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Limb-Girdle Muscular Dystrophy Associated with Fatness: A Case Report and Literature Review

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ABSTRACT

Limb-girdle muscular dystrophy LGMD is a rare genetic disorder characterized by progressive muscle atrophy in the scapular and pelvic regions. Associated with pregnancy, LGMD represents a management challenge, because of pre-existing or exaggerated pregnancy complications. In the light of a clinical case and through a review of the literature, we will discuss the different aspects of this pregnancy-related pathology.

Keywords

Limb-girdle muscular dystrophy, Pregnancy, Twins.

Introduction

Limb-girdle muscular dystrophy (LGMD) is a condition first described by Walton and Nattrass in 1954. It is an extremely rare progressive muscle disorder with an incidence less than 1 per 100,000. Limb-girdle muscular dystrophy is characterized by weakness and atrophy of the shoulder and hip muscles. It is a genetic disease that can be transmitted in an autosomal recessive mode in 90% of cases, or dominant in 10% of cases [1-3]. LGMD may be accompanied by impaired cardiac function, and has restrictive lung disease. This pathology exposes certain challenges during pregnancy. The literature is poor in this sense and there are few case reports of successful management.

Case Report

Mrs. C.S., 26 years old, primigravida, nulliparous, with a history of limb-girdle muscular dystrophy for which she had been followed up since the age of 5 years, and who presented to us for pregnancy follow-up. The patient was in a wheelchair, the first clinical examination revealed a patient in a fairly good general condition, with impossibility to stand up, and difficulty to sit down, with a slight thinning of the shoulder musculature, the tone was otherwise preserved. In addition, the patient presented a spinal deformity of the type of scoliosis. She had shortness of breath on effort without respiratory discomfort. The electromyography performed as part of the diagnostic workup for her pathology showed signs of early myopathy. The muscle biopsy showed a myeloid degeneration with subtle myopathic changes. The creatinine phosphokinase level was elevated to 412 u/l. The pregnancy was estimated at 19 weeks of amenorrhea. Obstetrical ultrasound showed a progressive twin pregnancy, bi-chorionic and bi-amniotic, with biometrics corresponding to the estimated term of the pregnancy. An echocardiogram was performed, showing a correct left ventricular ejection fraction of 65%. The pulmonary evaluation revealed a slight restrictive syndrome. Pregnancy proceeded without abnormalities. Routine paraclinical examinations revealed no abnormalities. Respiratory and cardiac function remained stable. The obstetrical ultrasound of the third trimester showed an evolving twin pregnancy. The first twin was in transverse presentation and the second in breech. The biometries corresponded to 34 SA for both twins. At 37 weeks and 2 days, the patient went into labor spontaneously. She was admitted to our training center for delivery. The obstetric examination revealed a cervix dilated to 1 cm, with a water sac that had broken two hours

before admission. The amniotic fluid was clear. The first twin was still in transverse presentation, so a cesarean section was indicated under spinal anesthesia, which proved to be technically difficult. She gave birth to two healthy male babies with a 5-minute Apgar score of 09/10 and 10/10 and a birth weight of 1950g and 1800g respectively. The cesarean section was uneventful. However, during the incision, there was significant thinning and sagging of the abdominal wall muscles. The post-operative course was simple. Thromboprophylaxis was instituted. The patient had been in intensive care for three days and was transferred to an inpatient unit for four days. The patient received continuous support to improve her mobility after delivery.

Discussion

Limb-girdle muscular dystrophy LGMD is a rare genetic disease affecting the muscles of the scapular and pelvic girdles [4]. These muscles will be disintegrated and replaced by connective and fibrous tissue, resulting in muscle atrophy associated with myasthenia [5]. The severity, age of onset, and characteristics of limb-girdle muscular dystrophy vary among the subtypes of this disorder. Symptoms can appear at any age, and worsen with time [6].

During pregnancy, there is a progression of LGMD. It therefore presents several challenges to the pregnant woman [7]. During pregnancy, the maternal heart has the added burden of supplying blood to the placenta, which can lead to a deterioration of the maternal condition if cardiac function is already compromised. The same is true for respiratory function, as the fetus grows throughout the pregnancy, the diaphragm will be compressed by the gravid uterus, which can decrease respiratory capacity. Preterm cesarean section may be necessary due to the risk of hemodynamic complications, and therefore a high rate of cesarean section would be associated with this condition [8]. However, a case of successful natural delivery has been reported in the literature in a multiparous woman with LMGD who did not have respiratory failure [9]. The lumbar lordosis exaggerated by pregnancy makes spinal anesthesia technically difficult. Patients with this condition are sensitive to anesthetic agents, including nondepolarizing muscle relaxants, analgesics, and sedatives. Patients may also develop malignant hyperthermia with succunylcholine or inhalation anesthetics [7].

The largest study to date of pregnant LGMD patients includes 22 parturients, giving birth to 29 children. In this study, a high proportion of breech presentation was noted, which may be related to mobility during pregnancy and may explain the increased rate of caesarean section associated with this disease [10,11].

In our case, the patient underwent a cesarean section at a term of 37 SA and 2 days from spontaneous labor, given the transverse presentation of the first twin that imposed the mode of delivery. The operation was performed under spinal anesthesia despite the technical difficulty encountered due to the spinal deformity.

In patients with impaired cardiac function, management must be multidisciplinary, essentially by a team of obstetricians, cardiologists, pulmonologists, and anesthetists. An echocardiogram should be performed before, at the beginning and at least three times later in the pregnancy depending on the symptomatology. A breath test should be performed in the second part of pregnancy to assess the impact of the gravid uterus on respiratory function, as well as to decide on the mode of anesthesia later.

Fetal monitoring should be close by growth ultrasound. Low weight for gestational age has been described in women with GDM. A risk of uteroplacental insufficiency and fetal growth restriction should be sought especially in the presence of compromised cardiac function [12].

Close postpartum monitoring is important, even if the maternal condition remains stable. Because of the heterogeneity of the pathology, and the paucity of the literature in this sense, obstetrical management must be adapted to the individual case.

Conclusion

The management of a pregnant woman with LGMD must be multidisciplinary. Associated with pregnancy, this pathology presents a challenge for the medical team to establish an appropriate follow-up and delivery strategy, especially in the presence of complications.

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