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Case Report

Intrauterine fetoscopic closure of myelomeningocele: Clinical case and literature review

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ABSTRACT

Objective: Spina bifida (SB) is a congenital birth defect defined as a failure of the neural tube formation during the embryonic development phase. Fetoscopic repair of SB is a novel treatment technique that allows to close spinal defect early and prevent potential neurological and psychomotor complications.
Case report: We present a case report of a 32-year-old-multigravida whose fetus was diagnosed with lumbosacral myelomeningocele at 23rd week. Fetoscopic closure of MMC was performed at 26 weeks. At 32 weeks, due to premature amniorrhexis and placental abruption, an emergency C-section was performed. Newborn's psychomotor development was within normal limits.
Conclusion: Although intrauterine treatment has an increased risk of premature labor, placental abruption, prenatal closure is associated with improved postnatal psychomotor development. Prenatal surgery decreases the risk of Arnold-Chiari II malformation development and walking disability. Fetoscopic closure of SB is becoming a choice for treatment with beneficial outcomes for mother and fetus.

Introduction

Spina bifida is a developmental defect of the vertebra and spinal cord which prevalence varies among different countries and ethnic groups. Treating and living with this disease is a challenge for the patient, parents, and medical staff as it takes a lot of knowledge and effort to make the quality of life as best as possible. We present a case report of fetoscopic closure of myelomeningocele which was the most suitable treatment of spina bifida that allowed to achieve the best results in a patient's care.

Case presentation

A 32-year-old multigravida presented to the tertiary hospital at 23 weeks for a detailed fetal ultrasound scan. The ultrasound showed isolated lumbosacral myelomeningocele measuring 30 × 15 × 19 mm with no skin coverage, signs of Arnold-Chiari II malformation, and mild lateral ventriculomegaly up to 11 mm (Fig. 1A and B). The patient underwent amniocentesis at 24 weeks of gestation. Fluorescence in situ hybridization and culture had been performed to obtain karyotype results, which revealed normal karyotype with XY sex chromosomes. After counselling and obtaining the patient's consent, the minimally-invasive fetal surgery at 26 weeks of gestation was conducted.

The surgery was performed under general anesthesia. An ultrasound-guided amnioinfusion of 500 ml warm ringer lactate was performed to gain enough space in the amniotic cavity to allow safer trocar insertion. Three trocars (11 Fr) were inserted into the uterine cavity using the Seldinger technique under ultrasound control. After the removal of amniotic fluid, CO_2 insufflation was started, and the initial intrauterine pressure was measured. The upper pressure limit was set at 3–5 mm Hg above the initial uterine pressure, which was maximum 14 mmHg and gas flow was set at 15 L/min. The neural placode was released using a circumferential incision of monopolar needle at the transition zone. The skin was further undermined to allow approximation of the edges in the midline. In this case primary skin closure over the placode was possible. The skin was closed with a single running suture with a 3-0 self-blocking absorbable monofilament stitch (Fig. 2A, B, 2C). The amniotic fluid was replaced with warm ringer lactate to obtain

