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Case Report:

Arthrogryposis: A Rare Manifestation in Infant of Diabetic Mother

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Abstract:
Arthrogryposis multiplex congenita is characterized by non-progressive, multiple joint contractures present at birth. The major cause of arthrogryposis is fetal akinesia due to fetal abnormalities like neurogenic, muscle, connective tissue abnormalities or maternal disorders Here we report a rare case of arthrogryposis in infant of diabetic mother with multiple congenital anomalies.
Key Words: Infant of diabetic mother, Arthrogryposis, Lower limb

Introduction:

Infants of diabetic mother (IDM) are at an increased risk of morbidity and mortality related to the respiratory distress, growth abnormalities, hyperviscosity secondary to polycythemia, hypoglycemia and congenital malformations. Central nervous system, renal, cardiovascular, and gastrointestinal anomalies are more frequently found in the IDM babies.[1] The overall risk is 8-15%, with 30-50% of perinatal fatalities related to major congenital malformations. Poor glycemic control early in pregnancy directly correlates with a higher incidence of congenital malformations.[2] Previous studies suggest that the birth defects in IDMs may be related to reduced arachidonic acid and myoinositol levels and elevated sorbitol and trace metal levels in the fetus.[2-4] Here we report a neonate who was born to mother affected with type 2 diabetes had arthrogyrosis with multiple congenital anomalies..

Case Report:

The full term female newborn was product of a non-consanguineous parent delivered by emergency cesarean section due to breech presentation with inadequate pelvis with previous scar to a 32 year old second gravid mother. The newborn had an agar score of 6,9,10 at 1, 5, and 10 minute. The pregnancy was complicated by Type 2 diabetes in mother and she was on insulin injection. The birth weight of the child was 3123gm, head circumference 35 cm and approximate length was 44cm.

On physical examination, the infant had characterstic facies with depressed nasal bridged, widely open mouth and low set ear with preauricular tags on left side. Temperature was 37.2º C, heart rate 124 beats/minute, and respiratory rate 54/ minute. Anterior fontanel was open. Hypospadiasis with left sided -

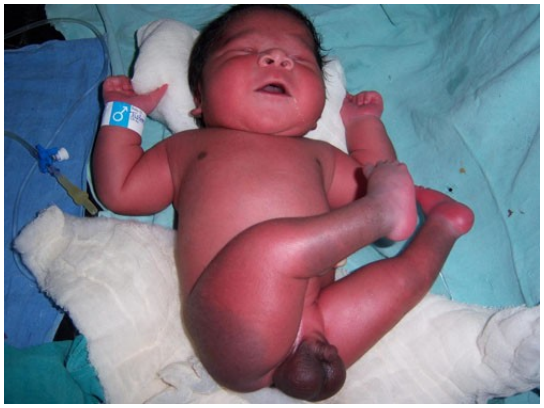

Figure 1: Photograph shows infant of diabetic mother baby with lower limb arthrogryposis

inguinal hernia was present. Lower limbs were arthrogrypotic characterized by the hip was fixed in flexion, knees were extended, ankles were dorsi-flexed and the foot had eqiunovarus deformity (Fig.1). Bilateral congenital hip dislocation with restriction of the lower limb movement was present. Shortening of the lower limb was present. No history of similar illness was found in the family. Based on these skeletal abnormalities, she was diagnosed as arthrogryposis. On cardiovascular examination, soft systolic murmur was present. Other systemic examination was within normal limit. On investigation, complete blood cell count was: Hb: 15gm%, TC: 10,600/cmm (LY32%, MO 10%, GR 58%) and platelet count: 5.1 lac/cmm. Transthoracic Colour Doppler echocardiographic revealed small size patent ductus arteriosus (PDA). Infantogram showed sacral agenesis. Neurosonography revealed normal brain parenchyma with a normal ventricular system. USG abdomen and pelvis reported normal. We conservatively managed the neonate and discharged after 7 day without any complication.

Discussion:

Arthrogryposis multiplex congenita (AMC) is a congenital anomaly characterized by non-progressive, multiple joint con-

tractures present at birth. In the classic form of AMC all four limb are involved, but the condition can also occur in the upper or lower limb. An autosomal dominant variant called distal arthrogryposis involves the hand and feets with severe deformation.[5] Multiple joint contracture occur at birth because of limitation of movement in utero and the causative factors like a) Neuropathy b) Myopathy c) Abnormal connective tissue involving joints d) In utero restraint and e) Maternal illness.[6] Infants born to mothers affected with myotonic dystrophy, myasthenia gravis, or multiple sclerosis are at risk of AMC. After extensive search in medical literature no single case is reported that infant of diabetic mother are at risk of arthrogryposis. In our case, IDM baby who had lower limb arthrogryposis along with multiple other congenital anomalies was present.

The clinical manifestations in arthrogryposis is mainly involved the extremities with thin subcutaneous tissue and absent skin creases. Symmetrical deformities increases distally, with the hands and feet deformed and joint dislocation, especially the hips. Normal sensation and deep tendon reflexes may be diminished or absent. Contractures, especially of distal joints are affected more frequently than proximal joints. Limb deformities include pterygium, shortening, webs, compression, dislocated radial heads, and dimples. It is very rare that upper limb is spared and only lower limb is affected which was present in our case. Other deformities include scoliosis, genital deformities, and hernia (inguinal, umbilical). There may be many other malformations of the skeleton, respiratory tract, urinary system and nervous system.[4-6]

The incidence of congenital anomalies mainly cardiac malformation and lumbosacral agenesis is increased threefold in IDM. Other anomalies include neural tube defect, hydronephrosis, renal agenesis, duodenal or anorectal atresia, situs inversus, double ureter and holoprosencephaly. Congenital anomalies in IDM babies correlelate with poor metabolic control during the periconception and organogenesis periods and may be due to hyperglycemia induced teratogenesis.[7] The mechanism by which hyperglycemia disturb embryonic development is controversial, but reduced arachidonic acid and myoinositol levels and accumulation of sorbitol and trace metals in the conceptus have been reported.[7-8] X-ray of all joints may show bony abnormalities. Neurosonography can help in evaluating the CNS and abdomoinal sonography is helpful for other viscera for anomalies. CT scan and MRI can be used to evaluate the CNS and the muscle mass. Lower-limb alignment and establishment of stability for ambulation and upper-limb function for self-care is the main goal. Improves passive and active range of motion if early manipulation started soon after birth. Physical therapy to improve the range of motion in and stretch surrounding tissues is very useful, especially in amyoplasia and distal arthrogryposis.[9] Surgery is rarely needed to correct soft tissue contractures and joint deformities.[10] In conclusion, lower limb arthrogryposis is a very rare manifestation in infant of diabetic mother and should be managed conservatively.

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