Case report

Choroidal Lymphoma Masquerading As A Rhegmatogenous Retinal Detachment : When Reasoning Prevails Over Appearances- A Rare Case Report

ABSTRACT

AIMS
The objective of this case report is to promote a structured reasoning that enabled us to overcome the masquerade syndrome in the setting of choroidal lymphoma.

INTRODUCTION
Choroidal lymphoma is a rare pathologic entity commonly misdiagnosed as a variety of other disorders. We report a rare case of choroidal lymphoma that was revealed by a chronic decrease in visual acuity and a total retinal detachment initially labeled as a rhegmatogenous retinal detachment.

CASE PRESENTATION
We report the case of a 45-year-old woman who suffered from chronic vision loss in her right eye. She was referred to our department with the diagnosis of a rhegmatogenous retinal detachment and was scheduled to undergo vitreoretinal surgery. However just before the surgery, the chief doctor noted some inconsistencies in her case that prompted further investigations. The diagnosis shifted to a suspicion of choroidal lymphoma, which was later confirmed by histopathological examination.

DISCUSSION
Choroidal lymphoma is a subset of uveal lymphoma. It is a rare condition and the diagnosis is often challenging as it may masquerade as other ocular diseases. The typical manifestations of fundus involvement are solid thickening of the choroid, serous retinal detachment and anterior and/or posterior epibulbar extension. The diagnosis is confirmed by histopathology with immunohistochemistry and flow cytometry, B-cell non-Hodgkin lymphoma remaining the most predominant type.

CONCLUSION
Choroidal lymphoma is a frequently misdiagnosed condition, as was the case for our patient who was initially diagnosed with a rhegmatogenous retinal detachment. Structured reasoning allowed us to establish the correct diagnosis by remaining vigilant in the face of clinical inconsistencies.

KEY WORDS
Choroidal lymphoma ; retinal detachment **;** masquerade syndrome

1. INTRODUCTION

Choroidal lymphoma is a rare pathologic entity. As a consequence, many ophthalmologists are unfamiliar with it and commonly misdiagnosed it as a variety of other disorders.

We report a rare case of choroidal lymphoma that was revealed by a chronic decrease in visual acuity and a total retinal initially labeled as a rhegmatogenous retinal detachment.

The objective of this case presentation is to describe choroidal lymphoma and to promote a structured reasoning that enabled us to overcome the masquerade syndrome.

2. CASE PRESENTATION

We report the case of a 45-year-old woman who suffered from chronic vision loss in her right eye, which had been progressing for the past 3 years. She was not being monitored for any other particular pathology and was in a good general condition with no other complaints. The patient was financially disadvantaged and preferred to wait for improvement through self-medication rather than to seek medical advice. She eventually sought medical help due to the progressive worsening of her vision and was diagnosed with a rhegmatogenous retinal detachment in her right eye at a city doctor office. The patient was then referred to our department at the university hospital for treatment of her retinal detachment, this department serving as a reference center.

Upon admission, the patient was taken care of by the doctor in charge of retinal cases. At the examination, the visual acuity was at counting fingers at near in her right eye, the eye pressure was normal, the anterior chamber was calm and deep, iris normal, crystalline lens transparent. The fundus was examined with a contact lens revealing a total retinal detachment, bullous in some quadrants with peripheral lattice degenerations, without signs of proliferative vitreoretinopathy. The examination of her left eye revealed a corrected visual acuity of 20/20 and was unremarkable.

