A Case Report of Maternal Spina Bifida that had a successful Pregnancy Outcome

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Abstract

Spina bifida is a neural tube defect which results from failure of closure of the neural arch during embryogenesis. The extent of the defect and physiologic impairments vary, it may be associated with neural, sensory, and motor losses, musculoskeletal impairments, urinary bladder and bowel sphincteric dysfunction. Term pregnancy among women with spina bifida is uncommon. With advances in management and care of children affected by spina bifida, more women survive to reproductive age group and achieve conception, pregnancy and delivery. There is dearth of information on the management of pregnant women with spinal bifida, which is why this case was reported.

This is a case report of a 23year old booked nulliparous woman with spina bifida occulta which was repaired at 3 months of life. She had a successful pregnancy and was delivered by caesarean section at term. This report describes the antepartum, intrapartum and postpartum management of a pregnant woman with Spina bifida.

Abstrait

Le spina bifida est une anomalie du tube neural résultant d'un défaut de fermeture de l'arc neural au cours de l'embryogenèse. L'étendue du défaut et les déficiences physiologiques varient, il peut être associé à des pertes neurales, sensorielles et motrices, des déficiences musculo-squelettiques, un dysfonctionnement de la vessie et du sphincter intestinal. Les grossesses à terme chez les femmes atteintes de spina bifida sont rares. Grâce aux progrès réalisés dans la prise en charge et les soins des enfants touchés par le spina bifida, davantage de femmes survivent jusqu'au groupe en âge de procréer et réalisent la conception, la grossesse et l'accouchement. Il y a peu d'informations sur la prise en charge des femmes enceintes atteintes de spina-bifida, c'est pourquoi ce cas a été signalé.

Il s'agit d'un rapport de cas d'une femme nullipare réservée âgée de 23 ans avec un spina bifida occulta qui a été réparé à 3 mois de vie. Elle a eu une grossesse réussie et a accouché par césarienne à terme. Ce rapport décrit la prise en charge antepartum, intrapartum et postpartum d'une femme enceinte atteinte de spina bifida.

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Introduction

Pregnancy in a woman living with spina bifida is not common, however, with improved care, spina bifida patients are able to survive to reproductive age, become pregnant and deliver successfully. The clinical presentation of this group of women varies based on the type of spina bifida, the severity of clinical symptoms, the pregnancy-associated challenges and other co-morbidities.[1] Spina bifida (SB) is a neural tube defect (NTD) resulting from failure of closure of the neural tube during embryogenesis. NTD is classified into occult (closed), open (meningomyelocoele or meningocoele), and dysraphism based on the size, location and tissues or structures involved. SB may be associated with neural, sensory, and motor losses, musculoskeletal impairments, bony deformity such as scoliosis, bladder and bowel sphincteric dysfunction, and occasional cognitive impairment.[2,3,4]

Worldwide, the incidence of spina bifida was estimated at 1-2 cases per 1000 births and this incidence varies with the degree of genetic predilection among different population.[5] The meningomyelocoele variety of spina bifida is the commonest type and accounts for about 75% of all cases of NTD.[4,5] Neural tube defects are the second most common type of birth defect after congenital heart defects. The rate of meningomyelocele and other neural tube defects has declined since the late 20th century and this is attributable to the widespread availability of prenatal diagnostic services and to improved nutrition among pregnant women.[7-9] The aetiology of spina bifida is multifactorial, involving genetic, family history, racial, drugs and environmental factors, in which nutrition, particularly folic acid intake, is the most crucial.[10]

Advances in the surgical correction and management of spinal bifida cystica and its related complications in infancy improved the survival rate, with 50% of patients surviving to the age of 30 years and above. Pregnancy in a patient with spinal bifida maybe associated with higher risk of delivering a baby with neural tube defect compared with the general population (4% vs 0.1%–0.3%),[9] worsening of bowel and bladder dysfunction, back pain and movement disorders among others.[9] However, in some instances, the pregnancy may be normal with no adverse events to the mother or fetus. There is paucity of literature on the management of pregnant women with spinal bifida in our environment and necessitates this case report. This report describes the antepartum, intrapartum and postpartum management of a pregnant woman with Spina bifida.

