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Obstetrics and Gynaecology Section

Successful Pregnancy Outcome in a Case of Muscular Dystrophy

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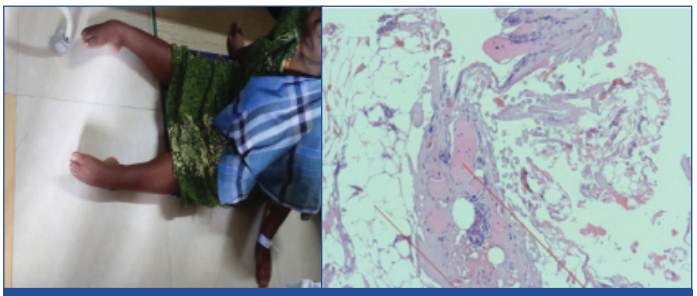
A 29-year-old primigravida female was referred as a case of asymmetrical IUGR and oligohydramnious at 39 weeks six days of gestational age with quadriparesis. She had a history of one episode of convulsion at 13 years of age followed by progressive weakness of upper limbs followed by lower limbs and proximal muscle weakness was greater than distal muscle weakness. The patient movement was restricted and she had to move by sitting and pushing herself through hands since last one month [Table/Fig-1]. On examination, bulk of both muscles were normal, power of both upper and lower limbs were, 0/5 as measured by Medical Research Council (MRC) scale [1]. Plantar reflexes were withdrawal and sensory sensation was intact. Vitals were as follows pulse rate was 78 per minute, Blood pressure was 110/70 mm of Hg and respiratory rate was 14 per minute. Respiratory system and cardiovascular system examination were within normal limits. On obstetrics examination, per abdomen-Fundal height was around 32 weeks, with cephalic presentation. Fetal Heart Sound (FHS) was good, per vagina-bishop score was 3 and pelvis was adequate. Doppler was normal and after counselling with the patient and her husband an attempt of vaginal delivery was made. Ripening was carried out with Foley's catheter in view of Intra Uterine Growth Restriction (IUGR) and decreased foetal movement. Post ripening, oxytocin was started however; emergency LSCS was done under general anaesthesia because of fetal distress. The caesarean and recovery from anaesthesia was uneventful. There were no complications like pre-term labour, pre-eclampsia and postpartum haemorrhage.

Atrophied rectus muscle was replaced by fatty tissue which was seen during operative period and a sample of tissue was taken for biopsy. There was no improvement in muscle weakness in postoperative period. On investigating her serum Creatine Kinase (CK- MB) value was 22, creatine kinase (CK-T) value was 306 and Lactate Dehydrogenase (LDH) value was 946. Echocardiography (ECHO) showed normal valves, chambers and normal ejection functions but her electromyography was not done.

Rectus muscle biopsy showed muscle fibres replaced by adipose tissue, fascicular architecture was not maintained and few hyalinised round fibres were seen [Table/Fig-2].

In our patient muscle weakness was started at age of 13 years and it was progressive symmetrical weakness and proximal weakness. Her sensory system was normal. No evidence of facial, ocular, bulbar and neck muscle weakness was seen. No family history of quadriparesis. So it is most probably a case of autosomal recessive variety of limb girdle dystrophy. Differential diagnosis include Duchenne's muscular dystrophy, Becker's muscular dystrophy, myotonic dystrophy but Duchenne's and Becker's muscular dystrophy both are X-linked recessive disorders and are rare in female patients. Myotonic dystrophy presents with progressive weakness of face, which was absent in our patient, so it was excluded. Limb girdle dystrophy, is an extremely rare disorder with the incidence quoted as <1/100,000



[Table/Fig-1]: Typical position of the patient in case of limb girdle dystrophy.
[Table/Fig-2]: (H&E 10X) Histopathology image of rectus muscle showing adipose tissue and hyalinised round fibres. Straight line= adipose tissue and arrow line= Hyalinised round fibres.

[2]. It may be inherited in an Autosomal Recessive (AR) (90%) or dominant (10%) fashion [3]. Limb girdle dystrophy typically manifest with weakness of pelvic and shoulder girdle muscles. Respiratory insufficiency and cardiomyopathy may occur as results of weakness of muscles. In limb-girdle muscular dystrophy muscle fibres slowly break down and are replaced by connective tissue and fat. Muscle degeneration leads to muscle weakness and atrophy. The name of the condition refers to the muscles affected, mainly those around the pelvic and shoulder girdles, also known as the limb-girdles. The clinical course of this group of muscular dystrophies is variable. Onset may be from the first to the fourth decade of life with severe forms of the disease beginning in early life and late-onset disease running a milder course [4]. In our case there were no signs and symptoms of respiratory insufficiency and cardiomyopathy. Very few case reports have been reported on patients of limb girdle Dystrophy and pregnancy outcome [5]. One such case was reported by Ayoubi JM et al., they reported a case of limb girdle dystrophy and pregnancy which was delivered by vaginal delivery [6]. Another case was reported by Von Breunig F et al., where they had managed a case of limb girdle dystrophy in pregnancy with LSCS [7]. Pregnancy in case of muscular dystrophy is rare and challenging to manage. But with multidisciplinary modality approach with a team of obstetricians, anesthesiologists, neurologists and ICU care we can manage such cases successfully.

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