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## Nodular anterior scleritis in systemic sclerosis: a case report



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### ABSTRACT

**Introduction:** Scleritis is an inflammation of the sclera's lining, characterized by cellular infiltration, collagen damage, and vascular changes. The incidence rate of scleritis accompanied by systemic disease generally is around 39% to 50%. Scleritis can be divided into two, namely anterior and posterior scleritis. Anterior scleritis can be divided into four, diffuse anterior scleritis, nodular anterior scleritis, necrotizing anterior scleritis with inflammation and without inflammation. One of the causes of nodular anterior scleritis is systemic sclerosis which is an autoimmune disease. Nodular anterior scleritis in systemic sclerosis is very rare, with an estimated prevalence rate of 4.4%.

**Case Description:** A man, 46 years old, complaining of red, watery, painful, and visible white membrane on his right eye since two weeks ago and was referred from an ophthalmologist with scleritis on his right eye. The patient had a history of asthma and cataract surgery.

Physical examination showed right eye VA was 6/45 pinhole (PH) 6/18, conjunctival and scleral injection, nodule on superior conjunctiva with yellowish-white in color. Laboratory examination showed a clinical representation of autoimmune disease, consulted to the rheumatology division, and was diagnosed with systemic sclerosis. The patient has been prescribed corticosteroids and immunosuppressants for the therapy, the patient's eye condition improves after.

**Conclusion:** Systemic sclerosis with a presenting ophthalmologic complaint is a rare disease. This case illustrates that nodular anterior scleritis needs comprehensive intervention to seek the etiology and provide the treatment. This case has shown that nodular anterior scleritis with systemic sclerosis improved with corticosteroids and immunosuppressants, a typical treatment for autoimmune disease.

**Keywords:** nodular anterior scleritis, systemic sclerosis, autoimmune diseases, corticosteroids.

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### INTRODUCTION

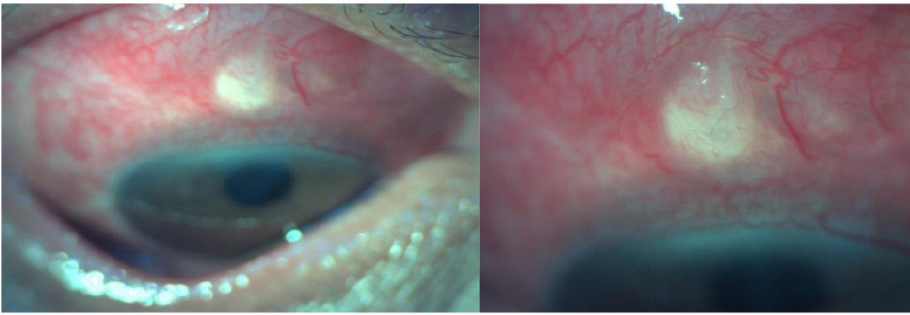
Nodular anterior scleritis was an ophthalmologic disease characterized by the presence of one or more erythematous inflammatory nodules. It is usually immobile, purplish-red scleral nodules, separated from the overlying episcleral tissue, which are unintentionally lifted by nodules and cause a pain sensation in the anterior sclera. About 20% of these cases may develop into necrotic scleritis. Systemic sclerosis is a multi-system autoimmune disease characterized by extensive vascular injury, progressive fibrosis of the skin and internal organs.<sup>1</sup> Eye disorders due to systemic sclerosis

can affect any part of the eye, either the adnexa or the visual system. There are various clinical manifestations and varying prognoses. Most of the patients experiencing thickened skin and some involving internal organs.<sup>2</sup> Inflammatory conditions such as uveitis, episcleritis, scleritis, and ulcerative peripheral keratitis, manifested by connective tissue involvement, have been reported in patients with systemic sclerosis.<sup>3</sup>

