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Case Report

Arthrogryposis multiplex congenita in twin 1 of a twin pregnancy: case report

Stephanie Rachel Dsouza[1]*, Abhey Wasdev[2], Prashanth Gowda[3]

[1]Department of Neonatology, Motherhood Hospital, Bengaluru, Karnataka, India
[2]Department of Orthopaedic Surgery, Bengaluru, Karnataka, India
[3]Department of Neonatology, Motherhood Hospital, Bengaluru, Karnataka, India

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***Correspondence:**
Dr. Stephanie Rachel Dsouza,
E-mail: stefdza21@gmail.com

ABSTRACT

Arthrogryposis multiplex congenita (AMC) is a rare congenital syndrome characterized by multiple joint contractures. It usually affects the joints of hands, wrists, elbows, shoulders, hips, feet, knees, jaw and back. This case report discusses a mild variety of AMC in a twin where the other twin had no musculoskeletal abnormalities. There has been no published case report of such a case in a twin delivery. Prompt recognition of the syndrome complex and early start of stretching exercises and physiotherapy is the mainstay treatment protocol to give relative contracture free mobility to the child.

Keywords: Arthrogryposis, Joint contractures, Twin, Physiotherapy

INTRODUCTION

Arthrogryposis multiplex congenita (AMC) is a rare congenital syndrome characterized by multiple joint contractures and, may also include, muscle weakening and fibrosis. It is a non-progressive disease. The word “arthrogryposis” in Greek directly translates as "curved or hooked joints."[1]

Fetal movements in the amniotic fluid are a necessary condition for adequate fetal growth. Limited or absent joint movements can cause various functional or morphological abnormalities for the fetus.[2]

Few joints may occasionally be impacted or may have a virtually complete range of motion. Amyoplasia, the most prevalent form of arthrogryposis, affects the hands, wrists, elbows, shoulders, hips, feet, knees, jaw and back.[2,3] Muscle weakness frequently coexists with the contractures, which further restricts motion. AMC normally involves all four extremities and is symmetrical, though there may be some variance.[3]

CASE REPORT

A 32-year-old woman; Primigravida; 36+4 weeks dichorionic diamniotic (DCDA) twin gestation by *in-vitro* fertilization (IVF) with a history of gestational hypertension was admitted to our hospital for elective lower (uterine) segment caesarean section (LSCS). Antenatal scans were told to have limited mobility of one twin in the final scan. Twin 1 was under fetal distress for which emergency LSCS was performed. Twin 2 was a female baby with low birth weight (LBW-1.88 kg), cried immediately after birth and had no obvious abnormalities. Twin 1 was a female baby with 1.64 kg, didn’t cry after birth; was intubated, shifted to NICU and connected to mechanical ventilator-SIMV mode with FiO_2 of 80%, PEEP-6 cm, PIP-16 cm, VR-40/min, Ti 0.3 sec. APGAR score was 3/10 at 1 min and 5/10 at 5 min. Baby was started on IV fluids and antibiotics after sending sepsis screen work-up, blood gas and chest x-ray. The baby didn't fit into the HIE criteria. 2 doses of surfactant were given. Saturation was labile with suspected Persistent pulmonary hypertension of newborn (PPHN). Baby was

weaned off from mechanical ventilation to CPAP on day 4 of life to room air on day 5. Baby had high total counts and thrombocytopenia due to which antibiotics were upgraded to Meropenem and FFP was transfused. Hypokalemia, hyponatremia and hypocalcemia were corrected. Aminoven infusion was started on day 2. ECHO at birth showed small PDA BD, moderate secundum ASD, severe PAH and high PVR. Neurosonogram at birth showed diffuse increased echogenecity in bilateral subcortical white matter, basal ganglia and thalami region, suggestive of, prolonged hypoxic ischemic changes with reduced resistive index of anterior cerebral arteries (0.48). Follow up ECHO at 1 week showed moderate secundum ASD. Follow up neurosonogram at 1 week showed only grade 1 PVL.

Figure 1: Twin 1 with multiple contractures of all joints.

Musculoskeletal examination of the baby revealed contractures with persistent flexed posture of all four limbs (Figure 1) with increased tone. Right shoulder was adducted and internally rotated. Both wrists were flexed with both thumbs adducted. Left wrist was ulnar deviated. Both knees and elbows were in flexed position. Range of motion was severely limited in both elbows, both knees and right ankle. There was moderately limited range of motion of both wrists, 1st CMC joints, both shoulders and left ankle. Baby was also diagnosed to have right sided congenital talipes equino varus (CTEV) with classical cavus, adductus, varus and equines deformities. Spine and hip examination was normal. Baby also had difficult mouth opening due to which feeding was delayed. Bilateral hip ultrasound was done and found to be normal. Paediatric orthopaedic surgeon was consulted and stretching exercises were started soon after birth. Gradually tone reduced over next 5 days in the hospital and range of motion improved. Jaw contracture improved with gradual stretching over next one week and feeds were started after one week. Serial casting for CTEV was started by Ponsetti method. At present (3 months of age), the baby is feeding well with full mouth opening. Joint contractures have gradually improved with around 10 degrees of extension lag in both knees and around 20 degrees lag in both elbows. Right shoulder abduction is limited by around 20 degrees. Right ankle is on a foot abduction splint after correction of CTEV. All other joints have almost full range of motion. Baby has gained weight well and is active. Baby is on regular physiotherapy for remaining joint contractures. Follow up ECHO and USG at 3 months are normal.

DISCUSSION

Athrogryposis affects between 1/3000 and 1/10000 newborns, according to estimates. Arthrogryposis is not a specific diagnosis, but rather a syndrome that is present in a variety of disorders and is characterized by the presence of several congenital contractures. The prognosis mostly depends on the etiology, which might be maternal or fetal, neurogenic or myogenic, among other things.[4] This case report discusses a mild variety of AMC in a twin where the other twin had no musculoskeletal abnormalities. There has been no published case report of such a case in a twin delivery. There are very few researches available for role of physiotherapy and serial stretching in arthrogryposis multiplex congenita and other related congenital abnormalities. Prompt recognition of the syndrome complex and early start of stretching exercises and physiotherapy is the mainstay of treatment protocol to give relative contracture free mobility to the child.

CONCLUSION

The challenge for the radiologist, obstetrician and neonatologist in such cases, is to identify limited mobility and oligohydramnios antenataly which becomes a challenge in twin pregnancy. Once recognized, the prognosis and treatment plan has to be carefully counseled to the parents. Early Paediatric Orthopaedic surgeon and Physiotherapist opinion is of utmost importance. Finally, the role of parents in understanding the prognosis and performing regular exercises at home to give their child a relatively unassisted contracture free life can't be more stressed upon.

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