The patient was scheduled for retinal surgery to address her rhegmatogenous retinal detachment in the right eye. In accordance with protocol, the patient was re-examined by the chief physician the day before surgery, one week after her initial admission. At re-examination, the visual acuity remained stationary at counting fingers at near. She noticed a sub conjunctival pinkness superonasally hidden by the upper eyelid and sensitive to palpation (Fig.1). Upon further questioning, it was revealed that this pinkness was chronic and that the patient treated it periodically with some unidentified eye drops. The anterior segment of her right eye was still unremarkable and calm. At fundus examination, our chief physician did indeed find a total retinal detachment but she was suspicious of it. The detachment appeared bullous, suggesting a retinal tear with vitreous traction to allow the liquid to seep in with this quantity. However, there were only peripheral lattice degenerations without tears. By delving deeper into the examination, the senior doctor detected a choroidal detachment behind the retinal detachment almost completely masking the optic nerve head (Fig.2). At the conclusion of her examination, the chief physician was highly suspicious of this detachment and performed a B-mode ocular ultrasound. When performing it, the subconjunctival lesion was tender to pressure from the probe. The ultrasound revealed a total retinal detachment with an underlying choroidal detachment of intermediate echogenicity in accordance with what was found during the clinical examination (Fig.3). But the most surprising was to find extraocular lesions following the contour of the sclera of similar echogenicity of the choroidal detachment (Fig.3). They had lobed contours with a nodular appearance near the optic nerve. An orbito-cerebral angio-Magnetic resonance imaging (MRI) was ordered in this setting as our chief physician was suspicious of an ocular tumor with epibulbar extension. The results of the MRI were as follow: An irregular, poorly defined thickening of the posterior wall of the right orbital sclera, measuring 7 mm in maximum thickness, showed homogeneous enhancement after contrast injection, with

scleral and intraconal fat invasion reaching close proximity to the optic nerve and the medial and lateral rectus muscles, suggestive of a lymphoma (Fig.4). The brain sequences showed no abnormalities.
We also obtained a blood test. Red blood cell and white blood cell counts and platelet counts were within normal ranges . A lactate dehydrogenase (LDH) assay was also requested, revealing a high level of 485.75 IU/L, for a normal rate between 207 and 417 according to the laboratory references.
The overall picture of the patient was compatible with a choroidal lymphoma with anterior and posterior epibulbar extension.
The next step was to perform a biopsy to confirm the diagnosis of lymphoma. The choice of biopsy site was initially focused on the most accessible lesion, namely the pinkish subconjunctival lesion, with emphasis placed on the patient not taking any self-medication before the procedure, especially steroids, at the risk of distorting the anatomopathological results. The histopathological examination confirmed the presence of a B-cells lymphomatous proliferation. The patient was referred to the hematology department for a complete staging workup and appropriate management.

At the end of this process, we were particularly surprised by the way things turned out. The shift in diagnosis, treatment, and prognosis was a complete 180-degree turn. We commend our chief physician's reasoning, her maintenance of a high level of suspicion, and her refusal to be swayed by appearances but rather to seek consistency between the findings. We believe this message deserves to be shared with our community, and we hope to pass on these reasoning reflexes to practitioners.

3. DISCUSSION

Choroidal lymphoma is a subset of uveal lymphoma, which can be further classified as primary and secondary based on the presence of systemic lymphoma at the time of ocular presentation (Aronow et al. 2014). It is a rare condition and the literature on it is sparse, limited to a few case reports and case series. This may explain why many ophthalmologists are not sufficiently familiar with this condition, which delays diagnosis.

The diagnosis of choroidal lymphoma is often challenging as it may masquerade as other ocular diseases. The typical manifestations of fundus involvement are solid thickening of the choroid, serous retinal detachment and anterior and/or posterior epibulbar extension (White 2019).

Choroidal thickening can be clinically evident, especially if accompanied by choroidal detachment. However, ultrasonography remains superior with a high diagnostic value as it allows for better characterization of lesions in eyes with suspected choroidal lymphoma. Indeed, it allows visualization of choroidal thickening, which may be accompanied by areas of posterior epibulbar extension in the form of nodular masses strongly suggestive of choroidal lymphoma (Chang et al. 1996; Holz et al. 1999; Neudorfer et al. 2004; Theophanous et al. 2018). Nevertheless, it is important to remain very cautious, as a similar ultrasound appearance can be obtained in cases of diffuse choroidal tumor with extraocular extension.
In addition, anterior extension of the lymphomatous proliferation can also occur, taking the form of a pinkish salmon subconjunctival patch (Coupland et al. 2001; Sarraf et al. 2005; Mashayekhi et al. 2014; Jusufbegovic et al. 2015). The presence of the latter is an important indicator for the lymphoid infiltration and represents an accessible biopsy site for histopathological examination (Mashayekhi et al. 2014).
Besides, serous retinal detachment may be observed in eyes affected by choroidal lymphoma but is not a universal finding and when present, it is generally shallow (Aronow et al. 2014; Mashayekhi et al. 2014).

