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Congenital Myasthenia Gravis in Newborn

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ABSTRACT

Congenital myasthenia gravis are very rare, which is an autosomal recessive disease originating from rare genetic disorders. Symptoms of congenital MG usually start at birth and are lifelong.

Our case weakness started from birth, need respiratory support and failure of extubation many times, and absent fetal movements with positive family history his cousin was neonatal myasthenia gravis and died.

Keywords

Genetic disorders, Congenital myasthenia gravis.

Introduction

Myasthenia gravis is either transient, which was mother affected and antibodies, are transferred to the baby and take time to get rid of the body, and it is present between 3rd and 10th day of life with poor feeding and hypotonic.

Congenital myasthenia gravis which severe type and autosomal restive, other siblings were affected like our case presented antenatal by decreased fetal movements and respiratory depression needing a mechanical ventilator and feeding problem. The prognosis is very poor due to the ventilator depend and recurrent infection and generalized hypotonia and feeding gavage leading to malnutrition.

Incidence

In general, neonatal MG is noted to be rare and may present in 10% to 15% of newborns born to mothers with MG. However, the risk of neonatal MG in a sibling is significantly higher in subsequent pregnancies. There is no reported race or gender preference. There is no definite association between disease severity in the mother and the clinical presentation in the newborn [1].

Types of Myasthenia Gravis

We have four type of myasthenia gravis:

1. Congenital myasthenia gravis is autosomal ressive.
2. Neonatal myasthenia gravis.
3. Juvenile myasthenia gravis.
4. Adult myasthenia gravis.

Differential Diagnosis

1. Spinal muscular atrophy.
2. Congenital muscular dystrophies.
3. Infantile myotonic dystrophy.
4. Mitochondrial myopathy.

Case History

32 years old para4+o was delivered by caesarian section due to fetal bradycardia, the baby was delivered flat, with bradycardia, with an APGAR score of 2,6.8 needed positive pressure and intubation and shift to the intensive care unit.

The baby was not dysmorphic, pink hypotonic with forge position, weight was 3:00kg and other parameters are within 5th percentile. Equal breath sound bilateral, 1st and 2nd heart sounds were normal no murmurs.

No hepatosplenomegaly and bowel sound is present, No joint contraction, the tone is decreased no sedation CBC was normal, chemistry was normal, CPK normal and liver functions and renal function were normal CBG was normal, chest –x-ray there was thymus shadow was normal, and no cardiomegaly. MRI brain no major anomalies, brain was normal. Baby admitted in intensive care unit, intubated and started antibiotic ampicillin and gentamycin for 48hour, asked for further history to explain hypotonia, Mather denied any sibling or casinos had hypotonia.

Consultation was done to metabolic and genetic consultant who advise to do Whole exome sequencing kit –DNA Sequencing Home Product study to determine the cause of hypotonia, and father gave history of one of sibling had same disease and died few year back. Baby still on mechanical ventilator, failure of extubation many times because weak muscle and drooling of secretion, she developed right lung collapse was managed by good physiotherapy and suction. Baby developed stridor, saw by ENT consultant advised neck virtual CT scan which reported normal.

Neurology consultant saw the baby and examine him to roll any abnormality. The baby still on Mechanical Ventilator, full OGT feeding. The result of Whole exome sequence came showed the boy had Congenital myasthenia gravies severe types so we started Anticholinesterase medications to treat myasthenia gravis include anticholinesterase agents such as mestinon or pyridostigmine, which slow the breakdown of acetylcholine at the neuromuscular junction and improve neuromuscular transmission and increase muscle strength.

Diagnosis

Congenital myasthenia gravis.

Discussion

Neonatal myasthenia gravis this is an autoimmune disorder and affects neonates born to mothers with autoimmune myasthenia gravis. Neonatal MG occurs due to the transplacental passage of antibodies directed against various neuromuscular junction antigens. The most common antigen is the nicotinic acetylcholine receptor (AChR). Another important antigen is the muscle-specific receptor tyrosine kinase (MuSKR) [2].

Congenital Myasthenia gravis is rare disease, it is autosomal ressive and the mother was completely healthy, the disease is affect newborn shortly after birth or antenatal by decrease fetal movements and baby delivered hypotonic, respiratory depression need respiratory support and very difficult to wean from mechanical ventilator. Most of the babies with congenital myasthenia were not response to the treatment and end with ventilator dependent, tracheostomy and ventilator at home.

Social support is very important to the parents; it is multidisplenary need perinatologists, neonatologists, and specialty trained nurses respiratory therapy and dietarians, neurologist and physiotherapy to optimize the care of these patients.

Transient myasthenia gravis is participated by stress and infections otherwise completely healthy. Neonatal myasthenia, mother was affected and antibodies transferred from mother to the baby and most of the baby presented at the age of 3rd and 7th day and they take time to get ride from this antibody and the tone is improving. There was no correlation between severity of the disease and affected newborns and there was no sex prevalence both male and females were affected.

Reference

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