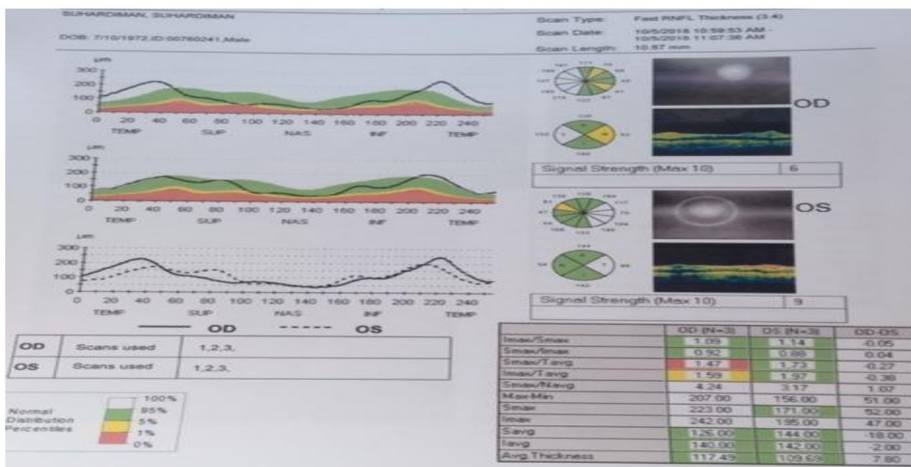
### CASE REPORT

Male, 46 years old, came to the Ophthalmology clinic, the External Eye Disease (EED) division at the Sanglah

General Hospital, Denpasar, on October 5<sup>th</sup>, 2018. The patient complained of red, watery, and visible white membrane on his right eye for the previous two weeks. The patient also complained about pain in the right eye, especially when moving the eyeball. He also complained of slightly blurred vision which has not been realized for a long time. In addition, he has also experienced headaches that occurred at the same time as the ophthalmologic complaint. The patient has a history of asthma and was treated regularly. He also had a history of cataract surgery on the right eye in 2016.



**Figure 1.** Photograph of the patient's right eye taken on October 5<sup>th</sup>, 2018. These showed a dilated conjunctival and sclera vessels around the yellowish-white nodule.



**Figure 2.** The results of Optical Coherence Tomography (OCT) of the Retinal Nerve Fiber Layer (RNFL) on October 5<sup>th</sup>, 2018. It showed the average thickness of the RNFL in the right eye was 117.49, and the left eye was 109.69.

Ophthalmological examination of the right eye revealed a visual acuity (VA) of 6/45, on Pinhole (PH) improved to 6/18. Close inspection of the conjunctiva revealed a conjunctival vascular injection (CVI), yellowish-white nodule measured around 2×3 millimeters on the superior part. He had an Intraocular Lens (IOL) implanted during the previous cataract surgery. On the other hand, the left eye was within normal limits, with VA 6/6. The right eye intraocular pressure was 10 mmHg, and the left eye was 12 mmHg. The patient was diagnosed with right eye nodular anterior scleritis + pseudophakia. He was prescribed Prednisolone 6 mg/0.6ml (P Pred™) eye drops, six times a day, each a drop on the right eye, Artificial tears (lyteers™) eye drops six times a day, each a drop on the right eye, and Diclofenac sodium tablets 50 milligrams twice daily. He was advised to

come back to the clinic in a week.

