



Scenario of MARFANS Syndrome in a Kashmiri Family

Gowhar Ahmad*

Professor, Department of Ophthalmology, Florence Hospital Multispeciality Center, Chanapora, India

*Corresponding Author: Gowhar Ahmad, Professor, Department of Ophthalmology, Florence Hospital Multispeciality Center, Chanapora, India.

DOI: 10.31080/ASWH.2024.06.0590

Received: June 03, 2024

Published: June 20, 2024

© All rights are reserved by **Gowhar Ahmad.**

Abstract

MARFANS syndrome is a kind of mesodermal entity involving eyes, skeletal muscular system and CVS. It is a kind of syndrome complex. Involving bones, cartilage, joints, bones, ligaments, blood vessels, skin, muscles, eyes, ears, valves, joints, so we have 3 major manifestations of eyes, CVS system and skeletal muscular system. One may not have manifestations of all three; their could be manifestations if one or two systems in that case it would be known as marfanoid stroma.

Ophthalmic manifestations are Subluxation and dislocation of lens, pseudophakia, ECTOPIA LENTES, nystagmus, squint, cataracts, KERATOCONUS, High myopia, retinal detachment. Skeletomuscular manifestations are Tall person, high arched palate, tower skull, macroglossia, long tapering fingers, span of arm is greater than height, Mera carpal index is positive, arachnodactyly, kyphosis, scoliosis, pigeon-shaped chest, tendency for fractures. CVS manifestations are Aortic dilatation, Mitral valve prolapse syndrome, Severe mitral regurgitation, PDA, Atrial septal defects.

Keywords: Ophthalmic, Skeletomuscular; CVS Manifestations; MARFANS Syndrome; Marfanoid

Introduction

MARFANS syndrome is a kind of syndrome complex, a mesodermal entity characterized by ophthalmic, skeletal muscular and CVS manifestations.

One may have one or two syndrome complex involvement.

Material and Methods

Are patients their families having features of MARFANS SYNDROME.

Ophthalmologist, surgeon, orthopedic surgeon, cardiologist, dermatologist play an important part in this syndrome complex.

Results and Discussion

They depend upon the preservation of ophthalmic, skeletal muscular and CVS involvement.

Conclusion

Is presentation of all three involvement or one or two and necessary treatment depending upon the preservation [1-13].

Bibliography

1. Carter N., et al. "Bone mineral density in adults with Marfan syndrome". *Rheumatology (Oxford)* 39.3 (2000): 307-309.
2. Ha HI., et al. "Imaging of Marfan syndrome: multisystemic manifestations". *Radiographics* 27 (4): 989-1004.
3. Rhee D., et al. "Incidence of aortic root dilatation in pectus excavatum and its association with Marfan syndrome". *Archives of Pediatrics and Adolescent Medicine* 162.9 (2008): 882-885.
4. Beals RK and Mason L. "The marfan skull". *Radiology* 140.3 (1981): 723-725.
5. Magid D., et al. "Musculoskeletal manifestations of the Marfan syndrome: radiologic features". *American Journal of Roentgenology* 155.1 (1990): 99-104.
6. Oosterhof T., et al. "Quantitative assessment of dural ectasia as a marker for Marfan syndrome". *Radiology* 220.2 (2000): 514-518.
7. De paep A., et al. "Revised diagnostic criteria for the Marfan syndrome". *American Journal of Medical Genetics* 62.4 (1996): 417-426.
8. Ruano MM., et al. "MR imaging in a patient with homocystinuria". *American Journal of Roentgenology* 171.4 (1998): 1147-1149.
9. Stuart AG and Williams A. "Marfan's syndrome and the heart". *Archives of Disease Child* 92.4 (2007): 351-356.
10. von Kodolitsch Y and Robinson PN. "Marfan syndrome: an update of genetics, medical and surgical management". *Heart (British Cardiac Society)* 93.6 (2007): 755-760.

11. Tunçbilek E and Alanay Y. "Congenital contractural arachnoidactyly (Beals syndrome)". *Orphanet Journal of Rare Diseases* 1 (2006): 20.
12. Gutierrez J., et al. "Dolichoectasia—an Evolving Arterial Disease". *Nature Reviews on Neurology* 7.1 (2011): 41-50.
13. Dyhdalo K and Farver C. "Pulmonary Histologic Changes in Marfan Syndrome". *American Journal of Clinical Pathology* 136.6 (2011): 857-863.