



Short Communication

Uveitis masquerade syndrome

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ABSTRACT

Uveitis Masquerade syndrome comprises a group of disorders simulating a chronic idiopathic uveitis with an underlying primary cause. This article reviews both benign and malignant conditions that mimic as uveitis. Though uveitis masquerade syndromes are generally uncommon constituting less than 3% of patients in uveitis referral clinics and well under 1% of patients with uveitis seen in general ophthalmology, their recognition is essential for proper management and in some instances may be life saving.

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1. Introduction

Simulation of an ocular inflammatory condition by a neoplastic or non neoplastic disease is called as masquerade syndrome. The term “Masquerade Syndrome” was first used in Ophthalmology by Theodore in 1967 to describe a case of conjunctival carcinoma that manifested as chronic conjunctivitis. According to Martin’s dictionary word MASQUE (mask) generates the verb Masquerade meaning to appear in disguise and the noun masquerade pronounced as MAS-KER-AID meaning FALSE SHOW. Certain conditions of anterior and posterior segment can mimic uveitis and are therefore called as Masquerade syndromes. The common finding between the two is the “presence of cells and flare”.

Uveitis masquerade accounts for 2-5% of the total uveitic patients in referral centres.^{1,2}

1.1. Neoplastic conditions presenting as uveitis masquerade

This includes neoplasms such as Primary intra ocular (vitreo retinal) lymphoma, Primary choroidal lymphomas and

lymphoidal hyperplasia, Uveal melanoma, Retinoblastoma, Ocular metastasis from the lung and breast, Leukemia.

1.2. Non malignant conditions presenting as uveitis masquerade

Retained Intra ocular foreign body, Rhegmatogenous retinal detachment, Myopic degeneration, Pigment dispersion syndrome, Retinitis pigmentosa and Ocular ischemic syndrome. paediatric conditions like Coat’s disease and PHPV can present as atypical uveitis.

1.3. Primary intra ocular (vitreo retinal) lymphoma

This is a Non Hodgkin’s diffuse B cell lymphoma with or without CNS involvement presenting in the 6-7 th decade and this is the most common condition^{1,3} presenting with ocular masquerade like retinitis, vasculitis, vitritis or an uveal mass.

Primary choroidal lymphomas can present as multiple choroidal infiltrates.

1.4. Uveal melanoma

Frequent neoplasm of the eye can present as uveitis with sentinel vessels, choroidal granuloma and posterior scleritis.

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Ciliary body melanoma which contributes to 10% of the uveal melanoma, occurring in the 6th decade of life can present with anterior uveitis.

1.5. Retinoblastoma

This is the commonest paediatric intra ocular malignancy and one of the most feared uveitis masquerade misdiagnosis causes death. Though some inflammation can occur from tumor necrosis, more commonly tumor cells are mistaken for inflammatory cells. Tumor cells can form pseudo hypopyon.

1.6. Metastatic carcinoma

Primary tumors from the lung and breast can cause bilateral choroidal involvement with multiple gray yellow lesions; can be associated with vitritis, exudative RD and papilledema. Aik Hau Tan and Soon Phaik Chee in American Journal of Respiratory and Critical Care Medicine reported a case of adeno carcinoma right lung in a 73 year old male presented with right eye vitritis and snow balls⁴

1.7. Leukemia

Can present as iritis with diffuse or nodular iris thickening and hypopyon.

1.8. Benign masquerade syndrome

Svoszilkora in his study on benign masquerade syndromes in differential diagnosis of uveitis found that 79 out of 1112 patients (7.1%) presented with masquerade out of which 37 are malignant and 42 are benign masquerades.⁵

1.9. Ocular conditions

1.10. Retinitis pigmentosa

Dutta Majumder PMenia N, Roy R et al in their study from 30 years consecutive data quoted that 22 out of 8364 retinitis pigmentosa patients presented with uveitis.⁶

Ocular ischemic syndrome: 20% of OIS patients may have inflammatory cells in anterior chamber which are rarely more than grade 2, occasional keratic precipitates or posterior synechiae.⁷

1.11. Intra ocular foreign body

The reaction around the foreign body can simulate pars planitis or toxocariasis. Copper foreign body can cause acute intense inflammatory reaction while siderosis causes chronic anterior or intermediate inflammation. A typical feature of siderosis is RUST SPOT beneath the anterior lens capsule.

1.12. Rhegmatogenous RD

Can sometimes present with anterior uveitis including cells, flare and posterior synechiae.

Intra ocular medications: Triamcinolone acetonide molecules can migrate anteriorly and produce pseudo hypopyon; can lead on to sterile endophthalmitis.⁷

1.13. Anti VEGF

After one or two injections anti VEGF can cause transient anterior uveitis. But the incidence is low (0.026/ 1000 injections)⁷

1.14. Systemic medications

Rifabutin: Anti HIV drug known to cause severe intra ocular inflammation which is dose dependent and reversible.

2. Conclusion

Masquerade syndromes represent the uncommon presentation of rare disease. Most important step to diagnose neoplastic masquerade syndrome is clinical suspicion. Early diagnosis of malignancy is crucial both for vision and life saving which can be achieved by multi disciplinary approach, thorough history, careful ophthalmic examination aided by ancillary testing.

3. Source of funding

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4. Conflict of interest

None.

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