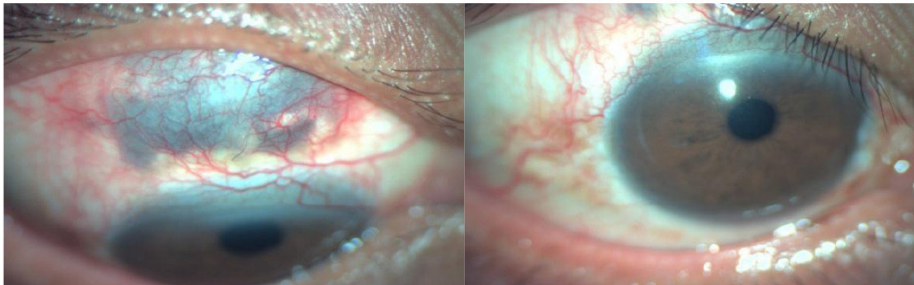
The patient came back for reevaluation on October 12<sup>th</sup>, 2018 after conducting a laboratory examination. The total cholesterol was 248 mg/dL, triglycerides 106 mg/dL, HDL cholesterol 60 mg/dL, LDL cholesterol 169 mg/dL, BUN 14.40 mg/dL, creatinine 1.09 mg/dL, fasting blood sugar 94 mg/dL, and blood sugar 2 hours post-prandial 123 mg/dL. The case was consulted to the Internal Medicine Department for further analysis of Antinuclear Antibody (ANA) Immunofluorescence (IF) and Rheumatoid Factor (RF) as well as management of dyslipidemia. He was advised to visit the clinic after he had done the consultation and laboratory examination. On October 16<sup>th</sup>, 2018, the patient underwent an examination in the Internal Medicine Department and was diagnosed with dyslipidemia and was

prescribed oral Simvastatin 20 milligrams once a day.

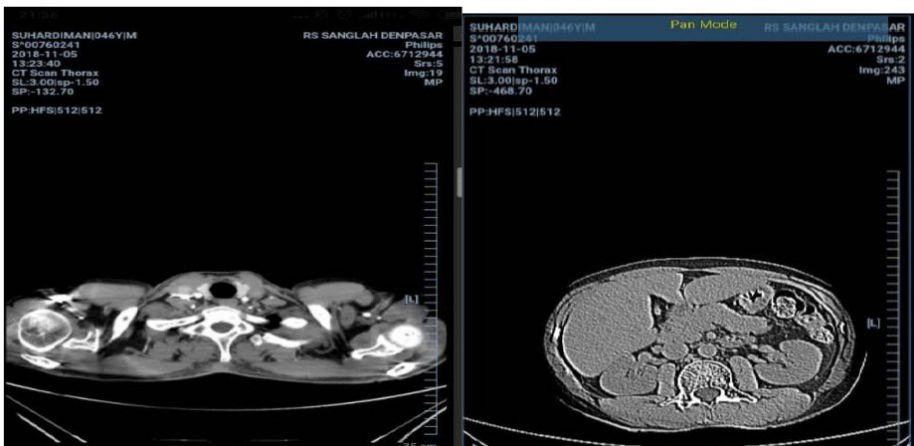
On October 28<sup>th</sup>, 2018, the patient came and showed ANA laboratory results. The titer was 1:1000, and the RF was negative. He was then referred for a reevaluation to the Rheumatology Division of the Internal Medicine Department. The patient underwent an examination at the Internal Medicine Department of the Rheumatology Division on October 29<sup>th</sup>, 2018, and was diagnosed with the right eye anterior nodular scleritis et causa suspected scleroderma differential diagnosis include Eosinophilic Granulomatous Polyarthritis and Systemic Lupus Erythematosus (SLE). He was prescribed immunosuppressant drugs, Imuran and oral steroid drugs with the doses according to the Ophthalmologist's prescription. He was planned for a chest computed tomography (CT) scan, consulted to the Pulmonology Department for spirometry and ANA profile examinations.

The patient underwent an examination at the Pulmonology Department on October 30<sup>th</sup>, 2018, with complaints of shortness of breath and heavy breathing accompanied by coughs and colds during that day. The patient was diagnosed with a mild attack of bronchial asthma in persistent partially controlled asthma. He was given Salbutamol 4 milligrams three times a day, Cetirizine 10 mg once a day, and a powder for inhalation containing a combination of Salmeterol xinafoate and Fluticasone propionate (Seretide™) inhaled twice daily for asthma controller.

