Arthrogryposis Multiplex Congenita: A Rare Case Report

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Abstract :

Arthrogryposis Multiplex Congenita is a descriptive term with various etiologies and complex clinical features including multiple joint contractures of various limb joints. It is associated with malformations, malfunctions and neurologic deficiencies. We report the case of a new born term female child admitted at Rural Medical College, Ahmednagar (Maharashtra) with positive family history and evident clinical features of arthrogryposis multiplex congenita. Multidisciplinary management was instituted. This case is presented for its rarity.

Keywords: Arthrogryposis Multiplex Congenita, multiple joint contractures, Club Foot, Teratologic Hip Dislocation, neurologic deficiencies

Introduction: Arthrogryposis Multiplex Congenita is a descriptive term with various etiologies and complex clinical features including multiple joint contractures of various limb joints. It is associated with malformations, malfunctions and neurologic deficiencies. It is associated with abnormal contraction of muscle fibers, causing reduced mobility with a decreased active and passive arc of motion. Arthrogryposis is not a specific diagnosis but a descriptive term with various etiologies and complex clinical features, including multiple congenital contractures of various limb joints. It is associated with over 300 different disorders encompassing malformation, malfunction, and neurologic deficiency. Approximately 1% of all births show some form of contractures of the joints ranging from unilateral clubfoot to the most severe amyoplasia, a condition characterized by pervasive, crippling contractures involving many joints. Although children with arthrogryposis have many other problems, such as micrognathia and feeding issues, focus is on the orthopedic problems frequently seen in this group of children. In the absence of central nervous system lesions, many children have normal intelligence.[1,2]

Case report:

A term, female, small for gestational age baby delivered to a G3P2A1 mother on 15-09-15 at 9:30 AM, with h/o immediate cry after birth at a private nursing home in periphery. She was referred to our institute and presented with abnormal posture.

Detailed clinical examination revealed features of Arthrogryposis Multiplex Congenita due to congenital contracture deformities. Contractures were more marked in lower limbs. There was marked adduction of arms, flexion of elbows, arms across chest and fixed flexion contracture of fingers. There was adduction of thighs, hyper-extension of knees, overlapping of toes, and bilateral club feet, X-ray revealed dislocation of both knees and both hips.(Fig 1 & 2)

Fig 1

Fig 2

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Discussion:

Incidence of arthrogryposis is reported to be 1 per 5000 to 10000 with equal gender ratios.

- This baby was born to parents with 3rd degree consanguinity.
- Mother had bad obstetric history i.e. 1st conceptus had similar features and expired soon after birth. However, their 2nd child is normal, a 2.5 years old boy. During present pregnancy, mother had no h/o DM, myasthenia gravis, infections, drug allergies or multiple sclerosis.
- The generalized stiffness and immobility of newborn with AMC is a sharp contrast to parents expecting a healthy baby. The stiffness and deformities need to be aggressively addressed through a combination of modalities. Team of clinicians including therapists for upper and lower extremities, orthotists and orthopedic surgeons will be involved.
- Her ANC USG revealed no oligohydramnios or uterine anomalies which are known to cause contractures of fetal limbs, nor was there any evidence of fibroids or tumors which are known to impinge on uterine space and prevent movements.
- The therapeutic and orthopedic goal for child with arthrogryposis limb deformity is to achieve maximal joint motion and optimize joint position for function.[3,4]
- Club foot deformities are most commonly seen in arthrogryposis. Clubfoot had hindfeet equinus, midfoot varus and forefoot adduction shortly after birth. It is managed by casting and ponseti method.[5,6]
- Knee issues respond well to therapy and splinting.3
- Teratologic hip dislocations are common within spectrum of arthrogryposis and usually require open reduction of hip.[4]
- Ambulation is undoubtedly delayed in those who achieved it, associated with muscle fatigue and pain.[3,4]
- Parents were counselled regarding prognosis and management of disease. They were reluctant to continue further hospitalization and treatment. Therefore, child was lost to follow up.

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