Case Report

The 23year old nulliparous woman with spina bifida occulta presented for antenatal care on account of ultrasound diagnosis of intrauterine pregnancy at a gestational age of 17 weeks at the University College Hospital (UCH), Ibadan, Nigeria.

The NTD was repaired at 3 months of life by the neurosurgery team at the UCH, Ibadan Nigeria. There was associated urinary and faecal incontinence which she is being managed by life style modification.

She started oral folic acid 5mg twice daily in the first trimester of pregnancy and booked the index pregnancy for antenatal care at our facility in the second trimester (at a gestational age of 19weeks). On physical examination; the general examination revealed a healthy young woman, ambulant with no gait abnormality, not in obvious distress, not pale, anicteric, afebrile and there was no pedal oedema.

On systemic examination, the neurologic and musculoskeletal examination revealed normal power, tone and reflexes in all the limbs; and no motor deficit. The scar of the repaired vertebral defect was at the S2-S4 vertebral level. Abdominal examination revealed an abdomen distended by the gravid uterus and the uterine fundal height was 18weeks sized. There was no tenderness and no other palpable abdominal mass.

She was counselled about the physiologic changes of pregnancy, her condition and its implication in pregnancy. She was also counseled on the risk of recurrent urinary tract infections from urine stasis and effect of pregnancy hormone on the urinary tract, increasing bowel and bladder dysfunction and worsening back pain in pregnancy. She was continued on high-dose folic acid supplement. She had fetal anomaly scan performed at 20 weeks gestation which showed there was no fetal anomaly. She was co-managed with the neurosurgery and urology teams. She was regular with routine antenatal care clinic visits, there was no report of urinary tract infection; but she had occasional urinary and faecal incontinence.

The patient had ultrasound done at 36 weeks gestation which showed a single live intrauterine fetus in breech presentation. She was counseled for elective caesarean delivery. The patient was reviewed at 37-weeks by the anaesthetist and paediatrician as part of pre-operative preparation for caesarean section. She had an elective lower segment caesarean delivery at 38 weeks and 5days under regional anaesthesia. A subarachnoid block was placed at the L3/L4 Vertebral level. She was delivered of a live male neonate with Apgar score of 9 in the first

minute, 10 in the fifth minute of life and a birth weight of 3.45kg. The neonate was examined, and there no abnormality was detected on him.

The patient had intravenous fluid, prophylactic parenteral antibiotics and postoperative analgesia given - intramuscular Pethidine 100mg 6hourly for 48hours with good pain control. Thromboprophylaxis was commenced with compression stockings (thromboembolic deterrent stockings-TED), subcutaneous enoxaparin combined with early ambulation. The postoperative period was good. She was counseled on exclusive breastfeeding, infant immunization, family planning and regular cervical screening for cervical cancer prevention. She was discharged home on the fourth postoperative day.

She attended postnatal clinic at the sixth week postpartum and she had no complaints. The surgical wound site healed well with no complication.

Discussion

The patient presented in this case report is a young woman with post-repair Spina bifida and residual urinary and faecal incontinence who had a normal pregnancy and caesarean delivery of a healthy male neonate at term under a multidisciplinary team management. As a result of improved medical and surgical intervention in the management of Spina bifida in infancy; patients with spina bifida survive, reaching adulthood and able to reproduce.[1] Although relatively uncommon in our environment, cases of successful pregnancies have been reported in the developed climes.[1,10,13]

Before pregnancy, the spinal bifida patient should receive preconception care[4,12] and genetic counselling on the risk of neural tube defect, folic acid supplementation from three months preconception until twelve weeks of pregnancy, medication review, review of other co-morbidities such as obesity, epilepsy, cerebrospinal fluid(CSF) shunt - ventriculo-peritoneal shunts, spinal deformities (scoliosis and kyphoscoliosis), venous thromboembolism (VTE) , musculoskeletal complications[4] and the implication of the maternal condition on the antenatal, delivery and postpartum care. The risk of recurrence is about 1-5% if one parent is affected and 15% if both parents are affected.[4]

