University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2019, Vol. 5(9)

# A CASE OF BILATERAL INTERSTITIAL KERATITIS AS THE SOLE MANIFESTATION OF A LATE CONGENITAL SYPHILIS

**UMA V**
Department of Ophthalmology, THANJAVUR MEDICAL COLLEGE

**Abstract :** Interstitial keratitis is a nonulcerative stromal inflammation, commonly an immune mediated response to antigen, syphilis being a leading cause. Congenital syphilitic keratitis accounts for 90 per centof untreated cases in the age group of 5 to 25years and 10 per cent were acquired syphilitic keratitis. IK can be a sole presentation in an untreated late congenital syphilis as in this case report. Hence diagnosis of interstitial keratitis must be followed by screening for Treponemapallidum to prevent and treat its systemic manifestations and transmission.

**Keyword :**Interstitial keratitis, congenital syphilis

**INTRODUCTION**:

Interstitial keratitis is a common inflammatory sign of untreated late congenital syphilis in upto 50% of cases, most commonly girls in the age group of 5-25 yrs. This journal highlights the importance of diagnosis of an interstitial keratitis and screening for syphilis in a case of late latent congenital syphilis presenting with corneal stromal inflammation without any associated systemic features.

FIG 1

**CASE REPORT**:

A fourteen year old girl presented with history of redness, watering, photophobia and diminished vision in both eyes for past 3 months. On general examination revealed afebrile, no pallor, no jaundice, no cyanosis, no skin lesions, no lymphadenopathy, normal dentition, clinically normal cardiovascular, abdomen and central nervous system.

Ocular examination of right eye (Fig 2) revealed visual acuity of 6/36(P), lids normal , no lagophthalmos, no eyelash abnormalities, conjunctiva-palpebral part normal, bulbar congestion and circum corneal congestion present,dense greyish white deep stromal infiltration of 5*5mm with irregular margins involving the pupillary and parapupillaryarea with multiple pinhead sized dense infiltrations in the periphery and iris pigments over the back of inferior cornea, pink coloured deep radial bundle like vascularisation in the stroma along inferior limbus from 5 to 6 o'clock hours extending towards centre, anterior chamber appears normal depth, pupil 3 mm, reacting to both direct and consensual light and near reflex present lens appears clear.

**OD**
FIG 2

**OS**
FIG 3

Left eye (Fig 3) visual acuity of 6/24(P),lids normal, no lagophthalmos, no eyelash abnormalities, conjunctiva-palpebral part normal, bulbar congestion and circum corneal congestion present, multiple pinhead sized dense stromal infiltrations throughout the cornea with iris pigments, fresh and old keratic precipitates over the back of cornea. pink colored deep radial bundle like vascularisation seen at 1 o clock and 5 o clock position extending towards the cornea, mild anterior chamber reactions present,pupil irregular 3- 4 mm, sluggishly reacting to light and posterior synechiae extending from 10-11 o'clock, iris pigment clumps over anterior capsule of lens. BE-Fundus examination showed hazy media with normal disc, vessels and macula. Corneal sensation was normal in both eyes. On refraction, BCVA RE with -1.00DSPH/-1.00DCYL at 30 deg is 6/24(P) and BCVA LE with - 1.00DSPH/-0.50DCYL at 45 deg is 6/18 (P) Clinical diagnosis of BE interstitial keratitis and LEchronic iridocyclitis was made

**INVESTIGATIONS**:

**Ocular investigation:**

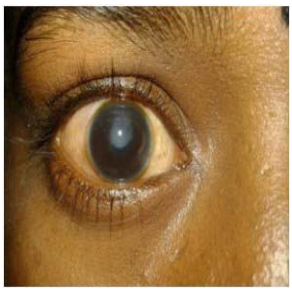
Tear film break-up time was normal, 20 seconds noted in both eyes. Schirmer's test(basic and reflex secretion) was normal, suggestive of normal tear production. On staining the corneal lesion using 2% fluorescein strip and examined using cobalt blue light of slitlamp revealed no epithelial defect. Staining with 1% Rose Bengal showed no devitalised epithelial cells, mucous or filaments. Intraocular pressure measured using Schiotz tonometer found to be 20.6mmHg and applanation tonometer could not be done due to distorted mires. Both the ducts were patent on syringing.

