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Autologous Stem Cell Transplantation to Treat Emery- Dreifuss Muscular Dystrophy

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Abstract

Emery- Dreifuss muscular dystrophy affects muscles such as Skeletal muscles, Cardiac muscles which is named after Eglin H. Emery and Fritz Emery- Dreifuss. Here we are going to demonstrate Autologous Stem cell transplantation done in our hospital to treat this dystrophy and discuss about post transplantation follow up out come

Key words: Muscular dystrophy, Emery-Dreifuss, Skeletal muscle, Cardiac muscle.

Introduction

Emery- Dreifuss muscular dystrophy is a condition that affects skeletal muscles and cardiac muscle. It is caused by mutation. This is x- linked autosomal dominant or autosomal dominant recessive fashion. The mutation are caused in Emery- Dreifuss muscular dystrophy and LMNA, FHLI genes. Joint symptoms are present in child hood and involve contractures of neck,elbows ankles ,children and adults with this dystrophy usually experience slowly worsening of muscle wasting and muscle weakness. By adulthood many people with this type of dystrophy develop cardiac problems such as arrhythmias and conduction defects. Here, we are going to demonstrate a male boy with age 18 who has muscle weakness in upper arms and lower legs and hips. He has elevated serum creatine phosphokinase levels with 2000 U/L and myotonia and pectus excavatum and a detailed explanation about autologous stemcell therapy was given to him and his parents with its pros and cons. A written consent was obtained from the patient and his parents to proceed for autologous stemcell therapy .Preoperative blood investigations and cardiac checkup was with in normal limits and the patient was kept Nill by mouth 6hrs before the procedure to ensure that he will be not as aspirated during general anesthesia. Under general anesthesia 100ml bone marrow was aspirated and 126 $x10^6$. Autologous stemcell were isolated under sterile conditions and were injected into all affected muscles deeply. Cardiac muscles were excluded because he didn't develop any cardiac manifestation. For cardiac muscles it is standardized protocol to inject autologous stemcells into coronary artery .The patient was recovered from anesthesia and he was kept on antibiotics and anti inflammatory medication for 10 days. On the next day he was discharged from the hospital as there were no post operative complications. He was followed for 2 yrs for every 15 days as his upper and lower limb power was improved gradually.

Conclusion

This autologous stemcell transplantation is a milestone in medical field particularly in neurological disorders especially in this Emery-Dreifuss muscular dystrophy.

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