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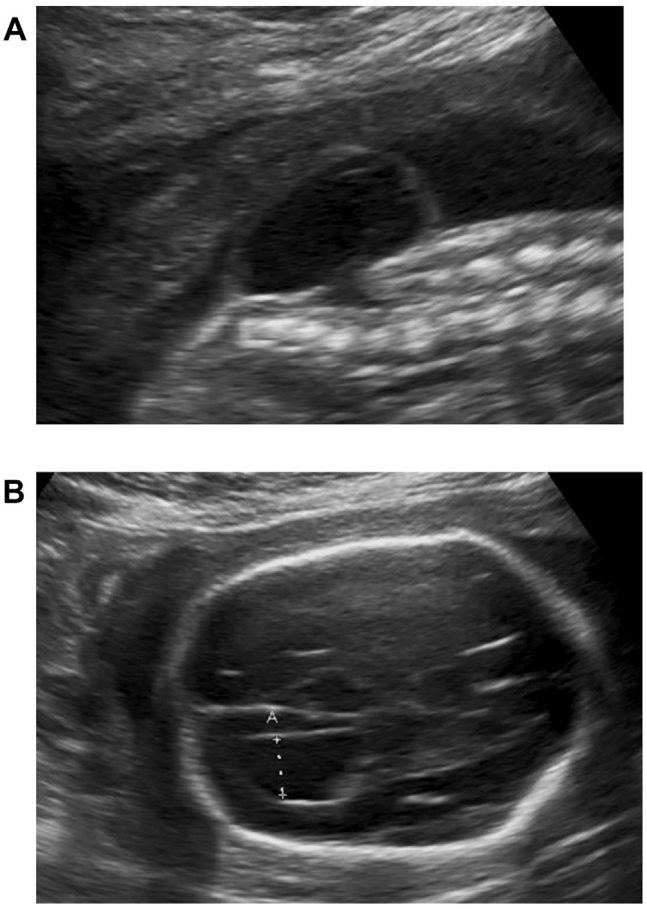

Fig. 1. A. Ultrasound image at 23 weeks gestation showing fetal lumbosacral Myelomeningocele. B. Ultrasound image at 23 weeks gestation showing fetal lateral ventriculomegaly.

the deepest pocket of 5 cm. Fetal heart rate and umbilical artery doppler were measured.

The patient arrived for the follow-up ultrasound at 30 weeks of gestation, which showed normal fetal growth, dynamically lowered amniotic fluid index, no signs of myelomeningocele, and slightly dilated third ventricle (Fig. 3A and B). The patient presented at the Emergency room at 32 5/7 weeks with premature rupture of membranes. Due to the increased uterine bleeding and suspected placental abruption, the patient undergone an emergency Caesarean section. A 2300 g male infant was born, Apgar scores were nine at 1 min and ten at 5 min. His vital signs were within normal limits, but he was transferred to the neonatal intensive care unit with continuous positive airway pressure. His physical examination revealed normal reflexes and muscle tone. On the 3rd day of life, neurosonography showed slightly dilated lateral ventricles up to 4 mm and hyperechogenic corpus callosum. On the same day, neurosurgeon counselled the infant and removed the skin stitches from the fetoscopic surgery. Neurological examination at the 5th day of life reported normal tactile senses and reflexes in legs, no defecation, urination abnormalities, intracerebral hypertension symptoms were observed.

One month after the birth, magnetic resonance imaging of the brain revealed partial dysgenesis of the corpus callosum with no other visible abnormalities of the following structures: brain, spinal cord, and spinal cavity. Neurological examination at seven months' age showed no urination and defecation problems. The amplitude of movements was not limited, but the trunk's posture was weak, and the extremities' muscle tone tended to be lower. Nonetheless, reflexes and tactile senses were explicit. The postoperative scar was seen in the lower part of the back (Fig. 4). Also, ultrasonography reported that the brain structures, except corpus callosum, were normally differentiated, lateral, third, and fourth ventricles were not dilatated. The patient currently is under the supervision of neurologist with positive dynamics in psychomotor development.

Discussion

Spina bifida is a congenital birth defect, defined by the failed development of neural tube during the embryonic development phase. The occurrence of the defect worldwide is 1 in 1000 newborns in Europe [1]. It varies among the ethnic groups, for instance, the occurrence is higher in Hispanics [2]. Unfortunately, the actual cause of spina bifida remains unknown; however, genetic and non-genetic risk factors are described. There is no specific gene related to spina bifida inheritance, but the genetic path is seen among the relatives in families where the malformation occurs. Studies show that siblings have a 2–5 percent higher recurrence risk than healthy families [3]. Also, genetic disorders, such as trisomy 13 and 18, mutations in MTHFR or DHFR genes, carry a higher risk of spinal defect occurrence [2]. Non-genetic factors, including well-known and described folate insufficiency, is usually associated with folic acid antagonists intake during pregnancy. Other known non-genetic risk factors leading to a higher risk of spina bifida are maternal diabetes mellitus, obesity, alcohol consumption, smoking [2].

