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CASE STUDY OF STARGARDT DISEASE

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ABSTRACT

Stargardt disease is the most common form of juvenile macular degeneration. Clinically, it is characterized by pisciform flecks at the level of the retinal pigment epithelium and a bull's-eye maculopathy. Inheritance is usually autosomal recessive, although dominantly inherited case have been described. Both sexes are affected equally. Even though significant research is taking place in this area, no treatment is available till date for this disease in contemporary science. In Ayurveda by considering the visual disturbance & field defect, this disease can be included under drishtigata rogas explained by Acharya Sushruta, Pitta vidagdha drishti is one among them, which holds more appropriate to define this clinical condition. As there is no common line of management following in these types of diseases in Ayurveda, an attempt has been made to explain the case treated as per the Ayurvedic principles. This study describes diagnosed as Stargardt's macular degeneration & underwent Ayurvedic treatment for one and half year. By executing Ayurvedic treatment the quality of the vision distant & near acuity is moderate improved.

KEYWORDS: Stargardt's disease, drishtigata roga, Tarpana.

INTRODUCTION

Stargardt disease is the most common type of hereditary recessive macular dystrophy. The estimated incidence for the disease is 1 in 10,000 live births, and it characteristically presents in juveniles and young adults.[1] Patients begin to experience a bilateral, gradual decline in their vision between the ages of 6 and 20 years, and can present with visual acuity ranging from 6/18 to 6/60. It was named for the German ophthalmologist Karl Stargardt, and may

also be called fundus flavimaculatus. In people with Stargardt disease the RPE collects lipofuscin, which can lead to vision problems. Vision loss in Stargardt disease is most intense in the macula. Stargardt disease is part of a group of diseases affecting the macular region of the retina, called macular degenerations. Stargardt disease is sometimes called a juvenile macular degeneration because it often appears at an early age.[2] Autosomal recessive Stargardt disease is caused by mutations in a gene called ABCA4. A second gene called ELOVDT has been found to be the cause of the autosomal dominant form of Stargardt-like disease.[3] The classical Stargardt phenotype is characterized by a juvenile -onset foveal atrophy surrounded by discrete, yellowish round or pisciform flecks at the level of the Retinal pigment epithelium.

AIMS AND OBJECTIVES

- To study the efficacy of Ayurvedic management in Stargardt's macular degeneration.
- To understand Stargardt's macular degeneration in Ayurvedic perspective.

MATERIALS AND METHOD

A 10 year old female child patient diagnosed as Stargardt's macular degeneration was selected and consent was taken.

Review of Literature

A. Pathological considerations - Modern perspective

- Retinal changes in Stargardt's disease there are two main findings on the retina are commonly seen. In the initial stages, usually an oval-shaped lesion, often referred to as 'beaten bronze' in appearance, around the macula is seen. This lesion tends to deteriorate over time leads to a loss of visual acuity, reduced colour perception and more discomfort in the day light or bright light vision. The second change involves yellowish flecks which surround this lesion. In some cases people have just these flecks without the macular lesion and this type of disease used to be diagnosed with fundus flavimaculatus.[4] However, some researchers believe that these two problems, the macular lesion and the yellow flecks, are both caused by the gene which causes Stargardt's disease and therefore are different versions of the same genetic problem. It has also been suggested that fundus flavimaculatus and Stargardt's disease vary in age of onset and severity where fundus flavimaculatus may appear in the 20's and 30's and vision may be more severely affected.

- **Consequences on vision in Stargardt's disease**

Stargardt's disease mainly affects the macula and hence central vision is affected more. So this disease will make the central vision unclear and then sometimes distorted or blurred. As the disease progresses, a blank patch may appear in the centre of vision (central scotoma). Stargardt's disease does not affect other parts of the retina so does not normally affect peripheral or side vision. But this can also cause problems such as glare and difficulties adapting to changing light conditions and colour perception. Stargardt's disease does not appear to have any effect on general health.

- **Diagnostic techniques**

Usually the diagnosis is apparent from the history, age of manifestation and the way of presenting the visual related problems. Initially the eyes may look normal, but as the years pass, the flecks and atrophic scars develop. A useful test is the fundus fluorescein angiogram (FFA) in which approximately 86% of the patients shows a sign called the "dark choroid effect" is present, which arises from the abnormal accumulation of the substance lipofuscin in the retinal pigment epithelium.[5] Monitoring the visual acuity (distant & near), colour vision, visual field analysis, ERG, Fundus Fluorescein Angiography and Amsler grid charting are the useful diagnostic techniques.

