Case Report

Nodular Posterior Scleritis Masquerading as Amelanotic Choroidal Melanoma

Asima Rafique[1], Muhammad Shaheer[2], Muhammad Suhail Sarwar[3]
Department of Ophthalmology, [1-3]Mayo Hospital, Lahore

ABSTRACT

We report a patient of choroidal mass, masquerading as amelanotic choroidal melanoma. A 48 years old male presented in outpatient department (OPD) with painless decrease in vision of left eye. Fundus examination revealed a two disc diameter, non-pigmented, sub-retinal lesion with approximately 3mm basal diameter, superior to the disc with no choroidal folds, retinal detachment or pigmentation over the lesion. B scan revealed a small nodular thickening superior to the optic nerve head with moderate to low internal reflectively and no choroidal excavation. Swept-Source optical coherence tomography (SS OCT) showed massive elevation of retina due to underlying scleral thickening. He was diagnosed as nodular posterior scleritis (NPS). The lesion regressed completely after treatment with topical and systemic Non-steroidal anti-inflammatory drugs (NSAIDs). Despite its low prevalence, NPS should be kept in differential diagnosis of an amelanotic choroidal mass.

Key Words: Scleritis, Melanoma, Optical Coherence Tomography.

How to Cite this Article: Rafique A, Shaheer M, Sarwar MS. Nodular Posterior Scleritis Masquerading as Amelanotic Choroidal Melanoma. Pak J Ophthalmol. 2022, **38 (4):** 292-295.

Doi: 10.36351/pjo.v38i4.1392

Correspondence: Muhammad Shaheer
Department of Ophthalmology, Mayo Hospital, Lahore
Email: mshaheer212@gmail.com

Received: March 30, 2022
Accepted: June 04, 2022

INTRODUCTION

Posterior scleritis is a potentially blinding, often misdiagnosed form of scleritis due to its inconsistent manifestations, often labelled as pseudo melanomas.[1] Most of the cases of posterior scleritis are isolated, but some of the cases can be associated with systemic diseases.[2] It can present as diffuse or sectoral scleral involvement. The latter is particularly notorious for resembling choroidal granuloma, hemangioma and most of all amelanotic choroidal melanoma, which can lead to inadvertent treatment options like enucleation, chemotherapy or radiotherapy.[3] A case review series of 400 referred ocular oncology patients showed that 1.5% cases of posterior scleritis were misdiagnosed as choroidal melanoma leading to misguided treatment even enucleation.[4] The relevant ancillary tests need to be interpreted correctly for making a correct diagnosis. In this case report, we aim to present a case of NPS masquerading amelanotic choroidal melanoma, treated successfully with topical and systemic NSAIDs.

Case Report

A 48 years old healthy male presented in OPD on January 15th, 2022 with complaints of loss of vision in right eye associated with redness and peri orbital pain for past 3 weeks and painless decrease in vision of his left eye. His past medical and ocular history was not significant. Visual acuity in the right eye was normal ''perception and projection of light'' in all four quadrants while best corrected visual acuity (BCVA) in the left eye was 6/60. Extra ocular motility was normal in all gazes. Pupillary reactions were normal in the left eye but could not be elicited in the right eye due to media opacity. However, there was no relevant afferent pupillary defect (APD). Examination of right eye showed mild periorbital swelling, diffuse conjunctival injection and hazy cornea with central 2.5 x 3 mm area of corneal thinning. There was shallow anterior chamber, Van Herick ''grade II'' and mature cataract. The anterior segment examination of left eye was unremarkable and no inflammatory cells were

noted in the anterior vitreous phase. The intraocular pressure of left eye was 16 mmHg while right eye was digitally soft. The dilated fundus examination of left eye revealed a 2 disc diameter (DD), approximately 3mm in basal diameter, non- pigment, sub-retinal lesion, superior to the disc with superonasal arcade crossing over it (Figure 1). There was no evidence of sub retinal fluid, hemorrhages, pigmentary changes, lipofuscin deposition, choroidal folds or disc edema. Red free photograph of the left eye depicted a corresponding elevated mass with no overlying vessels (Figure 1).

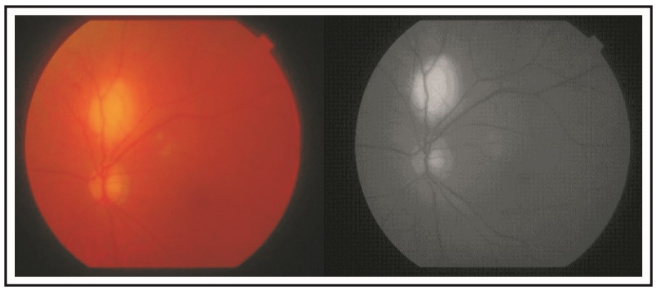

Figure 1: Colored Fundus Photograph of left eye showing 2 DD of non- pigmented, sub-retinal lesion superior to the disc with superonasal arcade crossing over it (Left).Red Free Photograph of left eye showing elevated mass with no overlying vessels (Right).