Types of spina bifida are divided into closed (*spina bifida occulta*) and open (*spina bifida aperta*) as we have had in our case. The variations of the open spina bifida are myelomeningocele (MMC) and myelocele [4]. MMC is the most common and clinically the most significant one. During the period of embryological neural tube development when it is supposed to close, the neuroectodermic layer disconnects from the adjacent cutaneous ectoderm resulting in MMC [5]. The majority of MMCs occur in the lumbar region, though they can occur anywhere along the spine. If a spinal cord continues distally, beyond the dysraphism level, the neural placode is described as segmental. When the placode occurs at the end of the spinal cord, it is described as terminal.

Spina bifida treatment consists of the closure of myelomeningocele, prevention of ventriculomegaly, and its complications along with other neurologic, urologic, musculoskeletal, dermatologic comorbidities through patient's life [6]. Fetal surgical repair of MMC has been associated with improved early neurological outcome compared to postnatal operation. The primary purpose of the closure of myelomeningocele is to reconstruct anatomic layers which failed to develop during gestation, thus prevent layers from reattaching and restore adequate cerebrospinal fluid flow [7]. The first step of treatment is the early closure of neural tube defect as spinal cord exposure to the amniotic fluid, or the air is damaging the spinal cord itself. Conventional repair is done as soon as possible, in most cases, within two days after delivery. When the operation is performed later, the risk of infections, such as meningitis and ventriculitis, severe neurogenic urination prognosis, mortality, impaired muscle strength, neurodevelopment delay is increased. However, this early treatment does not decrease the rate of hydrocephalus [7,8]. In postnatally repaired, MMC hydrocephalus occurs for most patients during the first year after delivery [9].

Whereas postnatal surgery protects against infections and is performed for cosmetic reasons, it cannot restore already lost neurological function. In contrast, fetal surgery technique results in improved neurological function in most cases since it prevents further progression of the disease for the final four months of gestation. There is no difference between neonatal mortality in prenatal and postnatal surgery of MMC [9]. However, when

