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Case Study

Severe periodontitis associated with Marfan syndrome- an existence or a co-existence.

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ABSTRACT

Marfan syndrome is the most common dominant autosomal genetic disorder of connective tissue origin. Over the years various diagnostic criteria have been developed for this entity, the earliest among them is the Berlin Nosology, which classifies this condition mainly based on its physical characteristics.Oral manifestations associated with this condition are poorly documented and periodontitis associated with Marfan syndrome is still a controversy. This article report a case of Marfan syndrome with periodontal involvement, the patient was successfully managed with nonsurgical periodontal therapy.

Keywords: Connective tissue disorders, Marfan's syndrome,Dolichocephalia,Fibrillin**.**

Introduction:

Marfan Syndrome (MFS) was first described by an pediatrician Antoine Bernard-Jean Marfan, who reported an out of proportioned length of the lower limbs and fingers.[1] Marfan syndrome is the most commonly inherited connective tissue disorder with equal distribution between the sexes.[2]

Marfan syndrome is caused by an autosomal dominant mutation in the gene encoding fibrillin (FBN1, chromosome 15q15). Fibrillin is a glycoprotein that is an integral part of the connective tissue in the body.[3] The normal fibrillin inhibits the growth of the long bones and elastic fibers, through its tension control. As fibrillin being altered, an exaggerated bone overgrowth is produced.[4] They also produce defects in various locations such as the ocular lens suspensor ligament, lungs, blood vessel walls and apparently, the periodontal ligament. Hence Marfan syndrome can be considered as a multisystemic disease where localization and degree of symptoms differ among individuals. The Periodontal findings are found in accordance with bacterial plaque and manifests as severe form of both horizontal and vertical bone resorption.[5] We report two cases of marfan's syndrome with periodontal manifestations.

Case report

A 26 year old woman reported to us with a complaint of swollen gums in relation to lower front teeth region. Clinical examination suggested presence of signs and symptoms such as flat feet [fig 1], arachnodactyly [fig2], dolichostenomelia, ectopia lentis[fig 3] and mitral valve prolapse. The patient also reported positive family history, positive wrist or Walker's sign [fig 4]

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and positive thumb or Steinberg sign [fig 5]. Considering the above findings as per Berlin's criteria, the patient was diagnosed to have Marfan syndrome.Intraoral examination revealed inflammation of the gingiva and destruction of the periodontium [fig 6], high arched and narrow palate, generalized bleeding on probing and 5mm of probing pocket depth in relation to 12, 13, 16, 22, 23, 26, 31, 36, 41 and 46. On examination of OPG there was horizontal bone loss in relation with 12, 22, 16, 26, 36, and 46.

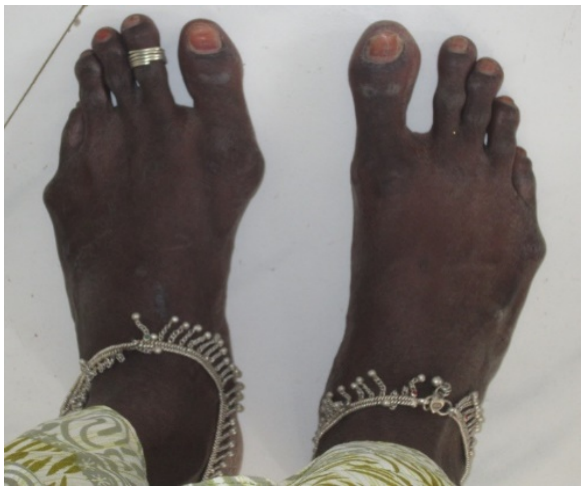

Figure 1: Extra oral photograph showing flat feet

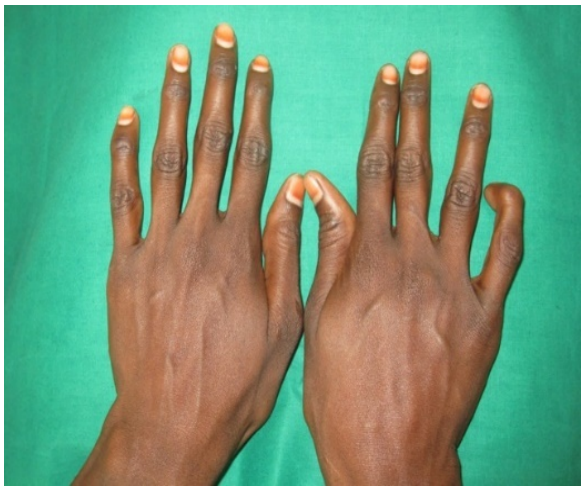

Figure 2: Extra oral photograph showing arachnodactyly.