Histopathological examination with immunohistochemical and flow cytometry evaluation remains mandatory to confirm the diagnosis of choroidal lymphoma. The specimen can be obtained from anterior or posterior epibulbar extension of the lymphoid proliferation (Jusufbegovic et al. 2015). B-cell non-Hodgkin's lymphoma remains the most predominant type, as it was the case with our patient (Doycheva et al. 2015; Jusufbegovic et al. 2015; Yang et al. 2022).

Regarding treatment options: monitoring, radiotherapy, chemotherapy and immunotherapy (with rituximab) are all options for the management of choroidal lymphoma, on a case-by-case basis (Mahdizad et al. 2022).

In summary, the diagnosis of choroidal lymphoma can prove difficult, as was the case for our patient. The practitioners who initially took charge of her case focused on the obvious lesion, namely retinal detachment, classifying it as rhegmatogenous due to the incidental presence of peripheral retinal degenerations. However, subsequent inconsistencies led to a series of investigations which resulted in the diagnosis of choroidal lymphoma. This is why we wish to emphasize the importance of clinical reasoning, particularly for pathologies where mimicry is frequent.

4. CONCLUSION

Choroidal lymphoma is a frequently misdiagnosed condition, as was the case for our patient who was initially diagnosed with a rhegmatogenous retinal detachment. Structured reasoning allowed us to establish the correct diagnosis by remaining vigilant in the face of clinical inconsistencies.

CONSENT

As per university standards, patient written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the authors.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Authors hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT , COPILOT , etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

Aronow, Mary E., Craig A. Portell, John W. Sweetenham, and Arun D. Singh. 2014. “Uveal Lymphoma: Clinical Features, Diagnostic Studies, Treatment Selection, and Outcomes.” Ophthalmology 121 (1): 334–41. https://doi.org/10.1016/j.ophtha.2013.09.004.

Chang, T. S., S. F. Byrne, J. D. Gass, J. R. Hughes, R. N. Johnson, and T. G. Murray. 1996. “Echographic Findings in Benign Reactive Lymphoid Hyperplasia of the Choroid.” Archives of Ophthalmology (Chicago, Ill.: 1960) 114 (6): 669–75. https://doi.org/10.1001/archopht.1996.01100130661005.

Coupland, S. E., H. D. Foss, N. E. Bechrakis, M. Hummel, and H. Stein. 2001. “Secondary Ocular Involvement in Systemic ‘Memory’ B-Cell Lymphocytic Leukemia.” Ophthalmology 108 (7): 1289–95. https://doi.org/10.1016/s0161-6420(01)00594-2.

Doycheva, D., M. Zierhut, D. Süsskind, K. U. Bartz-Schmidt, and C. Deuter. 2015. “[Diagnostics and treatment of choroidal lymphoma].” Der Ophthalmologe: Zeitschrift Der Deutschen Ophthalmologischen Gesellschaft 112 (3): 217–22. https://doi.org/10.1007/s00347-014-3206-x.

Holz, F. G., H. V. Boehmer, G. Mechtersheimer, G. Ott, and H. E. Völcker. 1999. “Uveal Non-Hodgkin’s Lymphoma with Epibulbar Extension Simulating Choroidal Effusion Syndrome.” Retina (Philadelphia, Pa.) 19 (4): 343–46. https://doi.org/10.1097/00006982-199919040-00015.

Jusufbegovic, Denis, Vivian Kim, and Devron H. Char. 2015. “Primary Uveal Lymphoma with Epibulbar Extension Masquerading as an Intraocular Inflammation.” Canadian Journal of Ophthalmology 50 (1): e24–26. https://doi.org/10.1016/j.jcjo.2014.11.018.

Mahdizad, Zahra, Elias Khalili Pour, Mohammadreza Mehrabi Bahar, et al. 2022. “Management of Primary Choroidal Lymphoma Presenting as Extensive Serous Retinal Detachment and Salmon Patch of Bulbar Conjunctiva: A Case Report.” Journal of Medical Case Reports 16 (April): 162. https://doi.org/10.1186/s13256-022-03373-z.