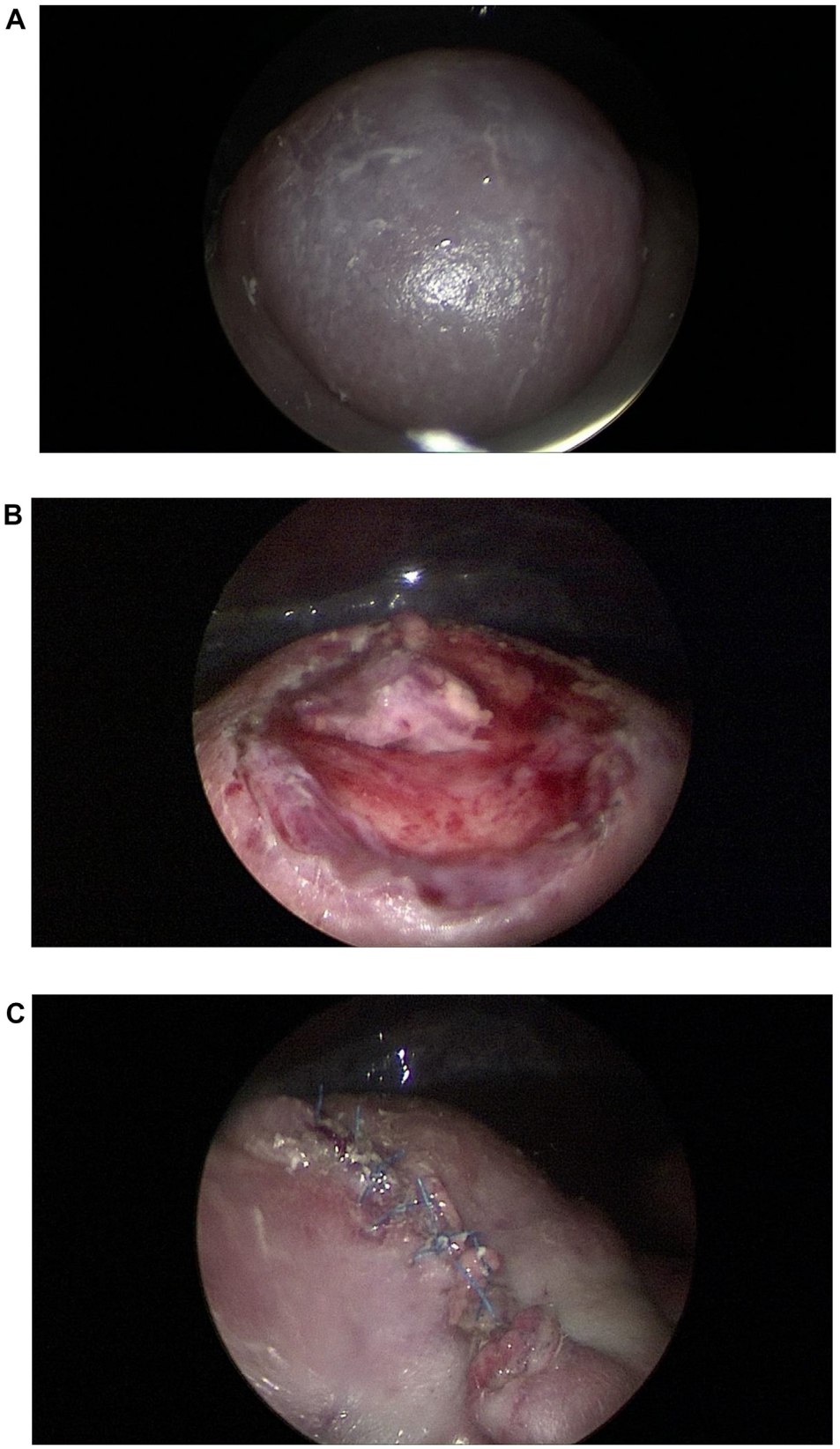

Fig. 2. A. Fetoscopic surgery: visualized Myelomeningocele. B. Fetoscopic surgery: placode dissected from Myelomeningocele. C. Fetoscopic surgery: sutured skin above vertebral defect.

