Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 8, Issue, 10, pp.39972-39974, October, 2016

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE STUDY

PERIODONTAL DISEASE ASSOCIATED WITH MARFANS SYNDROME

[1]Dr. Anupama Masapu, [2]Dr. Anusha Gummadi, [3]Dr. Greeshma M., [3]Dr. Salavadhi Shyam Sunder, [3]Dr. Manikantakumar T. [3]Dr. Ramanarayana Boyapati and [4,*]Dr. Sankalp Verma

[1]Assistant Professor, GSL Dental College Rajahmundry Andhra Pradesh
[2]Assistant Professor, Lenora Institute of Dental Sciences Rajahmundry Andhra Pradesh
[3]Assistant Professor, Department of Periodontics, Mamata Dental College, Khammam
[4]Consultant Oral Physician, Sri Sai Hospital, Moradabad

ARTICLE INFO

Article History:
Received 08th July, 2016
Received in revised form
25th August, 2016
Accepted 14th September, 2016
Published online 30th October, 2016

Key words:

Bone loss, Periodontitis,
Connective tissue disorders.

ABSTRACT

Marfans syndrome is the most common dominant autosomic genetic disorder of the connective tissue. This pathology's diagnosis is mainly based on physical characteristics. Herein, the authors' present report of a case of Marfans syndrome in which patient presented with bone loss involving seven teeth which was diagnosed as periodontitis in Marfans syndrome. This case reports confirms the hypothesis that connective tissue disorders increased susceptibility to periodontal breakdown.

Citation: Dr. Anupama Masapu, Dr. Anusha Gummadi, Dr. Greeshma, M. *et al.* 2016. "Periodontal disease associated with marfans syndrome", *International Journal of Current Research*, 8, (10), 39972-39974.

INTRODUCTION

Marfan Syndrome (MS) was firstly described by the pediatrician *Antoine Bernard-Jean Marfan* who reported an out of proportioned length of the lower limbs and fingers (Shirley *et al*., 2009). Marfan syndrome is the most common inherited connective tissue disorder, with a reported incidence of 1 in 10,000 individuals and equal distribution between the sexes (Dietz *et al*., 1991). It is caused by an autosomal dominant mutation in the gene encoding fibrillin (FBN1, chromosome 15q15) a glycoprotein that is an integral part of the connective tissue in the body (ligaments, blood vessel, eye lenses) (Pyeritz, 1997). It has been described that the normal fibrilininhibits the growth of the long bones and elastic fibers, through its tension control. As fibrillinbeing altered, an exaggerated bone overgrowth is produced. (Voermans *et al*., 2009) MS is a multi-systemic disorder with typical manifestations which affect the skeletal, cardiovascular and ocular systems. Skeletal manifestations are the cardinal signs of Marfan syndrome and usually gain the attention of a physician. The most common features include tall stature with the lower segment of the body greater than the upper segment and long, slender limbs, or dolichostenomelia; thin body habitus with increased arm span-to-height ratio; long, slender fingers, or arachnodactyly, deformities of the chest, such as pectuscarinatum or pectusexcavatum; scoliosis; and highly arched palate with crowded teeth and dental malocclusion. Other less common manifestations include hypermobility of joints, flat foot (pesplanus), reduced extension of elbows (< 170 degrees), and elongated face (dolichocephalia). (Ammash *et al*., 2008; Umamahesh *et al*., 2006) Cardiovascular manifestations are the most serious complications and determine the prognosis and survival in Marfans syndrome. Abnormalities include aortic root dilatation, aortic regurgitation, aortic dissection, and aortic aneurysm, which most commonly involves the ascending aorta but can involve the descending aorta. The rate of aortic root dilatation is unpredictable and usually requires surgery when it measures more than 50 mm. Mitral valve prolapse can also occur. Although cardiovascular abnormalities typically appearlate, they can occur during childhood. (Grimes *et al*., 2004) Ectopialentis (subluxation of lens) is a hallmark feature of Marfans syndrome and is present in approximately 60% to 80% of patients. Ectopialentis is usually bilateral, symmetrical, and upward. The diagnosis can be made by looking for iridodonesis (tremor of iris), phacodonesis (abnormal movementof lens), and a deep anterior chamber in the non-

Corresponding author: Dr. SankalpVerma,
Consultant Oral Physician, Sri Sai Hospital, Moradabad

dilatedeye. The dislocation may be complete, with the lensfloating free within the vitreous cavity. Other nonspecificocular features of Marfans syndrome includemyopia, elongated eye, flat cornea, and retinal detachment. (Fuchs, 1997)

Case report

A 26 year old woman reported to the outpatient department of Periodontics, St.Joseph Dental College, Eluru with a chief complaint of swollen gums in relation to lower front teeth region. Medical history revealed mitral valve prolapse.