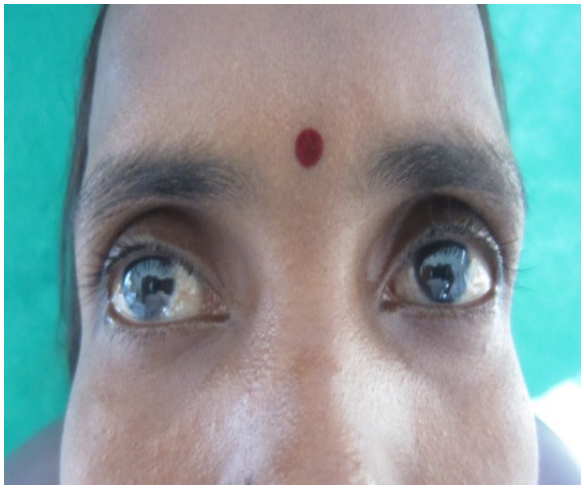

Figure 3: Extra oral photograph showing ectopia lentis.

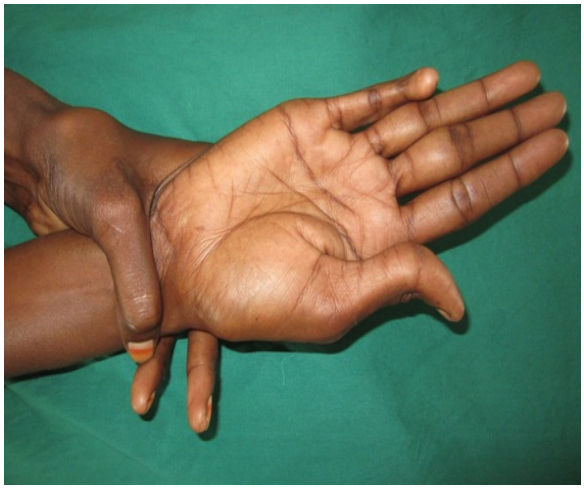

Figure 4: Extra oral photograph showing positive wrist sign

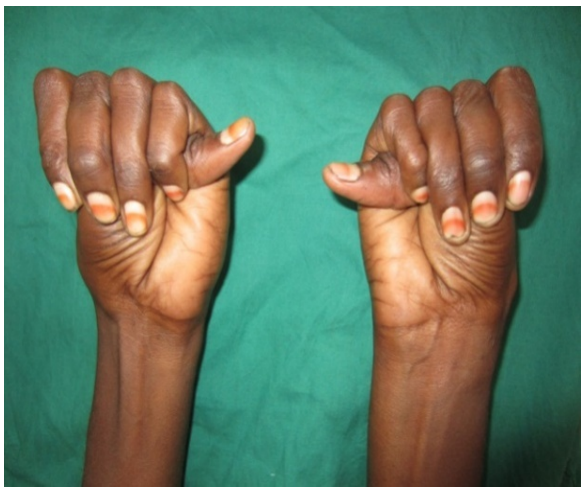

Figure 5: Extra oral photograph showing positive thumb sign

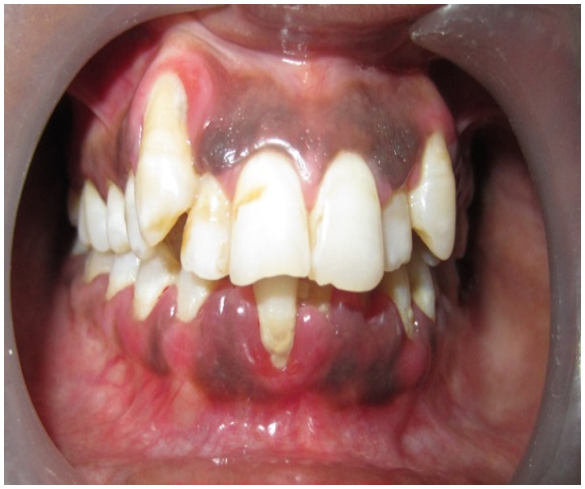

Figure 6: Intra oral photograph showing severe periodontitis involving 13, 23 and lower anteriors

