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CASE REPORT

TERRIEN'S MARGINAL CORNEAL DEGENERATION: A FOLLOW UP OF 29 YEARS: A CASE REPORT

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ABSTRACT: Terrien's marginal corneal degeneration is a rare disease of the peripheral cornea characterized by vascularisation, opacification, lipid deposition, and thinning. With progression, large degrees of astigmatism occur and perforation is a threat. Severe astigmatism may be managed with spectacles or rigid contact lenses. No single therapy has been shown to halt the progression of the disease. Usually pseudo pterygia occur in Terriens degeneration in 20 % of cases. Failure to recognise this pseudopterygium may cause unnecessary removal thus corneal perforation. So though a rare clinical entity we should keep Terrien's degeneration in mind with its possible variations, and treat the patient accordingly and once diagnosed ophthalmologists should be aware of the regular follow ups to see the progression of the disease to recognize impending perforations. Here in we report a case of unilateral Terrian's marginal corneal degeneration with a long follow up of 29 years at our institute showing the pattern of progression of disease.

KEYWORDS: Terrien's marginal corneal degeneration, astigmatism, corneal perforation, long term follow up, pseudopterygium.

INTRODUCTION: Terrien's marginal corneal degeneration is a rare disease (Terrien himself saw only three cases in thirty years of practice) of the peripheral cornea characterized by vascularisation, opacification, lipid deposition, and thinning, 75 % being males[1] Terrien's Marginal Degeneration is typically described as a disease of the middle to late age.[1] However there are several articles that have described a variant of this disease with prominent inflammation as episcleritis or scleritis occurring in a younger age group and in children.[1,2]

This condition may be bilateral or unilateral. Lesions begin usually superonasally rarely inferiorly with development of fine, white sub epithelial, peripheral opacities that spare the limbus. The opacities coalase and this is followed by corneal thinning, typically with a sloping central edge and a fairly steep peripheral edge to the resultant furrow.[3] The epithelium is typically intact with yellowish white lipid deposits in the centre of the gutter with associated vascularisation. Etiology is unknown, although inflammatory, degenerative, and immune mediation have been proposed.

With progression, large degrees of astigmatism occur and perforation is a threat.[4] As it progresses slowly and painlessly it is common the diagnosis to be made in an advanced stage. Perforation is believed to happen spontaneously after minor trauma in up to 15% of the patients.

Here in we report a case of unilateral Terrian's marginal corneal degeneration with a long follow up of 29 years at our institute well documented photographically.

CASE REPORT: A 55 year old male patient has been at a regular follow up at Indira Gandhi Medical College Shimla from last 29 years diagnosed as unilateral Terrien's Marginal Corneal Degeneration of right eye. He first presented to our hospital with symptoms of progressive diminution of vision in

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May 1986 at the age of 26 years. His visual acuity at the time of presentation was found to be 6/9 P in right eye and 6/6 in left eye with cylinder of -1.5 D at 100^0 in right eye and no error in left eye. His keratometric values (Bausch and lomb) K1 (horizontal) and K2 (vertical) at that time were 44D and 43D respectively. The patient underwent a thorough slit lamp examination which revealed a superior circumlinear corneal thinning with intact epithelium, without neovascularisation, mild lipid deposition and a clear area existing between the lesion and limbus. The degree of thinning varied from 20 to 30%. The extent of thinning was seen between the 10 and 2o'clock positions. There was no epithelial defect noted by fluoroscein staining. The left eye was normal.

He had no history of episodes of pain in the past suggesting non inflammatory type of disease. Complete blood profile was done which was normal. The ocular tension was 17.3 mm by Shiotz tonometry in both eyes. Gonioscopy revealed open angle in the affected eye. The patient's fundus was normal. He was diagnosed as a case of unilateral Terrien's Marginal Corneal Degeneration of right eye and was advised glasses for his refractive error. He was explained about the progressive nature of disease and advised regular follow ups and also to avoid any inadverdant trauma to his right eye.

In the subsequent follow ups his right eye vision progressively deteriorated along with progression of corneal thinning and against the rule astigmatism. In 1996 about 10 years from the diagnosis of disease his visual acuity in right eye was 6/36 with refraction of -8.00D x 100 degree showing worsening of astigmatism. His keratometric values K1 and K2 were 46D and 42D respectively in affected eye. The vision in left eye showed no deterioration.

His right eye vision has now deteriorated to the present visual acuity of 6/60P with refraction of -13.00 D x 105 degree due to progression of corneal astigmatism resulting in severe visual disability in his right eye. Presently his keratometric values were K1 47D and K2 39D suggesting high against the rule astigmatism.