Mashayekhi, Arman, Shripaad Y. Shukla, Jerry A. Shields, and Carol L. Shields. 2014. “Choroidal Lymphoma: Clinical Features and Association with Systemic Lymphoma.” Ophthalmology 121 (1): 342–51. https://doi.org/10.1016/j.ophtha.2013.06.046.

Neudorfer, Meira, Ada Kessler, Irene Anteby, Dafna Goldenberg, and Adiel Barak. 2004. “Co-Existence of Intraocular and Orbital Lymphoma.” Acta Ophthalmologica Scandinavica 82 (6): 754–61. https://doi.org/10.1111/j.1600-0420.2004.00354.x.

Sarraf, David, Atul Jain, Sander Dubovy, Allan Kreiger, Donald Fong, and John Paschal. 2005. “MUCOSA-ASSOCIATED LYMPHOID TISSUE LYMPHOMA WITH INTRAOCULAR INVOLVEMENT.” RETINA 25 (1): 94. https://journals.lww.com/retinajournal/abstract/2005/01000/mucosa_associated_lymphoid_tissue_lymphoma_with.18.aspx.

Theophanous, Christos, Melike Pekmezci, Bertil E. Damato, et al. 2018. “Choroidal Lymphoma Discovered on Ultrasound in a Patient with Suspected Corneal Tumor.” Ocular Oncology and Pathology 4 (5): 318–21. https://doi.org/10.1159/000486039.

White, Valerie A. 2019. “Understanding and Classification of Ocular Lymphomas.” Ocular Oncology and Pathology 5 (6): 379–86. https://doi.org/10.1159/000499845.

Yang, Ming, Taoran Zhang, Bojing Yan, and Yingxiang Huang. 2022. “Value of Combined Diagnosis for Choroidal Lymphoma: A Case Report.” Current Oncology 29 (11): 8835–45. https://doi.org/10.3390/curroncol29110695.

FIGURES

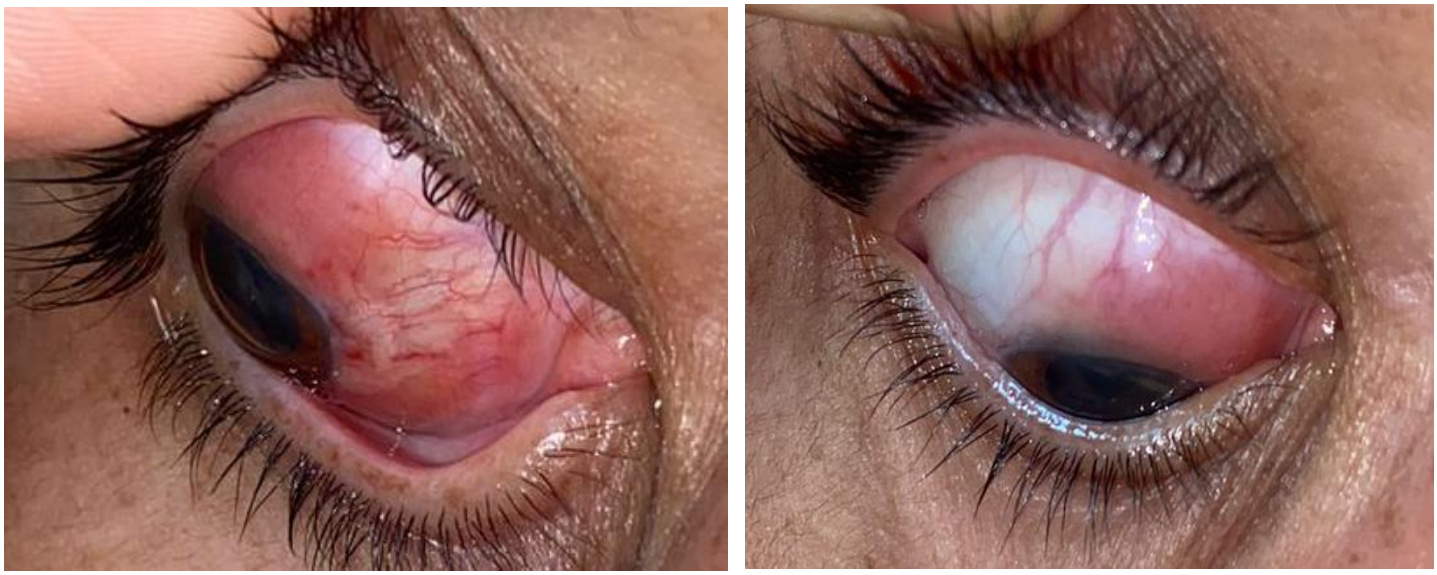

Fig.1. Salmon subconjunctival patch on clinical examination of the right eye, visible on lateral and down gazes