Discussion

Marfan syndrome is a rare multifaceted disorder with a worldwide incidence of 7-17 per 100,000 individuals.[6]Over the years, various diagnostic criteria have been evolved for the diagnosis of MFS. As early as in 1956, McKusick developed a criterion which was then modified with certain minor revisions in 1979. But then the most widely accepted classification was developed in 1986, famously called as Berlin nosology. The drawback of the above criterion is that it's purely clinical and does not have any molecular basis, hence conditions which resembles marfan clinically has to be ruled stringently. [7] Considering this a revised diagnostic criterion was proposed by Paepe et al in 1996, where additional major and minor criteria have been included to each system, with an emphasis for molecular analysis.[7] However, validation of this proposal has not been documented widely in the literature.

Typical manifestations of MFS are seen associated with the skeletal, cardiovascular and ocular systems. The skeletal manifestations include tall stature with the lower segment of the body greater than the upper segment and long, slender limbs & fingers (arachnodactyly); deformities of the chest such as pectus carinatum or pectus excavatum.[8] Cardiovascular abnormalities comprise aortic root dilatation, aortic regurgitation, mitral valve prolapse and aortic aneurysm.[9] Ectopia lentis (subluxation of lens) is a hallmark feature of Marfan syndrome. Other nonspecific ocular features of Marfan syndrome include myopia, elongated eye, flat cornea, and retinal detachment. [10]

Oral manifestations include maxillary protrusion, high arched palate, narrowed arch, crowding of teeth and fragility of the temporomandibular joint.[11-12] Periodontitis associated with Marfan syndrome has always been a topic of debate. Many reports suggest them as a prime finding whereas some reports them as a finding by chance.[13-18] Reports favour periodontal association in MFS suggests that the inherited abnormalities in extra cellular matrix may confer increased susceptibility to periodontal breakdown. Although, the cause of attachment loss is multifactorial, beside genetic predisposition patient-specific risk factors like smoking, oral hygiene,nutrition, obesity, etc., should also be considered.[19-22] On the contrary,others suggest that thehigher degree of inflammation in patients with MFS may have been the result of crowded teeth. As MFS patients suffer from crowding of theteeth compared to sound persons, these crowded teeth arevery much self-cleansable, thus plaque accumulation and consequentlyperiodontal inflammation is much more pronounced.[18]

Regardless of lower/higher susceptibility to periodontitis and slight/more tendency for periodontal inflammation in patientswith MFS, one has to consider that there is an increased risk for developing endocarditis in Marfan patients.[23] Thehigher risk for endocarditis is not the result of the diseasebut the consequence of a prosthetic valve or other prostheticmaterials that are implanted in patients with MFS in orderto treat cardiovascular complications. Therefore, an adequatedental monitoring and regular professional cleaningare recommended because of increasing number of patientswith prosthetic valves suffering from Marfan syndrome.[18]

Taking this into consideration in our cases, antibiotic prophylaxis with 2 grams of amoxicillin one hour prior to the procedure was given. Non surgical periodontal therapy was performed to the patient, after which there was reduction in probing pocket depth and in case two gingival inflammation and enlargement subsided following treatment.

Conclusion

Marfans syndrome affects the systemic health of patient hence it should be diagnosed early and treated.Damage due to inheritance of mutant gene or just crowding of teeth may be thereason for a slight tendency towards more periodontalinflammation in Marfan patients. Taken together, it isrecommended that Marfan patients should receive aprofessional oral hygiene on a regular basis due to theirincreased risk of endocarditis.

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THE AUTHOR HERE BY DECLARE THAT, THERE NO CONFLIICT OF INTEREST IN THE CONDUCTED STUDY