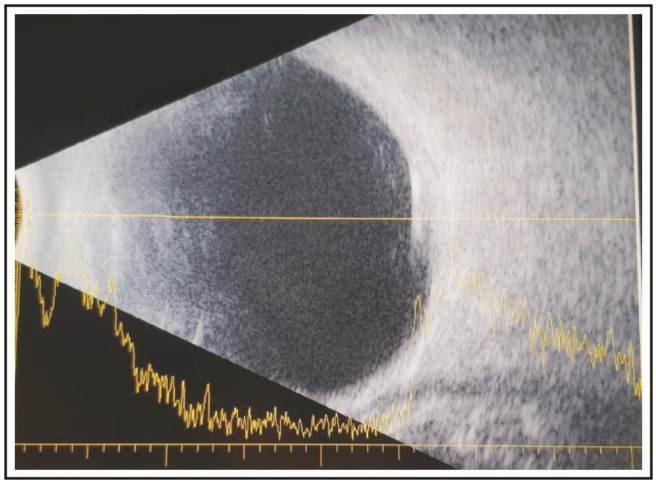

Figure 2: B scan of left eye showing small nodular thickening of sclera. A Scan shows medium reflectivity spikes at sclera-choroidal level.

B-scan of left eye revealed a homogenous lesion superior to the optic nerve head with moderate to low internal reflectively suggestive of small nodular scleral thickening approximately 3-4 mm in basal diameter and 1.5 mm in elevation with no exudative retinal detachment, choroidal excavation, collar stud sign, orbital shadowing or definitive T-sign (Figure 2). On A-scan, low to medium reflectivity spikes were noted at sclera-choroidal level; approximately 40% or less than the height of retinal spike.

Swept-Source optical coherence tomography (SS OCT) revealed elevation of retina due to underlying scleral thickening, with normal choroidal tissue and vasculature and no sub retinal fluid (Figure 3). The high resolution SS OCT clearly indicates the pathology to be in scleral tissue rather than choroid obliviating the need for invasive test like fundus fluorescein angiography (FFA).

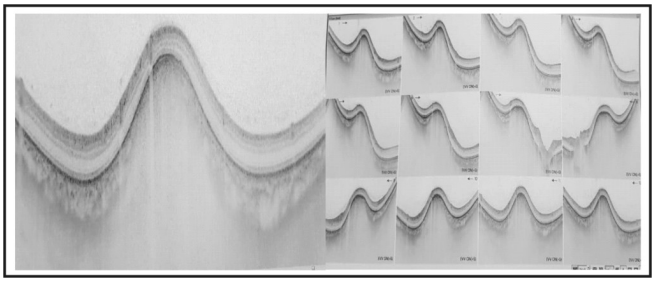

Figure 3: SS OCT of left eye showing elevation of retina due to underlying scleral thickening (Left). Sequential Photographs of SS OCT (Right).

Magnetic resonance imaging (MRI) of the orbit and brain revealed a focal, elevated, enhancing lesion in the superotemporal part of the left eye bulging into the vitreous cavity. Post contrast images depicted diffuse enhancement of the lesion along with enhancement of peri ocular tissue posterior to the globe (Figure 4).

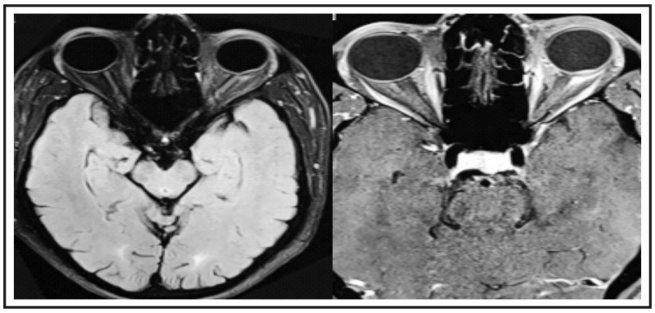

Figure 4: MRI Scan Axial Section of Brain and Orbit revealed focal, elevated lesion bulging into the eyeball (Left). Post Contrast Image showing diffuse enhancement of the lesion (Right).