Pregnancy in women with Spina bifida can be challenging due to severity of symptoms, physical limitation, previous abdominal surgery, urinary and bowel involvement, presence of ventriculoperitoneal shunt, and co-morbidities.[1] The management of pregnancy in patients with Spina bifida requires a multidisciplinary team approach.[1,4] The team care approach will involve obstetrician, paediatrician, anaesthetist and other specialists – urologist, neurologist/neurosurgeons – depending on the medical condition during pregnancy and delivery. Some patients with NTD may have uneventful antepartum, intrapartum and postpartum course as it was seen in this woman. In such cases, antenatal care will be given while assessing risk factors for complication and preventing complications. However, some women may have other pregnancy- or spina bifida- related challenges that may alter the course of the pregnancy. Women with spina bifida may have a risk of baby with (4-7%) spina bifida or anencephaly.[13] For this reason, a prophylactic high dose folic acid is recommended before conception and in the first trimester of pregnancy to reduce the risk of NTD.[13,14]

Antenatal management should include screening for NTDs using the 20-week fetal anomaly scan and the maternal quadruple test which includes the serum alpha fetoprotein assay. Women with NTD are at increased risk of antenatal admissions, especially women that are not or partially ambulant; recurrent urinary tract infections, urinary tract obstruction, bowel issues – constipation, faecal incontinence; preterm delivery, worsening decubitus ulcers, worsening back pain, reduced mobility, VTE, haematologic issues, blood transfusion and CSF shunt displacement.[4,15-17] Restrictive kyphosis and pelvic bone deformity may cause problem with accommodating the growing gravid uterus and/or reduce the pelvic measurements.[18]

The index patient did not experience pregnancy- or spina bifida-related complications and the pregnancy was uneventful. Despite the seemingly uneventful experience of our patient, women with NTD should always have antenatal care and childbirth in a specialized or tertiary health facility where specialist services can easily be accessed and utilized without undue delay. For example, patients with previous urologic or multiple vertebral surgeries may pose a challenge to caesarean section or neuraxial anaesthesia in view of difficult access to the epidural or spinal space.[19]

The patient had a successful elective caesarean section under regional anaesthesia at term. Our patient was a primigravidae that had breech presentation at term. There have been reports of safe vaginal deliveries among some women with spina bifida.[20,21] Arata et al, reported vaginal delivery in one in five and one in ten pregnancies among wheelchair dependent and independent women respectively.[21] In this case, the patient had an obstetric indication for caesarean section. Raji, et al reported a case of maternal spina bifida that was delivered by elective caesarean section at term due to abnormal pelvis, wasted muscles of the lower limbs, and previous multiple spina surgeries.[15]

Women with spina bifida are at increased risk of caesarean delivery and a higher risk of morbidity compared to women without spina bifida; the frequency of caesarean delivery also increased with severity of spina bifida.[16] General anaesthesia is an option for anaesthesia especially in severe cases, however, there is no absolute contraindication for regional anaesthesia in patients with spina bifida. The review, consideration and decision for route of anaesthesia should be made by a senior obstetric anaesthetist early. The route of anaesthesia was regional – subarachnoid block. Our patient had subarachnoid block because the level of the previous spinal defect was at the sacral spine and absence of motor deficit. The patient had urinary and faecal incontinence attributable to sacral nerve deficit due to the defect at the sacral level of the vertebrae. Women should be catheterized to prevent urinary retention.

There is limited evidence regarding postnatal care for the woman with SB. In the postnatal period, management of a postpartum women with SB should entail a review of pregnancy co-morbidities, risk assessment for VTE, VTE prophylaxis, infant feeding options, breastfeeding and challenges of breastfeeding such as positioning in women with kyphoscoliosis or limb contractures, maternal medications and safety for breastfeeding, and contraceptive options. Women with SB, who are at risk of VTE should avoid oestrogen-containing contraceptives and intrauterine contraceptive device may be difficult in women with limb contractures.[4] This patient had routine postnatal care and the postnatal period was uneventful.

In conclusion, this case highlights the possibility of safe pregnancy and delivery in a woman with NTD and further elucidated that a woman with NTD can get pregnant and deliver her baby without complication. We recommended that pregnant women with NTD should be managed at a specialized/tertiary health center where access to multidisciplinary care is achievable.

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Figure 1: Shows vertebral level of spina bifida repair – Sacral level (blue arrow)

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