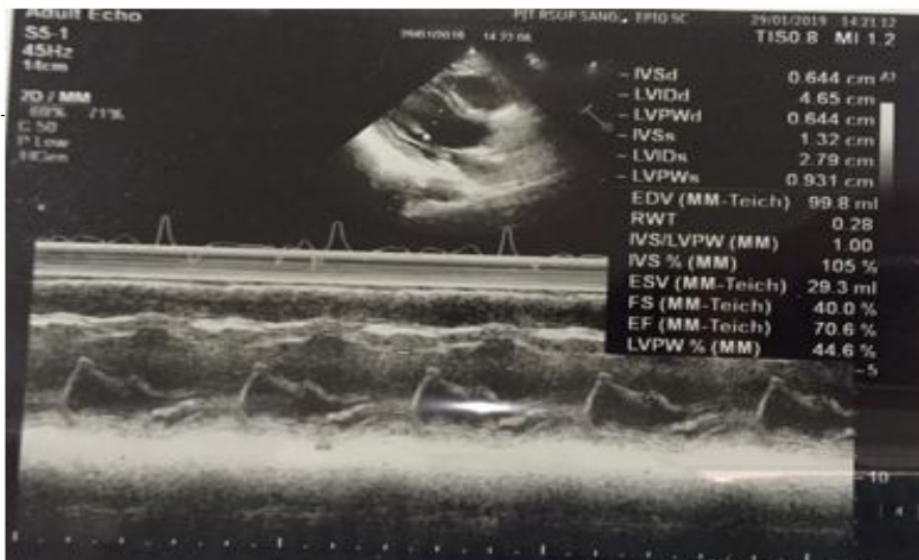
The patient came control to the polyclinic's Ophthalmology Department of the EED on November 5<sup>th</sup>, 2018 and performed an ophthalmological examination of the right eye. The result was VA 6/45, PH 6/18, minimal CVI, a yellowish-white, 1×1 millimeter nodule in the superior part of the conjunctiva, with IOL lens, fundus examination was within normal limit. The patient was diagnosed with right eye nodular anterior scleritis (improved) + pseudophakia. He has prescribed methylprednisolone tablets 16 milligrams three times daily, P Pred eye drops six times a drop on the right eye, artificial tears six times daily each a drop on the right eye, diclofenac sodium tablets 50 milligrams twice daily,



**Figure 3.** Photo of the patient's right eye on November 5<sup>th</sup>, 2018, where the dilated conjunctiva and sclera vessels had improved and the nodule size had reduced.



**Figure 4.** Chest CT scan (with contrast) imaging.



**Figure 5.** Photo of Patient's Echocardiography.

and revisit for evaluation in 3 days and graded dose adjustment (tapering off) the Methylprednisolone. In addition, a subjective refraction examination was planned for the next visit.

The patient returned to the clinic on November 8<sup>th</sup>, 2018, and the right eye intraocular pressure was 19 mmHg, and the left eye was 12 mmHg. The patient was corrected for subjective refraction. The right eye needed a spherical lens -3 D,

cylinder -2 D with the axis 90 degrees. The resultant visual acuity was 6/7.5, and the left eye was Plano. The patient had tried to adapt with the eyeglasses, but eventually, he felt uncomfortable and dizzy thus, he was not given any eyeglass prescription.

The patient underwent an examination at the Internal Medicine Department of Rheumatology Division on November 19<sup>th</sup>, 2018. The ANA profile was Centromere B (CB) +++ (3). The patient was diagnosed with systemic sclerosis. The therapy prescribed was Imuran tablet 50 milligrams twice daily, oral steroid medication doses following the Ophthalmologist's prescription, and Simvastatin 20 milligrams once daily.

The patient was consulted to the Glaucoma Division on January 7<sup>th</sup>, 2019, and the right eye intraocular pressure was 25 mmHg and 20 mmHg for the right and left eye, respectively. In the Goldmann Applanation Tonometry examination, the right eye was 26, and the left eye was 14. The patient was diagnosed with right eye nodular anterior scleritis (improved) + pseudophakia + secondary glaucoma. The therapy prescribed was Betaxolol 0.5% eye drops (Tonor<sup>TM</sup>), a drop twice daily each on the right eye, other treatment following the previously prescribed by the EED division.