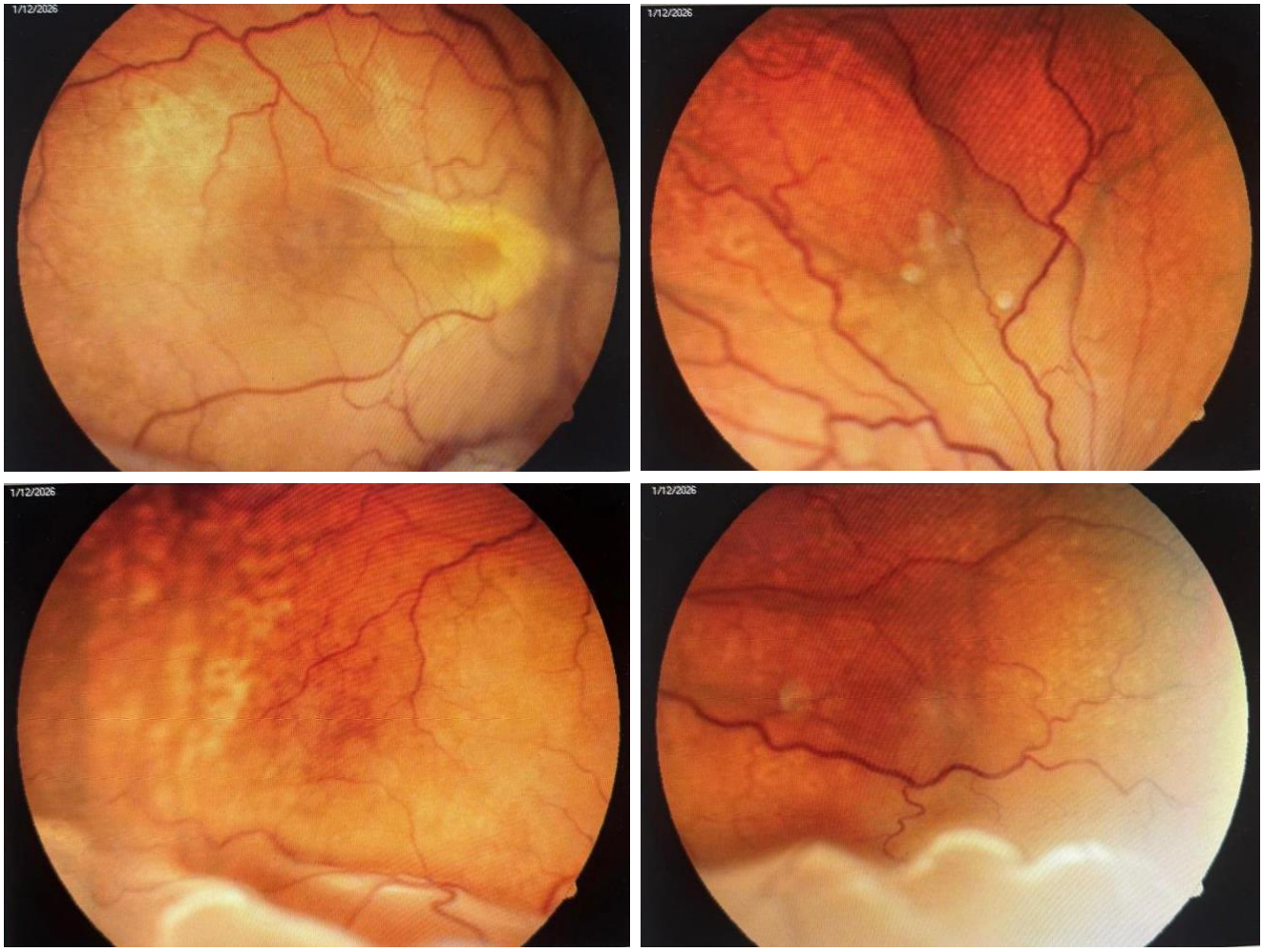

Fig.2. Fundus photographies of the right eye showing a retinal and choroidal detachment in all four quadrants