On general physical examination an ectomorphic (tall and lean) patient with disproportionate height (lower segment of the body greater than the upper segment) (Fig.1a) presented withdolichostenomelia (Fig.1a) arachnodactyly (Fig.1b, 1c) and ectopialentis (Fig.1d). Hyperextension ability of the joints was noticed. Patient presented with positive wrist sign (Fig.1d) and Steinberg sign (Fig.1e). Intraoral examination revealed a high arched and narrow palate (Fig.2a). Generalized gingival inflammation with profuse bleeding on probing was noted.3mm pockets were present i.r.t maxillary right and left upper teeth and mandibular molars bilaterally; FDI tooth number: 12,13,16, 22,23,26,36,46 (Fig.2b,2c). 4mm pockets were seen in mandibular anterior teeth (Fig.2d).

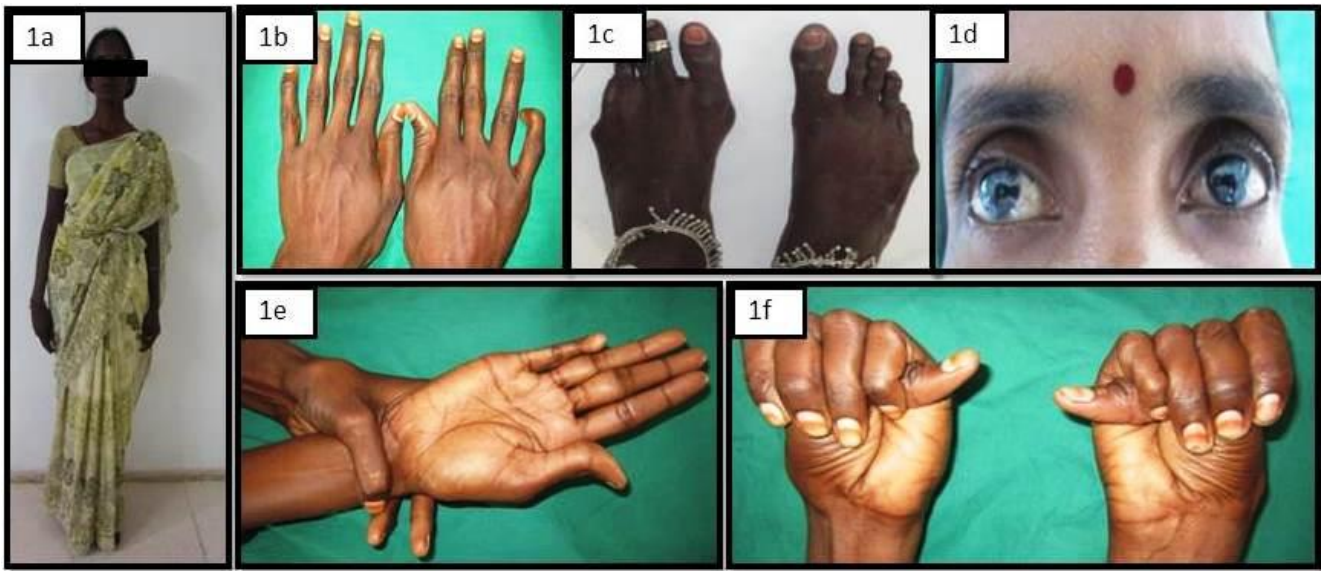

Fig.1.

1a: tall n slender built with lower body segment longer than the upper. Note the elongated face
1b, 1c: spiderlike fingers of hands and toes
1d: ectopialentis
1e: positive wrist or Walker's sign (The distal phalange of the first and fifth fingers of the hand overlap when wrapped around the opposite wrist)
1f: positive thumb or Steinberg sign (The thumb projects beyond the ulnar border while completely opposed within the clenched Hand)

