Case Report

Ocular Masquerade syndrome

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Abstract Masquerade syndromes comprises a group of disorders simulating a chronic idiopathic uveitis, having an underlying primary cause that is not immune mediated and that is associated with an apparent clinical picture of intraocular inflammation. The main disorders that can masquerade as a uveitis are intraocular tumours, postoperative infections or degenerative conditions. Reporting a 35-year male who presented with unilateral painless blurring of vision in left eye, 3 month old painless swelling in left testis. Ocular and systemic evaluation revealed chronic granulomatous panuveitis of left eye, spermatocytic seminoma of left testis, para aortic lymphadenopathy, bilateral enlarged adrenals and bilateral pneumatocele.
Key words: Chronic granulomatous panuveitis, Corticosteroids, spermatocytic seminoma of testis, Masquerade syndrome.

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INTRODUCTION

Masquerade syndromes are a rare group of disorders that mimic ocular inflammatory disorders can be infectious or neoplastic but they commonly refer to a neoplastic process.[1] The term 'Masquerade syndrome' was first used in ophthalmology by Theodore in 1967 to describe a conjunctival carcinoma that presented as chronic conjunctivitis.[2] Neoplastic masquerade syndromes constitute a minority of cases (2.3%) in uveitis clinics.[3]

Key features

It is usually bilateral but may have asymmetrical involvement. Cells are seen in aqueous or vitreous humour or both. Commonly associated with primary central nervous system lymphoma[1].

Associated features

They may initially respond to corticosteroids, but eventually becomes 'corticosteroid resistant'. Usually seen in older age group patients, may have a known history of malignancy elsewhere in the body. There is a lack of inflammatory features such as keratic precipitates and synechiae, but not universally[1].

We present a case of unilateral blurring of vision due to chronic granulomatous panuveitis as the initial manifestation of Masquerade syndrome.

CASE REPORT

A 35-year-old male presented with painless, unilateral blurring of vision in left eye of 15 days duration. On further enquiry history of infertility and a 3 month old mass in left testis was noted. On examination of left eye, visual acuity CF=2mt was recorded. Conjunctiva showed congestion with dilated, tortuous episcleral vessels present in all 4 quadrants. Cornea appeared steamy with loss of normal sheen. Posterior surface of cornea also showed multiple mutton fat KP's, which were brownish white in colour and of variable sizes, present in a triangular fashion in the inferior part. Anterior chamber contained 4+ cells and 1+ to 2+ flare. There was loss of normal iris pattern (muddy iris). Pupil was 5mm, round, regular and sluggishly reactive. In lens iris pigmentation was present in anterior capsule with early cortical cataract. Intraocular pressure of 18mm of Hg was seen on applanation tonometer. Fundus of left eye revealed hazy media with 4+ vitritis. Disc appeared slightly hyperaemic with normal vessels. Rest of the details could not be seen as media was hazy.

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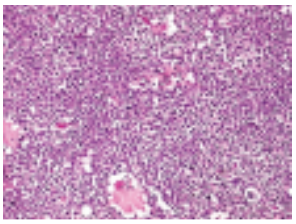
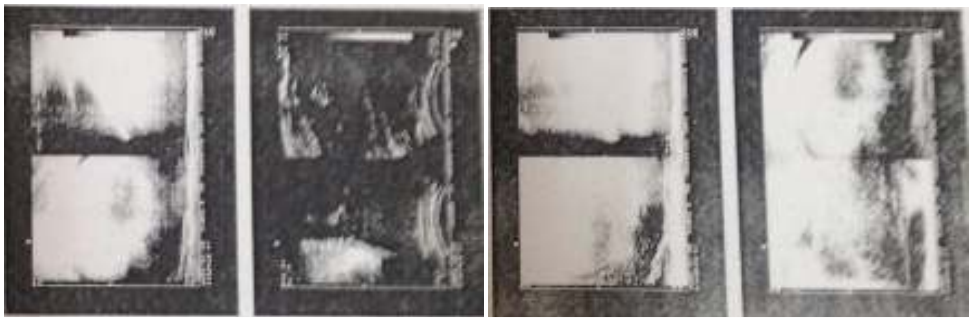


In right eye, visual acuity was 6/6. No abnormality was found in right eye other then early cortical cataract. Transscrotal open biopsy was taken from left testicular mass and send for histopathology, in which the reports suggested spermatocytic seminoma of testis. In view of clinical and pathological findings, left hemiscrotectomy with high inguinal orchidectomy was done. Post operative histopathology report confirmed it as spermatocytic seminoma of testis (stage Ib or T4NxMxSx). It also showed tumour cell infiltration in tunica vaginalis, albuginea and vasculosa, with infiltration into the spermatic cord. There was evidence of lymphovascular invasion also. Following this USG scrotum was done, which showed paraaortic lymphadenopathy, creating a suspicion of metastasis.



