

The Burden of Rare Disease in India the Eales Disease

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Abstract: Eales disease is a rare but a silent killer of the eye, it is an idiopathic obliterative vasculopathy which involves the peripheral retina in young adults. It is also known as angiopathia retinae juvenilis, periphlebitis retinae, primary perivasculitis of the retina, and it is characterized by inflammation and blockage of retinal blood vessels, abnormal growth of new blood vessels (neovascularization), and recurrent retinal and vitreal hemorrhages.

Keywords: eales disease, periphlebitis

1. Introduction

Eales disease is a rare but a silent killer of the eye, it is an idiopathic obliterative vasculopathy which involves the peripheral retina in young adults. It is also known as **angiopathia retinae juvenilis, periphlebitis retinae, primary perivasculitis of the retina**, and it is characterized by inflammation and blockage of retinal blood vessels, abnormal growth of new blood vessels (neovascularization), and recurrent retinal and vitreal hemorrhages .

Discovery

Eales' disease was first described by Henry Eales, a British ophthalmologist, in 1880 and 1882 who thought that it is a non-inflammatory condition.

Definition

Eales Disease: Is defined as rare disorder of eye, characterized by sudden vision loss, problems with eye blood vessels and reoccurring eye blood vessel hemorrhages which are characterized by oozing of clear thick fluid inside the eyeball.

Epidemiology

Eales' disease was described originally in England, but is observed **much more commonly on the Indian subcontinent** than in any other region. The incidence of Eales' disease has shown a significant raise as it occurs in 1 of every 135 to 260 patients who are evaluated at tertiary eye care centers in India.

Etiology

The cause of this condition is not known. However, in a significant number of patients, DNA of the bacterium Mycobacterium tuberculosis was detected by PCR. Studies show the evidences that hypersensitivity to tuberculo proteins plays avital role in the cause of Eales' disease.

Causes Of Eales Disease

The cause of Eales Disease is not yet known. Researchers believe that this condition spontaneously occurs without precipitating factors such as an injury, infection, or any genetic factors causing this abnormality in the retina of the eye.

Symptoms

- Sudden vision loss

- Sudden vision impairment
- Scotoma
- Floating spots
- Impaired ability to control voluntary movements
- Pins and needles sensation
- Speech disorder
- Inflammation of retinal blood vessels
- Retinal hemorrhage
- Detached retina

New Staging System for Eales' Disease

Stage	Description
I	Periphlebitis of small (Ia) and large (Ib) caliber vessels with superficial retinal hemorrhages
IIa	Capillary nonperfusion
IIb	Revascularization elsewhere/of the disc
IIIa	Fibro vascular proliferation
IIIb	Vitreous hemorrhage
IVa	Traction/combined rhegmatogenous retinal detachment
IVb	Rubeosis iridis, neovascular glaucoma, complicated cataract, and optic atrophy

Medical Therapy

- Corticosteroids are preferred to be the mainstream treatment in the therapy in active perivasculitis stage of Eales' disease.
- (Mantoux positive), Antitubercular treatment is given for a period of 9 months.
- If the individual is in the stage of Retinal periphlebitis and strongly positive for PPD test then systemic steroids and appropriate antitubercular drugs to be administered to avoid reactivation of the systemic illness.

Photocoagulation

The main treatment in proliferative stage of Eales disease is Photocoagulation, because the client is more prone to get vitreous hemorrhage in stage of Eales disease. But it's contradictory if laser or panretinal photocoagulation would be beneficial in inflammatory stage of Eales disease. After photocoagulation, Fluorescein angiography should be done to monitor the response to the treatment.

Standard Therapies

Treatment of Eales Disease is symptomatic and supportive. In case of Eales disease the blood vessels will be hemorrhagic and constriction of the blood vessels will be the main goal in the treatment, so laser beam (laser panretinal

photocoagulation) may be used to slow down excessive formation of blood vessel tissue. If Hemorrhaging of the clear jelly behind the lens of the eye occurs then removal of the dark pigmented disk and jelly-like substance to be done behind the retina.

Organization for Rare Diseases India

There are few organizations working to voice for the patients with rare diseases in Karnataka .ORDI is one of them ,ORDI is a national organization located in Karnataka (INDIA),which helps to note the incidence of the diseases and helps to bring awareness among the public .It is located in Bangalore in the state of Karnataka .

Application of Orem's Theory in Patients with Eales Disease

Dorothy Orem's self-care theory mainly concentrates on the individual's capacity to care for one self and the role of a nurse to help the individual to fill the gaps of care, when a person is unable to do so because of his or her illness.