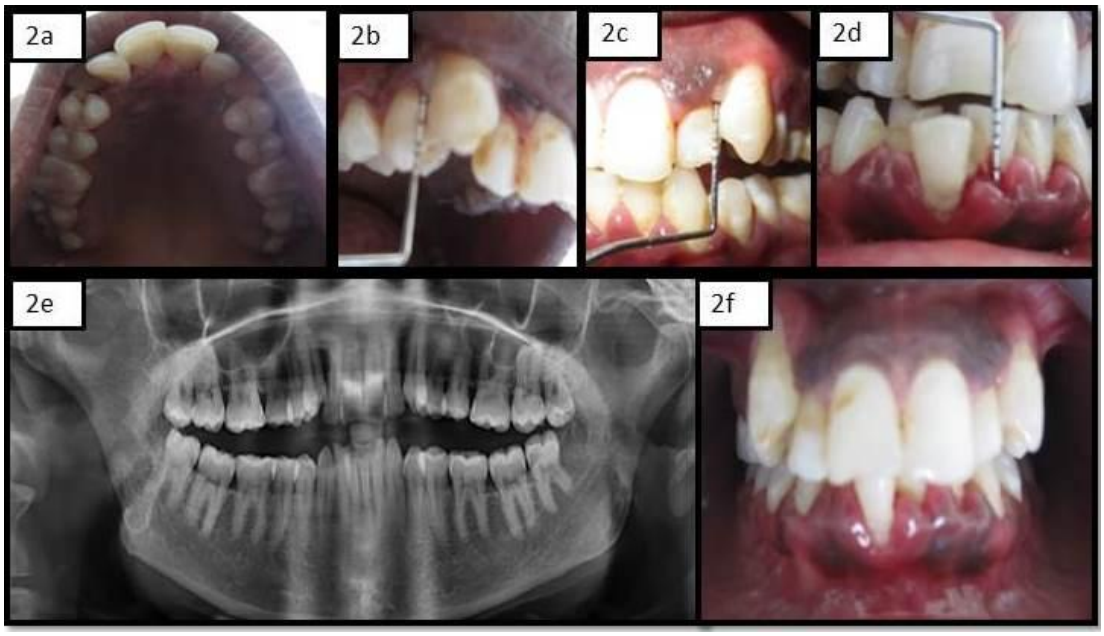

Fig.2
2a: narrow and high arched palate
2b, 2c: gingivitis with 3mm pockets
2d: 4mm deep pockets in mandibular anterior teeth
2e: Panoramic radiograph demonstrated generalized bone loss
2f: Follow up of the patient after non-surgical periodontal treatment

Based on the presence of clinical signs and symptoms and Berlins criteria, the patient was diagnosed with Marfans syndrome and subjected to panoramic radiography to investigate the severity of bone loss. On examination of OPG bone loss was seen in relation to tooth number: 12,22,16,26,36 and46 (Fig.2e). Thus, final diagnosis of periodontitis with Marfans syndrome was made and treatment was planned. Taking into consideration the cardiac pathology of the patient, it is imperative that preceding the performance of periodontal treatment, antibiotic prophylaxis by means of the intake of 2 grams of Amoxicillin one hour prior to the procedure is given.Non-surgical periodontal therapy was performed to the patient (Fig.2f).

DISCUSSION

Marfan syndrome is caused by mutations in the *FBN1* gene on chromosome15, which encodes the glycoprotein fibrilli n-1, a component of the extracellular matrix.Fibrillin-1 protein is essential for the proper formation of the extracellular matrix, including the biogenesis and maintenance of elastic fibers. Elastin fibers are found throughout the body, but are particularly abundant in the aorta, ligaments and the ciliaryzonules of the eye, these areas are among the worst affected (Sakai *et al*., 1986). In this case report the patient has periodontal break down and gingival enlargement .Inherited abnormalities in extra cellular matrix may confer increased susceptibility to periodontal breakdown (Tornos, 2002). This case reports confirms the hypothesis that connective tissue disorders increased susceptibility to periodontal breakdown.

Conclusion

This case report is an excellent example of careful history taking and thorough examination of the patient. Although the patient presented with swollen gums but the incidental findings as elicited by the vigilant investigators led to the diagnosis of a rare syndrome.

REFERENCES

Ammash NM, Sundt TM, Connolly HM. 2008. Marfan syndrome-diagnosis and management. *CurrProblCardiol.,* 33:7-39.

Dietz HC, Cutting GR, Pyeritz RE, *et al*. 1991. Marfan syndrome caused by a recurrent de novo missense mutation in the fibrillin gene. *Nature*, 352:337–9

Fuchs J. 1997. Marfan syndrome and other systemic disorderswith congenital ectopialentis. A Danish national survey. *ActaPaediatr*., 86:947–52.

Grimes SJ, Acheson LS, Matthews AL, Wiesner GL. 2004. Clinical consult: Marfan syndrome. *Prim Care*, 31:739–42.

Pyeritz RE. 1997. Disorders of fibrillins and microfibrilogenesis: marfan syndrome, MASS phenotype, contractural arachnodactyly and related conditions. In: Rimoin DL, Connor JM, Pyeritz RE, editors. Emery and Rimoin's principles and practice of medical genetics. 3rd ed. New York: Churchill Livingstone, 1027–66.

Sakai LY, Keenedr, Engavall E. 1986. A new 350KD glycoprotein is a component of extracellular microfibrils, J Cell Biology, 103; 2499-2509.

Shirley ED, Sponseller PD. 2009. Marfan syndrome. *J Am AcadOrthop Surg*., 17:572-81.

Tornos P. 2002. [Infective endocarditis: Are we managing our patients well?]. *Rev EspCardiol*., 55:789-90.

Umamahesh C. Rangasetty, MD Bernard M. KarnathClinical Signs of MarfanSyndrom MDHospital Physician, April 2006.

Voermans N, Timmermans J, Van Alfen N, Pillen S, Op Den Akker J, Lammens M, *et al*. 2009. Neuromuscular features in Marfansyndrome. *Clin Genet*., 76:25-37.

****