a hereditary form.

At the age of 30, chronic hypertension was diagnosed and treated with drug therapy with perindopril + Indapamide 2.5 mg one tablet a day until. The patient undertook the medically assisted Procreation (MAP) pathway for which the drug was replaced by Methyldopa 500 mg.

The patient II gravida 0 para (1 previous spontaneous abortion, pregnancy obtained by MAP), comes to our observation during her second pregnancy, obtained by medically assisted homologous procreation (ICSI). Her partner was also paraplegic because of an accident at work. During pregnancy several episodes of urinary infection (at least 3 episodes) have been reported and treated with antibiotic therapy. Due to immobility, 6000 IU heparin therapy was performed throughout the pregnancy to reduce venous thrombosis/pulmonary embolism.

At the 34th week of pregnancy, an upper and lower abdomen MRI was performed in order to study the relationship between the bladder and uterus for the planning of surgical access. From the Magnetic Resonance (MR) of the upper and lower abdomen, it was shown that the neobladder was located in the middle of the pelvic excavation. In the bladder lumen the end of the catheter previously positioned via the umbilical way was recognizable. The catheter was followed throughout its course in the subfascial area up to the para-umbilical site where its path became horizontal for about 5 cm reaching the navel from which it emerged (Figures 1,2).

Pregnancy was complicated by pre-eclampsia (arterial hypertension refractory to drug therapy, rise in uric acid and changes in coagulation factors). In addition, there is also IUGR with increase in PI-UA and brain sparing. For this reason a cesarean section at the 35 week of pregnancy, after disodium betamethasone phosphate cycle, was performed. The female infant weighed 1675 gr at birth, 42 cm in length, Apgar 8'-9'. During caesarean section a myomectomy of a pedunculated myoma was performed. The conditions of the baby at birth were optimal. The patient was discharged without maternal complications 72 hours after delivery.

DISCUSSION

We presented a quite complex case in the pre-conception, prenatal and perinatal period for maternal comorbidities and disability of both parents, congenital for the mother, acquired for the father with infertility. Maternal and neonatal outcome, however, was positive.

About 2000 - 3000 women in childbearing age suffer spinal cord injury each year, especially following trauma [9,10]. By itself, the fertility is not compromised in women with spinal cord injury: menstrual irregularities in the first 3-9 months and subsequent return to the situation pre damage with a maximum of 36% post trauma pregnancy [11-13].

In our case the etiology of spastic paraplegia seems to be unknown, even if genetic investigations have not been performed. Only prenatal data available is maternal alloimmunization, so our little patient underwent phototherapy for neonatal jaundice: perinatal suffering has not been documented and there are no studies correlating neonatal hemolysis to paraplegia.

In preconceptional management, in addition to information on present and potential problems, it is also necessary to consider the need for drugs, such as anticonvulsants, possible to take during pregnancy to avoid crisis.

Ghidini et al. [10], observed that 90% of women with SCI had not received the right information about pregnancy during the rehabilitation period.

Patients with paraplegia and pregnancy should be informed about the importance of caring for their skin, their gastrointestinal and genitourinary systems.

The known maternal-fetal complications in SCI patients are: recurrent urinary tract infections, kidney stones, constipation, pressure ulcers, preeclampsia on chronic hypertension, venous thrombosis, gestational diabetes and premature birth.

According to the literature, patients with acquired SCI don't have an increased risk of congenital malformations or