The patient returned on January 9<sup>th</sup>, 2019, and showed the chest CT scan results on November 6<sup>th</sup>, 2018. There was no representation of fibrosis, signs of inflammation, or other abnormalities in the right and left lung parenchyma and the mediastinum at the moment. The results of spirometry were the following, FVC 41.1% and FEV<sub>1</sub>/FVC 88.32%. It indicated a severe restrictive lung disease. The patient was diagnosed with controlled bronchial asthma with severe restrictive lung disease et causa systemic sclerosis. The therapy prescribed Seretide disc a puff twice daily, a combination of Ipratropium bromide and Salbutamol sulfate (Combivent<sup>TM</sup>) nebulizer if needed, and no specific treatment for the severe restrictive lung disease.

The patient came back for evaluation to the EED Division on January 21<sup>st</sup>, 2019, and got an intraocular pressure examination. It measured 19 mmHg in both eyes. The patient was diagnosed



**Figure 6.** Photo of the patient's right eye on March 5<sup>th</sup>, 2019, there were no dilated conjunctival and sclera vessels and no nodules.

with right eye nodular anterior scleritis (improved) + pseudophakia + follow-up secondary glaucoma. The therapy prescribed Methylprednisolone tablet 4 milligrams twice daily (tapered off to 4 milligrams once daily, artificial tears a drops six times a day on the right eye.

The patient underwent an examination at the Cardiology Department on January 29<sup>th</sup>, 2019, and underwent echocardiography. Echocardiography was normal left and right ventricle (LV and RV) systolic function, normal LV diastolic function. The patient was diagnosed with dyspnea et causa suspected pulmonary hypertension + systemic sclerosis.

The patient revisits the EED Division and Glaucoma Division on March 5<sup>th</sup>, 2019. He underwent an ophthalmology examination of the right eye. The VA was 6/45 PH 6/18, the anterior segment and palpebral was normal, clear conjunctiva, clear cornea, normal anterior chamber, round and regular iris, and positive pupillary reflex, IOL (+), clear vitreous, fundus examination revealed round papilla, the Cranial nerve II appears well-defined, CDR 0.4, aa/vv 2/3, retina within normal limit, and macular reflex (+). Meanwhile, the left eye VA was 6/6 visual, the anterior segment and palpebral was normal, clear conjunctiva, clear cornea, normal anterior chamber, round and regular iris, positive pupillary reflex, clear lens, clear vitreous, fundus examination reveals papilla II nerve appears well-defined, CDR 0.4 aa/vv 2/3, and the macular reflex (+). The right eye intraocular pressure was 18, and the left eye was 19. The patient was diagnosed with right eye nodular (improved) anterior scleritis + pseudophakia + follow-up secondary glaucoma. The therapy prescribed was Methylprednisolone tablets 4 milligrams once every two days,

artificial tears eye drops six times a day on the right eye.

## DISCUSSION

Scleritis is frequently associated with systemic immunology disease that causes significant pain and causes vascular abnormalities in the eye. This scleritis rarely occurs in children, often occurs at the age of 40 to 60, and is more common in women. The onset of scleritis is usually gradual, lasting over several days. Patients with scleritis experience sharp pain in the eye that worsens at night and interferes with sleep. The pain may radiate to the head or face on the involved side, and usually, the eyeball is felt soft, unlike glaucoma. The inflamed sclera has a visible purplish color.<sup>4</sup> This case is about a male patient, 46 years old, who presented with complaints of red, watery, and visible white membrane on his right eye for two weeks. Patients complained of pain in the right eye, especially when moving the eyeball. Pain that feels like stabbing and persists. The patient also complained of headaches for two weeks. The patient complained of blurred vision, which has not been noticed for a long time.

