Case Report

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Successful Pregnancy Outcome in an Operated Case of Lipomeningomyocele: A Rare Case

Obstetrics and Gynaecology Section

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ABSTRACT

Lipomeningomyocele is one of the types of occult spinal dysraphism associated with tethered cord syndrome, which is characterised by neurodeficit symptomatology due to stretch- induced functional disorder of the spinal cord with its caudal part attached by an in elastic tissue. There is stretching of conus medullaris and nerve roots due to disproportionate growth between vertebral column and spinal cord, resulting in various neurological deficits. Its incidence is approximately 1 in 4000 births in the USA, with a slightly higher female preponderance, although its true incidence is not known. There are very few studies on pregnancy outcome in these patients and the management dilemma in this group has prompted us to report this case.

Keywords: Management dilemma, Neurodeficit, Occult spinal dysraphism

CASE REPORT

A 24-year-old primigravida, with 20 weeks gestation was referred with complaints of abdominal pain and burning micturition since 6 days. She was a known case of tethered cord syndrome because of lumbar lipomeningomyocele for which she was operated at 2.5 years of age. Even after surgery she had symptoms of neurogenic bladder and constipation. Later on, she developed left lower limb weakness since 10 years. She was taking folic acid (0.4mg/day) in early gestation period. She used to do intermittent self catheterisation 3-4 times a day using urethral catheter, R – 90, FG – 14 [Table/Fig-1]. With increase in gestation period, her voiding frequency increased to 5-6 times a day, for which silicone catherisation was done, following admission. On examination, patient's vitals were normal. Uterus was 20-22 weeks in size, relaxed. On per speculum examination there was no leaking or discharge, on vaginal examination, cervical os was closed and pelvis was constricted. There was no other skeletal deformity. Her haemoglobin and total counts were within normal limits. Urine microscopy examination revealed severe urinary infection with *Pseudomonas aeruginosa* colonies more than 10^5, for which she was given intravenous antibiotics according to culture sensitivity. Her sonography was suggestive of right sided gross hydronephrosis with thin parenchyma and left sided mild to moderate hydronephrosis with thick walled trabeculated bladder. She took antenatal care in some other hospital where her symptoms were treated as simple urinary tract infection. Her complaints of recurrent urinary infection had increased with time, for which she was routinely screened till term. Rest of her antenatal period was uneventful.

At 37 weeks, elective caesarean section was done for pelvic inlet constriction followed by delivery of healthy male child weighing 2.8 kg. Post partum, both mother and baby was good and silicone catheter was removed within 48 hours of delivery. After 6 weeks of follow-up, her bilateral hydronephrosis was relieved and her renal function tests were within normal limits. She continued using self-intermittent catheter for voiding.

DISCUSSION

Lipomeningomyocele is one of the types of occult spinal dysraphism associated with tethered cord syndrome, which arise from the disorder of embryogenesis and usually consist of single cell type. It constitute subcutaneous fibrofatty mass that transverses the spinal region causing a spinal laminar defect, penetrating dura and tethering the spinal cord [1]. Its incidence is approximately 1 in 4000 births in the USA [2]. The age of presentation can vary from early childhood to the 80s [3]. The symptoms are commonly present in childhood, but the diagnosis is usually established in the adulthood [4]. However, it can be diagnosed with spinal ultrasound in neonates and Magnetic Resonance Imaging (MRI) in older children. Radiological abnormalities such as low lying conusmedullaris, lumbosacral lipoma, filum terminal elipoma, or thick filumterminale are the keys for the diagnosis of tethered cord syndrome [5].

Clinical features vary according to the area of traction on the lower spinal cord, leading to various neurological deficits like motor deficits, sensory disturbance, spasticity, urological, rectal dysfunctions and skeletal deformities. Pathogenesis and natural history of these complex anomalies are not clearly understood. These deficits may increase over course of time.

The patients may have congenital anomalies of the female reproductive tract, such as bicornuate uterus or prolapsed uterus at a much younger age due to laxity of pelvic muscles and uterine supports, which may be difficult to treat as standard surgeries

[Table/Fig-1]: Patient used to do intermittent self catheterisation using urethral catheter.

may not be effective [6]. Although sexual dysfunction can occur due to neurogenic bowel and bladder, which often necessitates evacuation of bowel and bladder just before intercourse, still they are fertile and can have successful pregnancy outcome as in our case [7].

There are many studies on the medical management of these patients but, a standard for obstetrical care still does not exist [8]. Pregnancy complications are unique because of their neurological deficit (like neurogenic bladder, gastrointestinal incontinence) and secondary conditions due to spinal deformities. The most common complication is urinary tract infection. In 10% of pregnancies, growing fetus, causes hydronephrosis, intestinal obstruction, and renal complication. They can have early pregnancy loss and pre-term delivery leading to low birth weight babies. Skeletal abnormalities often lead to respiratory compromise, cardiac failure, and difficulties with labour and delivery [9]. Urinary obstruction and renal failure can occur, especially if the abdomen is further compromised by exaggerated spine curvature [10].

Infants can also develop postnatal complications like hypoglycaemia, jaundice, respiratory distress syndrome and tachypnea [11]. They can have congenital anomalies but in most of the cases outcome is good. Although, caesarean-section is a common mode of delivery in 42-50% of these patients, because of pelvic bony abnormalities and lower extremity contractures. But vaginal delivery can be conducted uneventfully [12].

It is recommended that women should have sufficient folate supplementation to lessen the risk of having a newborn with neural tube defects. Past literature has recommended an intake of 0.4 mg of folate per day for pregnant women [13]. However, women with neural tube defect should have even larger doses of folate than their counterparts (4.0mg of folate/day) during periconceptional period.

The risk for neural tube defects in offsprings of women with spinal defect is 4 to 7% [14]. Although the risk is high, the actual cases of offsprings with spinal defects have not been documented due to improved genetic counseling and high rate of miscarriages.

CONCLUSION

Preconceptional counseling is universal, but special care and attention is required in these groups of women about changes of pregnancy and its effect on her clinical condition. They should also be recommended, to have 4.0mg of folate every day before and during pregnancy to prevent neural tube defects in their offsprings. These patients can have good pregnancy outcome by managing their conditions and being vigilant about anticipated complications during their antenatal visits.

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