Detailed inflammatory disease workup including Erythrocyte sedimentation rate (ESR), C-Reactive protein (CRP), serum Angiotensin converting enzyme

(ACE), Quantiferon TB Gold standard test, Rheumatoid arthritis (RA) factor, antinuclear antibodies (ANA) and venereal disease research laboratory (VDRL) for syphilis was done. Chest x- ray was also performed. All tests were unremarkable. The patient was referred to an internist for detailed evaluation. The non-pigmented appearance of the lesion and normal intrinsic choroidal vasculature along with B scan, MRI and SS OCT findings led to the diagnosis of NPS ruling out amelanotic choroidal melanoma.

Topical NSAIDs (three times a day) and Flurbiprofen tablet 100 mg (twice daily) were advised. Oral steroids were avoided due to corneal thinning in the right eye. A temporary tarsorrhaphy was performed in the right eye to prevent inadvertent trauma to eyeball due to blinking. Patient was called on follow up after 2 weeks.

Our case showed that NPS can simulate a choroidal tumor. In this case, B scan and SS OCT findings led to the diagnosis of NPS. However, malignancy could still not be ruled out confidently but complete resolution of lesion with topical and systemic NSAID's confirmed the diagnosis of NPS.

DISCUSSION

Posterior scleritis is inflammation of sclera posterior to ora serrata. NPS has been reported only as case reports.[5] Clinically, pain in eye is a common symptom that can lead the ophthalmologist towards the diagnosis of inflammatory disease with conjunctival injection or painful extraocular movements. On fundus examination, absence of halo and no lipofuscin deposition are indicative of NPS.[6] On reviewing the literature, it has been noted that mean age of presentation of a patient of NPS is 50 years with female preponderance.[7] The age of the patient is consistent with the available literature; however, there were no complaints of pain or redness. B scan plays a pivotal role in differentiating NPS from malignant conditions. Malignant melanomas will demonstrate mixed echogenicity with medium to low amplitude echoes and a collar stud shape.[8] MRI serves as an additional diagnostic modality to differentiate the inflammatory process from malignant lesion. MRI of the orbit and brain of our patient revealed a focal, elevated, enhancing lesion in the superotemporal part of the left eyeball bulging into the vitreous cavity.[9] FFA has been considered to be diagnostic of choroidal melanoma with characteristic double circulation while such a pattern is absent in case of posterior scleritis. We did not perform FFA in our patient considering it an invasive test because enough information was gathered from B scan, MRI and SS OCT. In our opinion, OCT is a very important diagnostic tool for NPS, particularly when the sub retinal mass is not very large. In this particular case, the scans depicted that choroidal tissue was normal and vessels of choroid were of normal configuration.[10] Treatment options vary according to the severity of disease, which include NSAIDs to even immunosuppressant. Our patient was successfully managed with topical and systemic NSAIDs for a period of 12 weeks. Post treatment visual acuity of the patient improved to counting fingers in the right eye and 6/9 in the left eye. Post treatment colored fundus photograph revealed complete resolution of mass (Figure 5). It is this therapeutic response which helped to confirm the diagnosis to be of inflammatory nature rather than neoplastic.

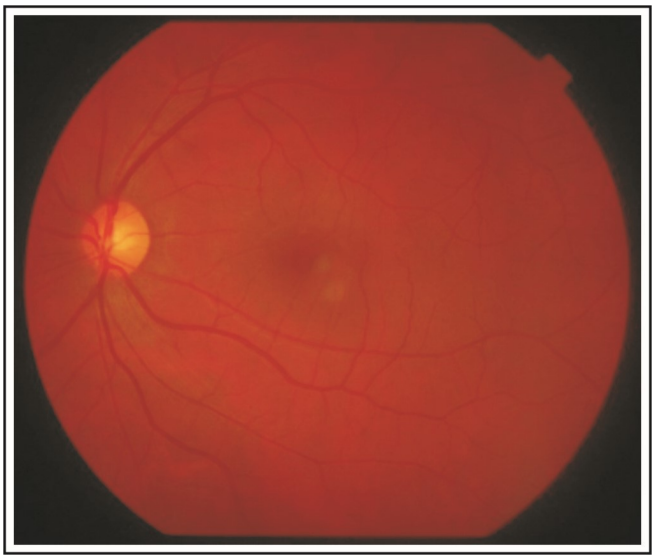

Figure 5: Post Treatment Colored Fundus Photograph of left eye showing complete resolution of mass.

The prognosis of NPS is excellent, with no recurrence reported in the literature (Table 1). Most of the cases respond to systemic steroids with a few cases treated with immunosuppressive agents (Table 2).

