

CASE REPORT

Successful Pregnancy Outcome by Caesarean Section in a Woman with Arthrogyrosis Multiple Congenita (AMC)

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Abstract

Arthrogyrosis Multiplex Congenita (AMC) is a symptom complex of congenital joint contractures associated with neurogenic and myopathic disorders. It is diagnosed at birth and often progresses to a state of significant disability. Pregnancy in a woman with AMC is at high risk due to diminished pulmonary reserve, increased risk of thromboembolism and anesthesia. Successful pregnancy in patients with AMC is very rare and only five cases have been reported in literature. Present case is being reported due to its rarity (*Afr. J. Reprod. Health* 2010; 14[3]: 233-234).

Résumé

L'arthrogryose congénitale multiple (ACM) est un symptôme complexe des contractures articulaires congénitales liées aux troubles neurogènes et myopathiques. Il est diagnostiqué à la naissance et progresse souvent jusqu'à un état d'incapacité significative. La grossesse chez une femme qui souffre de l'ACM est à haut risque dû au réserve pulmonaire réduit, un risque élevé de la thromboembolie et de l'anesthésie. La grossesse réussite chez les patientes atteintes de l'ACM est très rare et il n'y a que cinq cas qui ont été signalés dans la littérature. Ce cas présent est signalé à cause de sa rareté (*Afr. J. Reprod. Health* 2010; 14[3]: 233-234).

Key words: Arthrogyrosis multiplex congenita, pregnancy, successful outcome.

Introduction

Arthrogyrosis Multiplex Congenita (AMC) is a multifactorial syndrome caused by a variety of neurogenic and myopathic disorders. It is first diagnosed at birth and often progresses to a state of significant disability. It includes abnormalities of cardiovascular, respiratory, nervous and genitourinary systems¹. AMC is sporadic and is classified by the pattern of involvement of the joints, amyoplasia being the most common form, comprising one third of cases. In this form, the limbs are affected symmetrically and in 84% of cases all four limbs are affected². We came across a case of AMC with full term pregnancy which was managed successfully. There are only 5 cases of AMC with successful pregnancy outcome reported in literature.

Case Report:

A 25 years old fifth gravida with previous four abortions was admitted in a tertiary care centre with 36 weeks of pregnancy with labor pain since two hours. Her antenatal period was uneventful. On examination, she was short stature (130 cm) and iliac spine

to feet height was 65 cm. She had congenital deformity of both hands and feet and could not walk. She had to crawl on both hands and feet (Figure 1). Spinal involvement produced a marked lumbar lordosis and thoracic kyphosis with normal neck architecture. There was no family history of AMC. She had no other significant medical and surgical illness. Her blood pressure and hemoglobin were within normal limits. Ultrasound of the fetus revealed a single live fetus with no congenital malformations. Pelvic assessment revealed severe inlet contraction. Emergency cesarean section was done under general anesthesia in view of contracted pelvis. She delivered a full term healthy, baby girl weighing 2.5 kg with no deformity. She had uneventful postoperative recovery and was discharged in good condition.

Discussion

Arthrogyrosis Multiplex Congenita was first described in 1905 and affects 1 in 3000 births. It is a symptom complex of congenital joint contractures associated with both neurologic and myopathic etiologies. Joint deformities are primarily neurogenic in



Figure 1. Arthrogyrosis Multiplex Congenita (AMC).

origin (>90%), rest being myopathic. Various causes are genetic, environmental or rarely maternal drugs like alcohol, phenytoin, and drug abuse etc¹.

Deformities lead to moderate disability in patients which worsen during pregnancy. Respiratory function is adversely affected with advancing gestation due to lack of available space in abdomen for uterine enlargement as well as diminished pulmonary reserve. Respiratory function may be affected to such an extent that baby may have to be delivered preterm, as was done in two cases reported by Duffy et al³ and Hardwick et al⁴ in whom elective cesarean section was done at 31 and 30 weeks respectively. But in the present case, since the cardio respiratory function was not severely affected, so pregnancy was carried till term.

During pregnancy there is an increased risk of thrombo-embolism and anaesthesia. Most of the cases of arthrogyrosis are diagnosed at birth; however an antenatal history of decreased fetal movements may be suggestive of the condition in utero. Ultrasound may detect arthrogyrosis in second and third trimesters, showing decreased movements or abnormal posture. If other anomalies co-exist, it may be diagnosed even earlier⁵.

Only five cases of successful pregnancy have been reported in women with AMC³⁻⁷. In two cases, preterm emergency caesarean section was done due to deteriorating respiratory condition^{3,4}. Quance et al⁶ reported a case which was continued till term and underwent elective caesarean section for contracted pelvis, as was done in present case. In another case a successful, full term vaginal delivery was conducted⁵.

Pregnancy in women with AMC is very high risk due to physical and functional disabilities. X ray pelvimetry has some role in this condition as in most, the pelvis will be asymmetrical, depending on the degree of involvement of the joints and the posture,

patient adopts during walking. In present case radiological pelvimetry was not done as decision of caesarean section was taken in lieu of precious pregnancy (previous four abortions) and clinically contracted pelvis. The social circumstances of the women are also challenging in terms of caring of disabled mother and newborn baby which may be pre-term in some cases.

Thus obstetric management requires a multidisciplinary approach involving preconception counselling, planning care of mother and baby, during and after delivery and also contraceptive advice depending on the disability. Timing of delivery is challenging in view of maternal risks, especially when the fetus is healthy.

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