Research Paper

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Case Report: A Rare Case of Arthrogryposis Multiplex Congenita

Medical Science

KEYWORDS :

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ABSTRACT

Arthrogryposis multiplex congenital (AMC) is a rare congenital disorder which has multiple joint contractures involving more than one area of the body.Associated with different disorders like neurocognitive delay and malformations .Overall prevalence of 1:3000 live births.Antenatal ultrasound examination can establish the correct diagnosis. We would like to report case with the characteristic clinical & sonographic feature of fetus with AMC (Arthroglyposis multiplex congenita). A 35 years old G4P2L1D1A1 was referred for the first ultrasound scan at 19 weeks of gestation.Ultrasound examination of the present pregnancy at 19 weeks gestation demonstrated a single, live intrauterine fetus corresponding to 19 weeks. There was fixed flexion deformity of wrist, choroid plexus cyst, dextrocardia with associated right ventricular hypertrophy & right atrial enlargement. No normal limb movement noted..Bilateral fixed flexed deformity noted at elbow, wrist and interphalyngeal joints. Lower limb were in extended with everted foot persistently.Based on these findings arthrogryposis Multiplex congenita with cerebellar hypoplasia was made and the patient was advised for termination of pregnancy. Our aim is to present a case of rare incidence and aslo to highlight the fact that anomoly scan is an integral part of antenatal follow up around 18-22 weeks of gestation, so that if any anamoly is detected, it is possible to terminate the pregnancy within the period approved by MTP Act.

INTRODUCTION:

Arthrogryposis multiplex congenita (AMC) is a rare congenital disorder which has multiple joint contractures involving more than one area of the body. Overall prevalence of 1:3000 live births[1]and others say it is one in 11000 -12000 among European live births[2] It is just a clinical finding rather than a specific diagnosis, associated with different disorders like neurocognitive delay and malformations.[3] It is a nonprogressive disease. It occurs mainly due to fetal akinesia which may be because of multiple factors like neurogenic / myopathic process, a connective disorder, intrauterine compression, a vascular insult / teratogenic exposure.[3].Almost every joint in a patient with arthrogryposis is affected,because in 84% all limbs are involved, in 11% only the legs and in 4% only the arms are involved.[4] Antenatal ultrasound examination can establish the correct diagnosis. We report case with the characteristic clinical & sonographic feature of fetus with Arthrogryposis multiplex congenita(AMC).

CASE: A 35 yr old woman with G4P2L1D1A1 was referred for the first ultrasound scan at 19 week of gestation. Her past medical history was uneventful. Her first pregnancy ended up in spontaneous abortion at 2nd month of gestation (reason not known). While second pregnancy ended up with IUD at 8th month of gestation (reason not known). Now she has living normal third child which was PTVD. Her marriage is second degree consanguinous. Her husband was healthy. No familial history of any congenital disorders. There was no history of drug use. Ultrasound examination of the present pregnancy at 19 week gestation demonstrated a single, live intrauterine fetus corresponding to 19 week. There was fixed flexion deformity of wrist, choroid plexus cyst, dextrocardia with associated right ventricular hypertrophy & right atrial enlargement. No normal limb movement noted. Bilateral fixed flexed deformity noted at elbow, wrist and interphalyngeal joints. Lower limb were in extended with everted foot persistently. Based on these findings Arthrogryposis Multiplex Congenita with cerebellar hypoplasia was made and the patient was advised for termination of pregnancy. The pregnancy was terminated after explainaing the parents about the condition and poor outcome of pregnancy. The pregnancy was terminated with misoprostol tablet. Gross specimen showed the following features: Microcephaly with a space which admits only a tip of finger in anterior fontenella, Left eye cornea looks opaque, undescended testis and micropenia. Bilateral flexion deformity at elbow joint, radial decussation, flexion deformity at 3rd, 4th and 5th proximal interphalyngeal joints noted. Bilateral fixed flexion deformity of hip and knee and everted foot observed. Neck, Chest, Abdomen and Spine was normal.

TREATMENT:

Early detection on the basis of high suspicion.Parental counselling and consent for termination of pregnancy and pre conceptional counselling for next pregnancy.Termination of pregnancy before 20 weeks of gestation as per MTP Act.

CONCLUSION:

We present this case of its rare incidence and also to highlight the fact that detailed anamoly scan is an integral part of antenatal follow up around 18-22 weeks of gestation, so that if any anamoly is detected, it is possible to terminate the pregnancy within the period approved by MTP Act. Due to associated conditions in athrogryposis multiplex congenita, the prognosis is poor; termination should be considered when diagnosis is known before viability.

REFERENCE

1 Bamshad,Michael;2009.Arthroglyposis:A Review and Update.The Journal of Bone and Joint Surgery(American)91(suppl 4):40-6. | 2 Hoff,Jana Midelfart;Loane,Maria;Gilhus,Nils Erik;2011.Arthrogryposis multiplex congenital:a epidemiologic study of nearly 9 million births in 24 EUROCAT registers.European Journal of obstetrics and gynaecology and reproductive biology 159;347-350. | 3 'The Free Dictionary:Arthrogryposis'.retrieved 11 july 2013. | 4 Bevan,Wesley P;Hall,Judith ; Bamshad,Michael;Staheli,Lynn T;Jaffe,Kenneth M;Song,kit 2007.Journal of paediatric orthopaedics 27 (5):594-600. |