Table 1: *Literature Review of Patients with NPS: Demographics and Historical Findings.*

Study	Age	Gender	Laterality	Systemic Work up	Initial Visual Acuity	Final Visual Acuity
Current Case, 2022	48	Male	U/L	-ve	20/200	
Waleed, 2020	25	Male	U/L	-ve	CF 1 foot	20/20
Shibata, 2019	59	Female	U/L	Hepatitis B	20/200	N/A
Hatef, 2010	55	Female	U/L	-ve	20/40	20/20
Finger, 1990	66	Male	U/L	-ve	20/200	N/A

Table 2: *Treatment Modalities and Outcomes of Patients with NPS.*

Study	Management	Outcome
Current case, 2022	Topical and Systemic NSAIDs	Resolution
Waleed, 2020	IV steroids followed by oral steroids and Mycofenolate Mofetil	Resolution
Shibata, 2019	Oral Steroids (20 mg)	Resolution
Hatef, 2010	IV steroids (1g/day) followed by oral steroids (60 mg) + Mycofenolate Mofetil	Resolution
Finger, 1990	Enucleation	-

CONCLUSION

Choroidal mass can present a diagnostic challenge for ophthalmologists. Clinical misinterpretation of NPS as malignant melanoma can lead to misguided therapy and devastating outcomes, including enucleation. In any instance of diagnostic uncertainity, a trial of anti-inflammatory medication may serve as a therapeutic test and help spare patients from unnecessary intervention.

Conflict of Interest: Authors declared no conflict of interest.

REFERENCES

1. **Sin PY, Liu DT, Young AL.** Nodular Posterior Scleritis Mimicking Choroidal Tumor in a Patient with Systemic Lupus Erythematous: A Case Report and Literature Review. Asia Pac J Ophthalmol (Phila). 2016; **5 (5):** 324-329. doi: 10.1097/APO.0000000000000165.
2. **Lavric A, Gonzalez-Lopez JJ, Majumder PD, Bansal N, Biswas J, Pavesio C, et al.** Posterior Scleritis: Analysis of Epidemiology, Clinical Factors, and Risk of Recurrence in a Cohort of 114 Patients. Ocul Immunol Inflamm. 2016; **24 (1):** 6-15. doi: 10.3109/09273948.2015.1005240.
3. **Shukla D, Kim R.** Giant nodular posterior scleritis simulating choroidal melanoma. Indian J Ophthalmol. 2006; **54 (2):** 120-122. doi: 10.4103/0301-4738.25835.
4. **Liu AT, Luk FO, Chan CK.** A case of giant nodular posterior scleritis mimicking choroidal malignancy. Indian J Ophthalmol. 2015; **63 (12):** 919-921. doi: 10.4103/0301-4738.176038.
5. **Hage R, Jean-Charles A, Guyomarch J, Rahimian O, Donnio A, Merle H.** Nodular posterior scleritis mimicking choroidal metastasis: a report of two cases. Clin Ophthalmol. 2011; **5:** 877-880. doi: 10.2147/OPTH.S21255.
6. **Sainz de la Maza M, Foster CS, Jabbur NS.** Scleritis associated with systemic vasculitic diseases. Ophthalmology, 1995; **102 (4):** 687-692. doi: 10.1016/s0161-6420(95)30970-0.
7. **Alsarhani WK, Abu El-Asrar AM.** Multimodal Imaging of Nodular Posterior Scleritis: Case Report and Review of the Literature. Middle East Afr J Ophthalmol. 2020; **27 (2):** 134-138. doi: 10.4103/meajo.MEAJO_115_20.
8. Echography (Ultrasound) Procedures for the Collaborative Ocular Melanoma Study (COMS), Report No. 12, Part II. J Ophthalmic Nurs Technol. 1999; **18 (5):** 219-232.
9. **Osman Saatci A, Saatci I, Kocak N, Durak I.** Magnetic resonance imaging characteristics of posterior scleritis mimicking choroidal mass. Eur J Radiol. 2001; **39 (2):** 88-91. doi: 10.1016/s0720-048x(01)00278-9.
10. **Ozkaya A, Alagoz C, Koc A, Ozkaya HM, Yazıcı AT.** A case of nodular posterior scleritis mimicking choroidal mass. Saudi J Ophthalmol. 2015; **29 (2):** 165-168. doi: 10.1016/j.sjopt.2014.06.012.

Authors' Designation and Contribution

Asima Rafique; Medical Officer: *Concepts, Literature search, Data acquisition, Manuscript preparation, Manuscript editing.*

Muhammad Shaheer; Assistant Professor: *Literature search, Data acquisition, Manuscript review.*

Muhammad Suhail Sarwar; Professor: *Data acquisition, Data analysis.*