- **Prognosis and management**

There is currently no treatment for Stargardt's disease in modern medicine. Researchers have reported that exposure to ultraviolet light may cause further retinal damage. This is because an abnormal protein in retinal pigment epithelium (RPE) layer becomes more toxic when patient exposed to extreme sunlight. It is therefore recommended that wearing sunglasses with UV protection that conforms to Australian Standards and a hat with a wide brim can protect individuals from the sun's damaging ultraviolet rays.[5] In general, the same dietary recommendations that are made for the age-related form of macular degeneration are usually apply to people with Stargardt's disease.

B. Pathological considerations - Ayurvedic perspective

The features like central visual loss diminished distant and near vision are pointing towards the pathophysiology of Tritheeya Patala gata Timira.[6] Similar Dosha vitiation is also mentioned in the pathophysiology of Pitthavidagdha drishti.[7] In this disease patient feels more difficulty during the day vision also. As the disease starts manifesting in the Tritheeya

patala, special attention should be given to Raktha dhatu. Due to the involvement of deeper Dhatus, the prognosis is also become Yapya or Asadhya. Chakshu vaisheshika Alochaka Pittha and Prana vayu should be given distinct consideration while planning the treatment protocol. Rasayan, Nasya, and Tarpana are the potent treatment procedures found to be effective in this condition.

CASE STUDY

To report a patient with Stargardt's disease treated with classical Ayurvedic management, a 10 year old female child with Stargardt's disease treated in our hospital participated in the study. Examinations performed before and after treatment including visual acuity, fundus photography, Amslergrid. FFA was done to confirm the diagnosis.

10 year old female patient approached in OPD on 20-12-2021 with diminished distant and near vision in both eyes. She was also complaint of colour scotoma during reading. Her visual acuity at the time was 6/60 which improved to 6/12 and near vision N/10 which improved N/6 in both eyes.

	TREATMENT	**MEDICINE**	**DOSE**	**DURATION**
Poorva karma	Kostha shodhana	Avipatikara churna	5 gm BD before food with water	7 Days
Pradhan karma	Shamana chikitsa	1)Rasayana churna-3gm Jivanti churna -2gm Saptamruta loha-500mg	5.5gm BD after food with honey	One and half year
		2)triphala kwath-5gm	5gm BD twice per day	One and half year
Kriya-kalpa	Pratimarsha Nasya	Anutail	2 drop Morning time	One and half year
	Tarpana	Jivantyadi ghrita	15-20 minutes Tarpana was done for 7 days after gap of one week in every 15 days	One and half year

OBSERVATION AND RESULT

During the last one and half year, the visual acuity was not deteriorated and patient felt more clarity of vision with Moderate improvement in the distant and near vision.

Investigation		**Unaided Distant**		**Aided Distant**		**Unaided near**	
		OD	**OS**	**OD**	**OS**	**OD**	**OS**
Visual acuity	B.T	6/60	6/60	6/60	6/60	N/10	N/10
	A.T	6/12P	6/12P	6/12	6/12	N/6	N/6

DISCUSSION

DISCUSSION By considering the pathophysiology and clinical features of Stargardt's macular degeneration, it can be considered as a Drishtiroga in which utmost vitiation of Alochaka pittha. The Samprapthi and clinical features of Pitthavidagdha drishti is most suitable to understand this dangerous degenerative disease. Involvement of Sapthadhatus along with Vata-Pittha vitiation is the undergoing pathology in this case and Thrithiya patala is the site of lesion which is suddenly progressing to Chaturtha patala causing marked reduction of vision. Age of manifestation of the disease also points towards the role of Pittha in the disease process. While dealing such a chronic degenerative disease, ultimate line of management should be Vatapitthasamana and Rasayana. For the purpose of Tarpana, Jivantyadi ghrita is used due to its Vatapradhana drishtirogahara action. Anutail has a special role here because Prnavayu plays a key role for the stimulation of Chakshurindriya. Rasayana churna have three components Guduchi,Gokshura and Aamalaki. All these drugs have Anti-oxidant activity immunomodulatory and Anti-aging property. Jivanti churna and saptamruta loha both have chakshushya, anti Oxidant and Neuroprotective property. Triphala have vata-pitta shamak, chakshushya and anti oxidant properties. These disease is chronic progressive in nature all drugs have neuroprotective property which can bring moderate improvement. During follow-up period, Pratimarsha nasya, Ghrita Tarpana and Rasayana were advised to get supplementary effect in due to its Chakshushya and Rasayana action.

CONCLUSION

Even though Stargardt's macular degeneration is a chronic progressive disease; it can be managed by the principles of Ayurveda. By adopting the Ayurvedic treatment, the vision of the patient sustained and quality of the vision including acuity is improved at least for a period of one and half year. So to manage the diseases like macular degenerations, an Ayurvedic treatment protocol should be derived so that more data can be generated. The method adopted in this case can be taken as a guideline to manage the macular degenerative diseases which need extensive management to tackle the pathology.

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