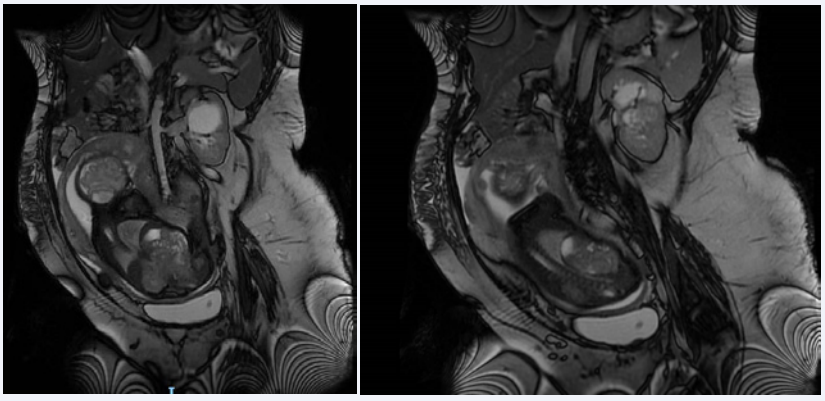

Figure 1,2 MRI at the 34th week of pregnancy. Neobladder with catheter previously positioned was located in the middle of the pelvic excavation. It clears dorsal and lumbar scoliosis.

intrauterine fetal death [14]. In our case, a genetic evaluation was not made during pregnancy, but second level ultrasound series.

Scientific studies have shown that 100% of self-catheterized pregnancies suffer from recurrent urinary tract infections in pregnancy and in the postpartum [2] and that paraplegia brings 25% more complications than women without disabilities [13].

For the prevention of premature birth it is necessary to investigate and treat any urinary, intestinal and vaginosis infections, check objective ultrasound parameters such as cervix length considering the poor perception of abdominal pain and the consequent cervical incontinence. There are known cases of precipitous birth without painful contractions [15].

Hypertension continues to be a major problem for SCI patients, but the possible medication taken during pregnancy is many, such as nifedipine or alpha methyl dopa.

In our case, chronic hypertension complicated by preeclampsia, recurrent urinary tract infections, kidney stones before pregnancy were found; there was no onset of gestational diabetes. Intermittent self-catheterization at the level of the umbilical stoma every 4 hours with a hydrophilic catheter has improved the recurrence of urinary infections in pregnancy, without episodes of renal colic. No Botox injections were necessary.

Autonomic hyperreflexia (AH), feared complication in paraplegic patients, was not found in our case, despite the stress factors such as contractions and pressure rises. It is important to discriminate between AH and preeclampsia, since inadequate treatment leads to an inadequate response. AH is characterized by excessive sweating, headache, patchy rash, pilo-motor erection, facial flushing, nasal congestion, convulsions and, consequently, placental uterine vasoconstriction, hypoxemia and fetal bradycardia [14].

About the mode of delivery and the type of anesthesia data are a bit contrasting: vaginal delivery is not contraindicated and anesthesia is often not necessary. It prefers epidural anesthesia especially in the onset of AH [16].

Often the caesarean section (up to 49% of cases) is carried out for the onset of maternal complications such as hypertension or related fetal complications such as prematurity, intrauterine growth restriction (IUGR) or fetal wrong position (10% of cases) [9].

In our case, caesarean section under general anesthesia was performed for maternal preeclampsia with IUGR and severe dorsal/lumbar scoliosis; surgical procedure was complicated by the anomalous neobladder, for which the execution of MRI before the surgical intervention and the collaboration with the urologists during the intervention was fundamental.

It is preferred to use metal clips for suturing the skin because of documented occurrence of infections or altered metabolism of the suture. The denervated area does not absorb the thread well with consequent sterile abscesses or suture dehiscence [17].

Therefore, an evaluation by a multidisciplinary team of experts, such as the gynecologist-obstetric surgeon, the anesthesiologist and the urologist, is necessary, as well as the neonatologist with neonatal intensive care in a tertiary center.

CONCLUSION

Despite the SCI is frequently nowadays are uncommon women with paraplegia pregnant than in cases of the past: it gives witness literature, full of examples until last decade.

It is important to know to manage pregnancies with this type of problem, because the situation could become very difficult. Often paraplegia pregnant arouses anxiety in caregivers because of lack of knowledge and the rarity of cases [18].

Above all, it is necessary to intervene in the preconceptional phase trying to prevent anything that could complicate this type of pregnancy, which disabled girls can still deal with due support.

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