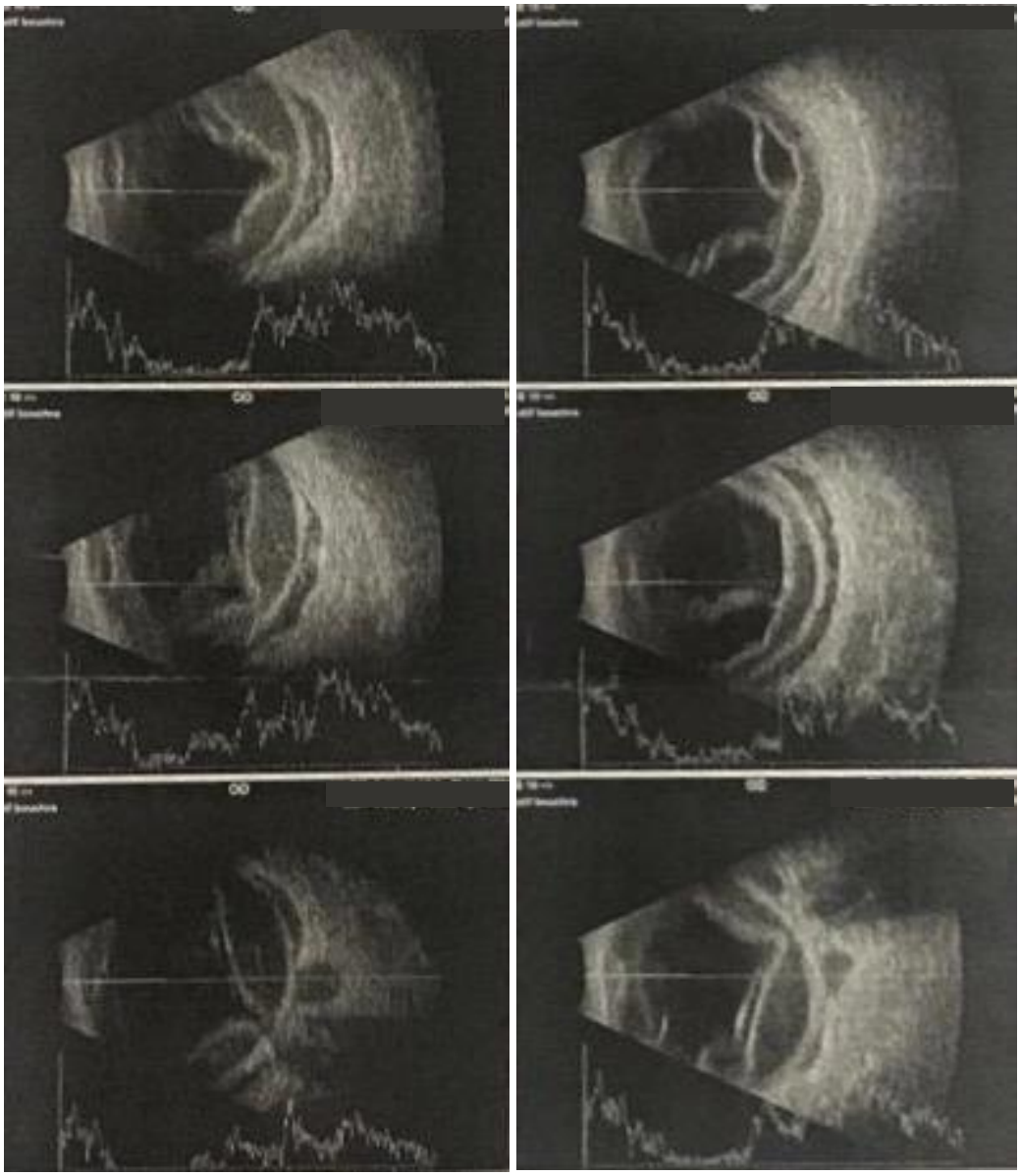

Fig.3. Mode-B ultrasound of the right eye showing a total retinal and choroidal detachments with posterior epibulbar extension as a multilobulated mass with a nodule adjacent to the optic nerve