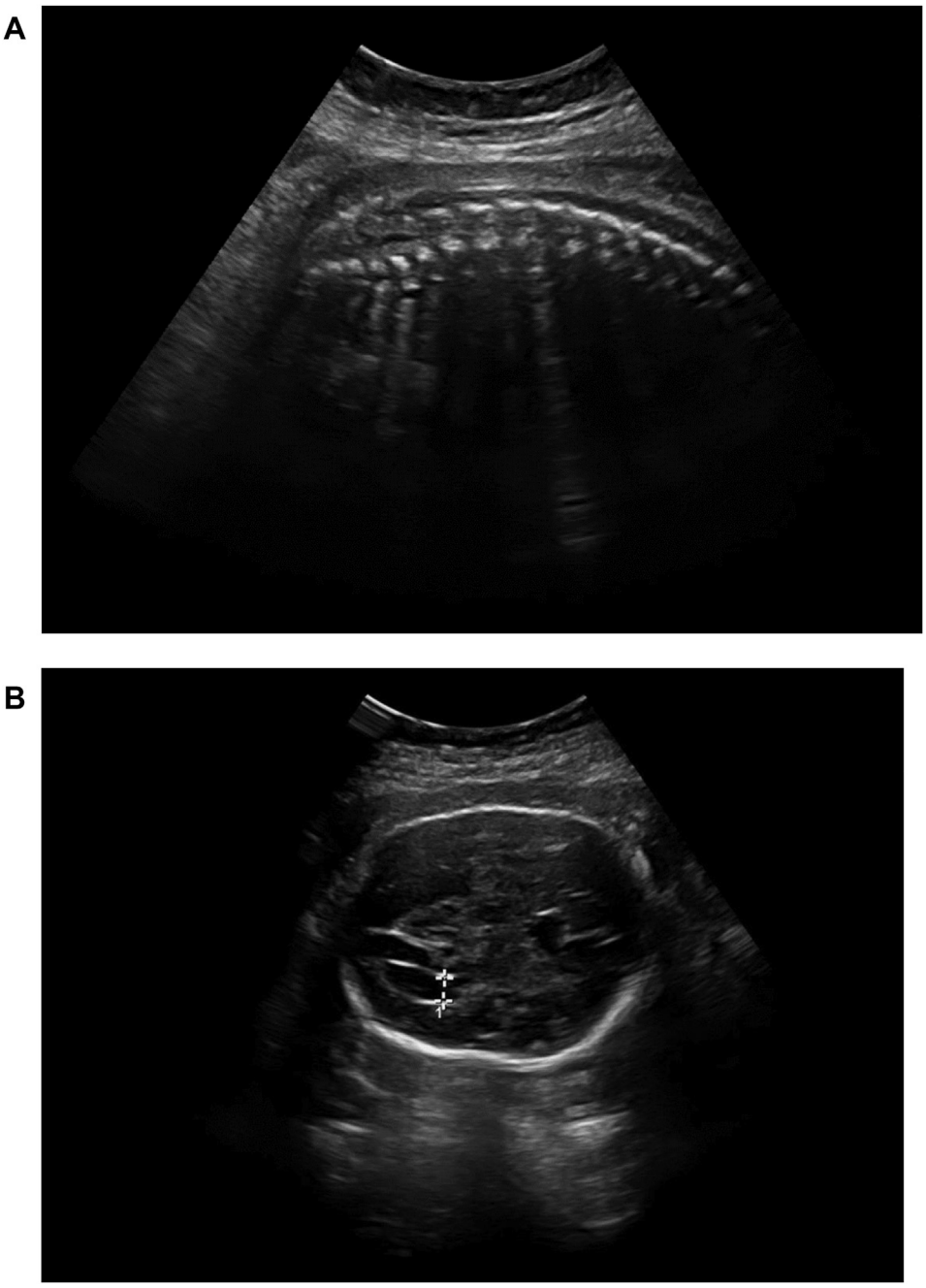

Fig. 3. A. Ultrasound scan of the fetus at 30 weeks: fixed Myelomeningocele. B. Ultrasound scan of the fetus at 30 weeks: slightly dilatated the third ventricle.

prenatal surgery is performed, the gestational age at birth is lower and the prevalence of placental abruption, separation of the membranes, preterm rupture of membranes is higher compared to postnatal surgery [9,10]. On the other hand, prenatal repair of MMC decreases the incidence of moderate to severe hindbrain herniation, hydrocephalus, and the necessity of cerebrospinal fluid shunt implantation [9,11]. Intrauterine surgery of MMC improves fetus postnatal ability to walk independently and increases the psychomotor evaluation score. However, this surgical approach is not associated with better mental outcomes than patients with postnatally closed MMC [9,12].