The present slit lamp examination shows progression of superior circumlinear corneal thinning with intact epithelium, with slight neovascularisation, significant lipid deposition and a clear area existing between the lesion and limbus. (Fig. 1, 2, 3). The degree of thinning has increased to 50 to 70 %. The extent of thinning is increased to between 9 and 3 o'clock positions. The left eye showed no signs of disease.

The idea of reporting this case is to emphasize that Terrien's Marginal Degeneration is a progressive disease and regular follow ups are necessary to rule out impending perforations which fortunately did not happen in our patient. Severe astigmatism may be managed with glasses or rigid contact glasses as our patient was managed with glasses. An option for full thickness or lamellar keratoplasty has been given to the patient.

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FIG. 1: Slit lamp photograph showing thinning of cornea superiorly.

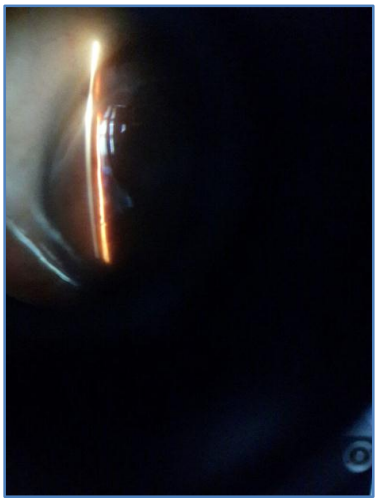

Fig. 1

FIG. 2: Showing thinning and peripheral stromal opacification of superior cornea.

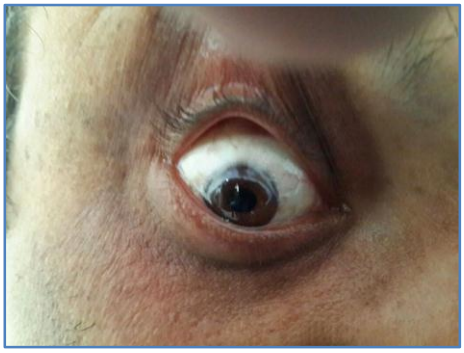

Fig. 2

FIG. 3: Showing showing thinning and lipid deposition in cornea superiorly from 9 to 3'oclock position in right eye.

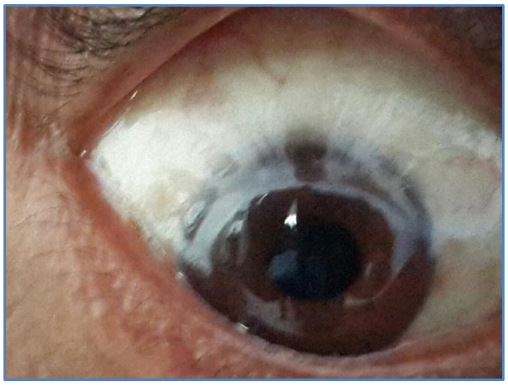

Fig. 3

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DISCUSSION: In Terriens degeneration when the thinning is restricted to the superior or inferior area of the peripheral cornea, there is relative steepening approximately 90^0 away from the mid-point of the thinned area resulting in astigmatism characteristic of this disorder. Histologically epithelium may be normal, thickened, and the thinned Bowmans layer and the lamellae may be split or fibrillated.[1] Terrien Marginal Degeneration usually starts at the superior nasal quadrant as fine yellow-white punctuate stromal opacities which progressively involves the entire circumference of the peripheral cornea with neovascularisation, scarring and lipid infiltration occurring at the leading edge.[5]

Though a lot of etiologies have been proposed, levels of circulating immune complexes are not elevated in patients with Terrien's Marginal corneal degeneration. Marginal furrow degeneration, Dellen, collagen vascular diseases, sclerokeratitis, staphylococcal marginal keratitis, pellucid marginal degeneration etc have been proposed as differential diagnosis.[6]

Usually pseudo pterygia occur in Terriens degeneration in 20 % of cases[7]. Failure to recognise this pseudopterygium may cause unnecessary removal thus corneal perforation[8]. Very rarely extreme thinning occurs when reconstructive surgery is indicated. A full thickness or lamellar corneo sclera graft (Often hand fashioned to fit the defect) may be necessary.[9] The progressive increase in against the rule astigmatism or oblique astigmatism with advanced disease can be arrested upto 20 yrs by grafting. Severe astigmatism can also be treated by a crescentric shaped excision of the gutter with suturing of the healthier margins.[10] Corneal thinning may progress sometimes, despite intact epithelium, to the point at which a deep corneal break leads to hydrops or even frank perforation in about 15 % of cases.[6] In these cases keratoplasty either a full thickness or deep lamellar is necessary.[6]

So though an uncommon clinical entity we should keep Terriens degeneration in mind with its possible variations, and treat the patient accordingly with regular follow ups to see the progression of the disease.

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