This theory is interrelated with 3 main sub systems

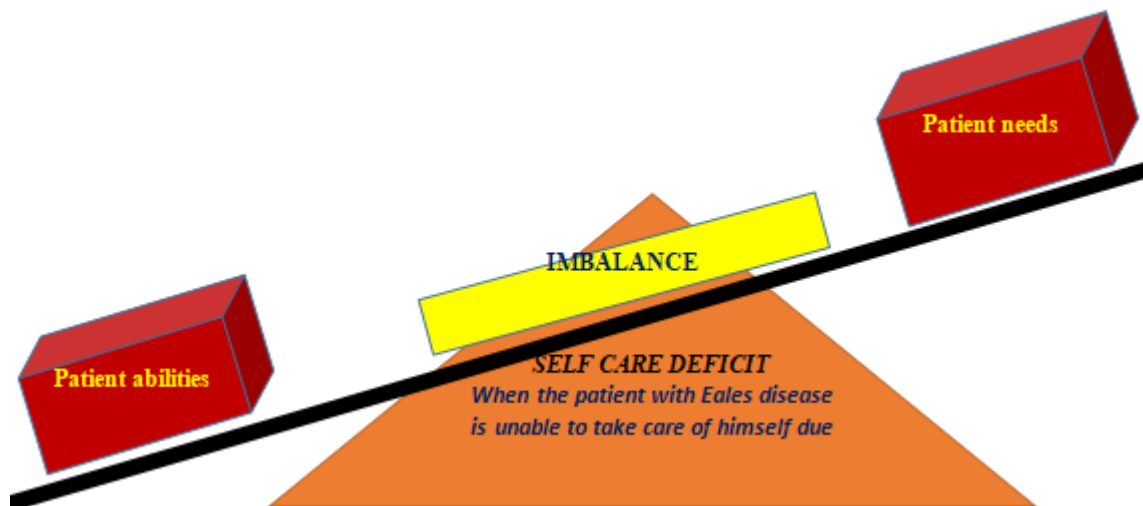
- Theory of self-care
- Theory of self-care deficit
- Theory of nursing system

Theory of Self Care

According to the theorist self-care means the activities which an individual should perform on his or her own for the wellbeing, when this concept is applied to a patient with Eales disease, due to the defect in the eye, it is hardly possible for the person to carry out the activities of daily living ,thus patient lands up into second concept of the theory.

Theory of Self Care Deficit

This concept explains when the individual is unable to take care of himself due to some deficits in the individual systems. These can be identified as universal, developmental and health deviation self-care requisites.



2. Theory of Nursing System

This subsystem illustrates when is nursing care needed by an individual and how can nurses help an individual to regain back his normal activities .So the theory explains about the 5 methods of helping ,

- **Nurse guides others** - here nurse can guide these patient about the care to be taken and the medical facilities which are available to help patient with Eales disease.
- **Supporting another** - this means the nurse supports the patient with Eales disease both with treatment and self-care.
- Providing an environment promoting personal development in relation to meet future demands.
- Teaching about the care to be taken.

Orem identifies three classifications of nursing system to meet the self-care requisites of the patient.

- **Wholly compensatory system,**
- **Partly compensatory system,**
- **Supportive-educative system.**

So being a nurse when we care for patients with Eales disease,

If patient is in **wholly compensatory system**, then these interventions can be implemented,

- Collect the history of the patient thoroughly
- Perform a quick physical examination and carry out the investigations
- Identify the signs and symptoms of the disease and classify in which stage is the disease progressing
- Provide proper medical and surgical management as needed by the patient

Partly compensatory system

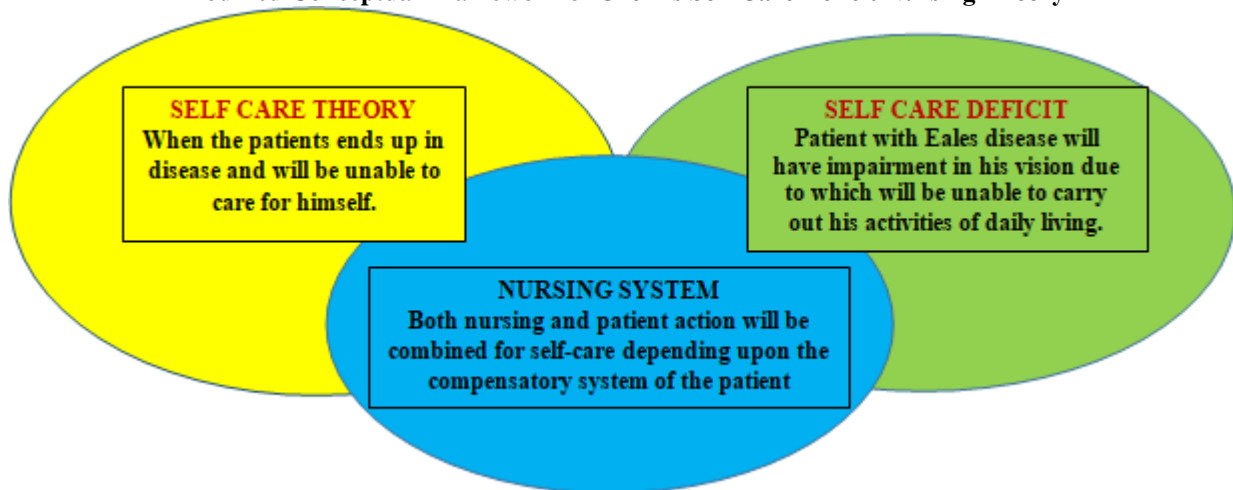
- if the patient is in this stage of disease progress
- take appropriate action both with medical and surgical management

Supportive-educative system

- Here nurses play a vital role in educating both patient and family about the care
- Education regarding the association which is available to help the patient

- Conduct awareness programmes and educate the public about these rare disorders in India

Modified Conceptual Framework of Orem's Self Care Deficit Nursing Theory



In this theory all this 3 concepts are interrelated depending on the condition of the patient .when the patient is unable to take care of himself /herself due to progression of disease, he or she becomes self-care deficit, and then nurses play a vital role in caring for the patients with Eales disease, by providing care both medically and psychologically and help the patient to cope up with the situation, by meeting the needs of the patient and making the patient to be comfortable.

3. Conclusion

Since Eales disease is one of the rare diseases in India and is a killer diseases, it is very important to know the prevention aspect and normal people should be aware of the disease .because “prevention is better than cure”.

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