Prenatal closure of spina bifida could be performed in two manners: using an open or minimally invasive (fetoscopic) approach. Nor long term outcomes between these techniques do not differ significantly, open surgery is associated with increased risk of uterine dehiscence and requirement of blood transfusion during labor [13,14]. After fetoscopic surgery preterm premature rupture of membranes, premature labor occur more often, and postnatal revision of surgical suture due to separated margins of the lesion and leakage of cerebrospinal fluid is performed more frequently [13,14]. Nor fetoscopic surgery lasts longer, according to Sacco et al. meta-analysis open closure of spina bifida is associated with increased risk of intraoperative maternal complications [15]. Mild (bleeding during procedure, infection, hemotransfusion) and severe maternal complications (sepsis, massive haemorrhage requiring delivery, placental abruption, pulmonary edema) occurred more frequently compared to fetoscopic technique. Endoscopic approach allows to deliver subsequent pregnancies vaginaly, whereas after open surgery

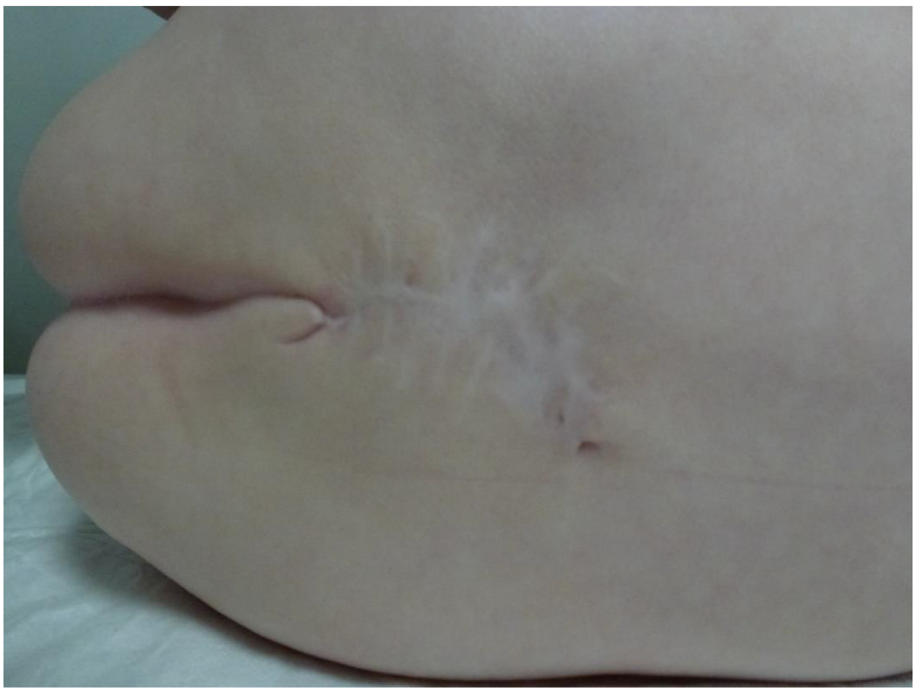

Fig. 4. Postoperative scar after intrauterine spina bifida repair.

to prevent rupture of uterine scar all fetuses have to be delivered during prelabor C-section [9].

Experimental novel approach of prenatal management of spina bifida is stem cells therapy. Both placental mesenchymal stem cells and amniotic fluid mesenchymal stem cells are being successfully used in rodent animal model to treat spina bifida [16]. They can induce adequate coverage of the skin and improve motor function. Spinal cord regeneration could be promoted by engineering fetal tissue with growth factors (VEGF, FGF2) or dura substitutes with biosyntethic materials [16]. After further studies, fetal tissue engineering technique combined with fetoscopic surgery could have a great impact in treating myelomeningocele.

Fetoscopic surgery is becoming the choice of treatment for carefully selected patients with myelomeningocele for its superior fetal outcomes and a lower rate of life-threatening maternal complications. Further studies are needed in order to investigate and develop fetoscopic surgical techniques and postoperative care.

Statement of ethics

We state that the patient has given written informed consent to publish the case (including publication of images) and the research was conducted ethically following the World Medical Association Declaration of Helsinki.

Conflicts of interest

The authors state no conflict of interest.

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We state that the material contained in the manuscript has not been published, has not been submitted and is not being submitted elsewhere. All authors have read the manuscript and approved of its contents. No funding to declare.

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