CT abdomen and pelvis was also done, which revealed retroperitoneal and left external iliac lymphadenopathy with bilateral enlarged adrenal glands. CT thorax showed no evidence of metastasis, but pneumatocele were present bilaterally in the lung field. Palliative therapy was planned, followed which chemotherapy with BEP 4 cycles was to be started. Blood and urine investigations were normal. Tumour markers were also within normal limit. Our patient was simultaneously started with treatment for uveitis. Nepafenac eye drop, Prednisolone acetate 1%eye drop, lubricant eye gel, Homatopine eye drop, Timolol maleate 0.5% eye drop, cap Indomethacine SR with vit A, vit C and vit B complex was given. With these treatment patient showed no improvement in both signs and symptom. Was referred for immunologist opinion Unfortunately, the patient passed away, in midst of the treatment due to cardio-respiratory arrest.

DISCUSSION

Masquerade syndromes are classically defined as those conditions that include, as part of their clinical findings, the presence of intraocular cells but are not due to immune-mediated uveitis entities. These may be divided into nonneoplastic conditions and neoplastic conditions. They account for nearly 5% of all uveitis patients at a tertiary referral center.[4] They are usually poorly responsive to corticosteroid treatment. Suspicion occurs when there is unilateral intraocular inflammation. The nonneoplastic masquerade syndromes classically include retinitis pigmentosa, ocular ischemic syndrome, and chronic peripheral rhegmatogenous retinal detachment.[4] Neoplastic masquerade syndrome may account for 2-3% of all patients seen in tertiary uveitis referral clinics. Majority of these are patients with intraocular involvement from primary CNS lymphoma.[4] Spermatocytic seminoma is a type of germ cell tumour (GCT), accounting for less than 1% GCTs. Unlike other GCTs, spermatocytic seminoma does not arise from ITGCN (intratubular germ cell neoplasia) and is not associated with a history of cryptorchidism or bilaterality.[5] The most common presentation of testicular cancer is a painless testicular mass. Although approximately two thirds of men with GCT have diminished fertility, it is an uncommon initial presentation.[5]

Diagnostic considerations

The family history, the past medical history, the ocular history, the review of systemic complaints, the general physical examination, the direct ocular examination, the clinical course and the response to treatment should always be considered to rule out not only infectious etiologies but also any malignant disorders that can cause an apparent intraocular inflammation. Systemic workup should include complete blood counting with differential classification, rheumatoid factor, antinuclear antibody, sedimentation rate, human immunodeficiency virus test, venereal disease research laboratory test, anticardiolipin antibody, antinuclear antibody, anticholinesterase enzyme, Mantoux and CT thorax etc. The tumour markers, Ultrasonography of scrotum, CT abdomen and

pelvis, CT thorax and histopathology of the tumour, help identify the testicular mass as spermatocytic seminoma of testis. It is also required to plan the treatment of the patient.

MANAGEMENT

A solid intratesticular mass in a postpubertal male should be considered a GCT until proven otherwise.[5] Inguinal orchidectomy with high ligation of the spermatic cord is performed. Testis sparing surgery for GCT is done if there is a small tumour in either a solitary testis or synchronous bilateral testicular masses. If elevated before orchidectomy, serum tumour marker levels should be measured after orchidectomy to determine if levels are declining, stable or rising. In our patient left hemiscrotectomy with high inguinal orchidectomy was done. Further palliative radiotherapy was planned, followed which chemotherapy with BEP 4 cycles was to be started.

CONCLUSION

It is an important impact on the life-expectancy of the patients, if malignant disorders that can masquerade as an uveitis could be early diagnosed for an early treatment. Hence a thorough history taking, like history of recurrent uveitis and not responding to steroids, including family history, ocular and systemic examination is to be emphasized. Timely diagnosis and intervention can save the life of the patient. The above said case conveys the importance of general examination in case of ocular pathologies, to identify potential diseases like neoplasms. Also within our limited knowledge, this is the only case reported showing uveitis masquerading in a patient of spermatocytic seminoma; but unfortunately it cannot be confirmed due to untimely demise of the patient.

INFORMED CONSENT

A written consent was obtained from the the patient before starting the treatment. They were also informed regarding
various treatment options and the publishing protocols.

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