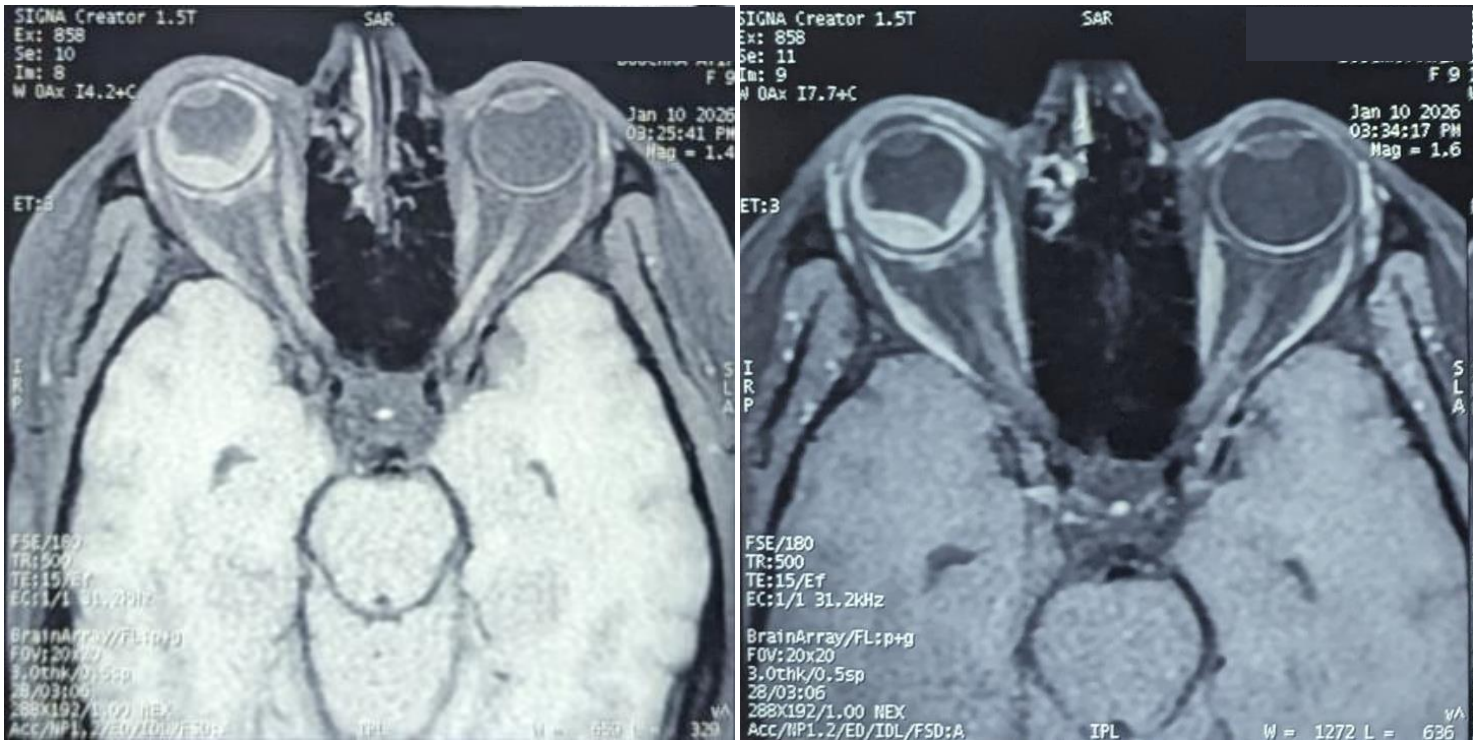

Fig.4. Magnetic Resonance Imaging of the orbits revealing an irregular, poorly defined thickening of the posterior wall of the right orbital sclera, measuring 7 mm in maximum thickness, with homogeneous enhancement after contrast injection, with scleral and intraconal fat invasion reaching close proximity to the optic nerve and the medial and lateral rectus muscles