ARTHROGRYPOSIS MULTIPLEX CONGENITA - A CASE REPORT FROM A TERTIARY CARE HOSPITAL IN KERALA

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Abstract: Arthrogryposis multiplex congenital is a syndrome characterised by presence of multiple joint contractures prior to birth, limiting the range of mobility of the joints. It is non progressive and occurs mainly due to fetal akinesia. Its etiology is multifactorial. Presented here is the case report of a 11month old girl child born with congenital anomaly of left hand and leg along with developmental dysplasia of hip and Tallipes Equinovarus Deformity, underwent posteriomedial soft tissue release 4 months after birth and was admitted for surgical fixation of dislocated hip.

Keywords: Arthrogryposis multiplex congenital, multiple joint contractures, fetal akinesia, Developmental Dysplasia of Hip, Tallipes Equinovarus

INTRODUCTION

Arthrogryposis multiplex congenital refers to development of multiple joint contractures prior to birth, which may affect two or more areas in the body. When a joint becomes fixed in a straightened or bent position for a long time, a contracture occurs and this may impact the range of motion and function of these joints[1][2][3]. In some cases the condition can be so severe that every joints in the body may be affected including jaw and back, while in other cases only few joints may be affected that the range of motion will be nearly normal[3]. Atrophy or underdevelopment of the muscles of the affected limbs are also observed[2][3]. When a joint is not moved for quite a long time, connective tissue may grow around it and fix it in its place. Lack of movement of joints may also mean that the tendons in that area is not stretched to its full length and this may further make joint movement difficult. Soft tissue webbing may also occur around the affected joint[3]. Although arthrogryposis multiplex congenita can be classified into many different forms, it can be essentially classified into limb involvement and limb involvement with other organ involvement[2][9].

The exact cause that lead to Arthrogryposis is not fully understood. The most suspected reason for occurrence of multiple contractures is decreased fetal movement in the mother’s womb. There are a variety of different causes that contribute to reduced fetal movements[1][2].

In general, the causes for decreased fetal movement prior to birth includes[3]

1. Abnormal development of muscles: The specific cause for this is cannot be identified but the suspected causes are maternal fever during pregnancy, muscles diseases, viral infections that can damage cells that transmit nerve impulses to muscle.
2. Insufficient space in uterus for normal movement: Oligohydraminos, multiple foetuses and uterine structural abnormalities.
3. Malformations of central nervous system (brain or spinal cord) which is usually accompanied by other symptoms.
4. Abnormal development of tendons, joints or joint linings.

Arthrogryposis multiplex congenita can be considered as a symptom associated with different medical conditions. Environmental and genetic factors are thought to play role in its development[1]. Genetic conditions that can cause AMC include connective tissue disorders, muscle disorders such as muscular dystrophies or congenital myopathies, certain mitochondrial disorders, single gene change or chromosomal abnormalities such as trisomy 18[2].

Treatment for AMC should be planned according to the condition of the patient. The importance of long term treatment for this condition should be explained to patients and caregivers[7]. A multidisciplinary approach is usually considered essential for treatment which includes serial casting or splints, range of motion exercises and passive exercises are beneficial for treatment of contractures[4][8]. Early surgical release of soft tissue around the contractures is crucial to prevent deformities[14].

CASE REPORT

A 11 month old girl child with multiple deformities born to P.L2 mother of non-consanguineous marriage was brought for surgical fixation of dislocated hip and Hip spica application to the Department of Orthopedics. She was a preterm baby born at 36 weeks of gestation, delivered by LSCS with birth weight of 2.86kg. She had short stature, neonatal hyperbilirubinemia and was kept in NICU and was given phototherapy and oxygen by mask. Mother had an uneventful medical history. Father and mother were young and healthy. Their first child is 7 years old and healthy. No family history of congenital deformities. No history of drug use. First child defect was noted in 2nd trimester scan. 5th month scan showed Tallipes Equinovarus(TEV) deformity of left foot, 7th month...
scan showed flexion deformity of left wrist, contractures and also polyhydraminosis. At birth the child was found to have Tallipes Equinovarus (TEV) deformity of left foot, Flexion deformity of left wrist, calcaneovalgus deformity of right foot and developmental dysplasia of left hip. Plantar application was done 1 week after birth for left upper limb and 2 weeks later for left lower limb. 4 months later, she underwent posteriomedial soft tissue release for TEV. She was also managed with antibiotics (Syrup cefixin 4.5ml) to prevent infections, analgesics (paracetamol suppository 80+40mg, Syrup paracetamol 2ml) for pain and Zinc oxide paste for abrasions due to Spica cast.

DISCUSSION

Arthrogryposis multiplex congenita is a rare but non progressive congenital disorder which is characterized by multiple joint contractures at the time of birth and can also be accompanied by muscle weakness. It is a clinical manifestation of different syndromes. The condition affects approximately 1 in 2–10,000 live births with no predilection to sex. It has a multifactorial etiology. In our case the child’s family history was normal. The mother of the child had polyhydraminosis at the time of gestation, which can be thought to be a reason for reduced fetal movements in womb thus leading to multiple deformities.

The treatment of AMC is based on patient condition and vary depending on the person. In this case the child underwent serial cast application, posteriomedial soft tissue release for TEV, open reduction of dislocated hip and k wire fixation of left hip, tendoachilltes lengthening and Hip spica application. She was also given antibiotics and analgesics after surgery.

The child had presented rashes which was because of the hip spica cast. Application of zinc oxide paste for rashes was suggested by Pharm D interns. Counselling about the disease, importance of multiple surgeries, the necessity of serial casting and passive exercises for the patient such as abduction – flexion exercise of foot, stretching exercises for arms and legs were explained to parents of the patient by Pharm D interns.

CONCLUSION

Early diagnosis of AMC can be done by prenatal ultrasound scan. The physical therapy should begin early in life in order to improve the range of motion of affected limbs. Soft tissue release surgeries, serial casting, passive stretching exercises, splints will help in improving the affected individuals condition. Appropriate counselling about the condition, the importance of multiple surgeries and physical therapy, should be given to the family members of the affected individual is also important so that they understand how they can contribute to betterment of the patient.

REFERENCES

[13] Dr. D.Y. Shrikhande,etal, Rare Case Series of Arthrogryposis Multiplex Congenita, Pravara Medical Review, December 2016, 8(4) 18-24