Scleritis can be classified clinically based on anatomy, frequent anterior scleritis, and posterior scleritis. Diffuse anterior scleritis is a scleritis with a form associated with rheumatoid arthritis, ophthalmic herpes zoster, and gouty arthritis. Nodular anterior scleritis is a scleritis characterized by the presence of one or more erythematous inflammatory nodules, immobile, and pain in the anterior sclera. About 20% of cases may develop into necrotic scleritis. Necrotic scleritis can be divided into two, which are necrotizing anterior scleritis with

inflammation and without inflammation. Patients with nodular anterior scleritis may develop necrotizing anterior scleritis and therefore need strict supervision.<sup>5</sup> The patient, in this case, had red eyes due to the dilation of the blood vessels under the conjunctiva and sclera. There is a nodule in the superior conjunctiva and a thinning of the sclera after the nodule's resolution. It is worth to note that we need to test it with topical 2.5% phenylephrine drops, and indeed it was seen that the blood vessels remained dilated and did not disappear. Inflammation of the sclera can progress into ischemia and necrosis, which will lead to thinning of the sclera.

Systemic sclerosis is a multi-system autoimmune disease characterized by extensive vascular injury and fibrosis of the skin and internal organs.<sup>1</sup> The marker is clinical heterogeneity with varying degrees of disease expression, organ involvement, and a good prognosis. The term systemic sclerosis or scleroderma describes a patient who presents with small vessel vasculopathy, autoantibody production, and fibroblast dysfunction, thereby increasing extracellular matrix storage.<sup>2,6</sup> Clinical manifestations and prognosis are diverse, with most patients experiencing thickening of the skin and some involving internal organs.<sup>2,7</sup>

In this case, the patient had an abnormality in the immune system after laboratory examinations showed that ANA IF is 1/1000 and ANA profile is centromere B +++ (3). The diagnosis was systemic sclerosis. Laboratory tests are needed to support the diagnosis and also determine the prognosis. In addition, supporting examinations were carried out by other laboratory examinations, including RE, chest CT scan with contrast, spirometry, and echocardiography to look for abnormalities in other organs. The patient, in this case, complained of a thickening of the skin on the hands and face so that sometimes the skin appears peeled on the hands.

Scleritis requires systemic treatment. Patients who were diagnosed with comorbidities will require specific treatment as well.<sup>8</sup> Nonsteroidal Anti-Inflammatory Drugs (NSAIDs) are generally effective in about one-third of patients with diffuse anterior

scleritis and two-thirds of patients with nodular anterior scleritis. The initial treatment for scleritis is generally with systemic NSAIDs, corticosteroids, or immunomodulatory drugs. Corticosteroid and immunosuppressive therapy is the most commonly used systemic scleritis therapy.<sup>9</sup> The patient, in this case, was given oral and topical corticosteroids, systemic non-steroidal anti-inflammatory drugs (NSAIDs), and artificial tears. Oral and topical corticosteroid therapy in this patient was tapered gradually. Additionally, the patient needed an oral immunosuppressive therapy combined with oral corticosteroids to counter the disease's systemic effect from the internal medicine perspective. The prognosis of scleritis in this patient is good and has improved substantially over the treatment period.

## CONCLUSION

This report is about a rare case of patients complained of redness and painful eyes due to systemic sclerosis and improved substantially with corticosteroid and immunosuppressant treatment. Systemic sclerosis is rare, even more so with a

primary complaint of scleritis. It is very important to understand that dilation of the sclera vessels is not always caused by a common inflammatory or infectious process but also due to the autoimmune disease. A proper examination from an ophthalmologist can help to diagnose and prevent the recurrence of the disease.

## PATIENT CONSENT

The patients had given consent regarding the publication of the case in an academic journal without revealing the personal identity.

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## AUTHOR CONTRIBUTION

All authors contributed equally in all phases of the study.

## CONFLICT OF INTEREST

The authors declared no potential conflicts of interest concerning the research, authorship, and/or publication of this article.

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