**Systemic investigation:**

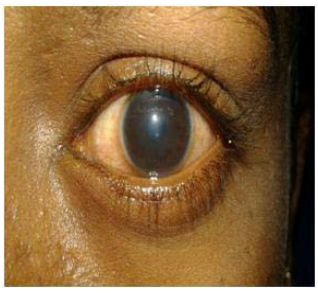
Ø Complete haemogram, random blood sugar, urine investigations and USG abdomen were normal.
Ø VDRL test found to be reactive in 1:32 dilution.
Ø Rapid plasma regain (RPR) test found to be reactive in 1:16 dilution
Ø InstacheckTreponemapallidum(TP)test found to be reactive
Ø TPHA test found to be reactive
Ø Testing the parents revealed reactive instacheck TP and RPRtests in father and in mother.
Ø Mantoux test non-reactive,sputum AFB negative,X-ray chest normal
Ø HIV ELISA testing found to be non-reactive.
Ø Cardiac evaluation with ECHO study was found to be normal.
Ø ENT evaluation found to be normal.
Ø CNS evaluation-CT brain was found to be normal.
Ø Lumbar puncture showed negative CSF VDRL, negative RPR, nil cells,protein-16mg/dl,sugar 50mg/dl ruling out the presence of neurosyphilis

**TREATMENT**:

Patient was started on topical 1% prednisolone acetate 2nd hourly,1% cyclopentolateb.d, and weeklyInj,Benzathine penicillin 2.4 million units; 1.2 million units intramuscularly in each gluteal region for 3 weeks.Patient showed signs of clearing of corneal stromal infiltrates andiridocyclitis and is being followed up(Fig 4,5).



**OD**
FIG 4



**OS**
FIG 5

**DISCUSSION**:

**INTERSTITIAL KERATITIS**:

IK is a non-suppurativeinflammation of the corneal stromathat features cellular infiltration and usually vascularisation without primary involvement of epithelium or endothelium. Keratitis is mostly associated with congenital syphilis. Late congenital syphilis occurs in untreated cases after 2 years of age.It is subclinical in 60% cases, clinical spectrum in remaining cases include interstitial keratitis.(5-25yrs),eighth-nerve deafness, recurrent arthropathy, Hutchinson teeth, Mulberry molars, sabre shin, saddle nose, rhagades, perforation of hard palate. Non-ulcerative stromal interstitial keratitis, often accompanied by anterior uveitis, is the most common inflammatory sign of untreated late congenital syphilis, occurring in upto 50% cases, most commonly in girls. Keratouveitis is thought to be an allergic type IV hypersensitivity response to T.pallidum in cornea. Congenitalsyphilis was the first infection to be linked with IK. Affected children typically show no evidence of corneal disease in their first years, stromal keratitis lasting for several weeks develops late in the first decade or later.

**CLINICAL PRESENTATION AND COURSE OF IK**:

Congenital syphilitic keratitis is usually bilateral (80%),although both eyes may not be affected simultaneously or to same degree.Initial symptoms are pain, tearing, photophobia, and perilimbalinjection. Inflammation lasts for weeks if left untreated. Sectoral superior stromal inflammation and keratic precipitate sare typically seen early. As the disease progresses, deep stromal neovascularisation develops. Eventually inflammation spreads centrally, corneal opacification and oedema develop. The vision may be reduced to the level of light perception only. Late stages show deepghost (non- perfused) stromal vessels and opacities. The constellation of interstitial keratitis, VIII cranial nerve deafness and Hutchinson teeth is called the Hutchinson triad. Sequelae of stromal keratitis include corneal scarring, thinning, ghost vessels in deep layers. Stromal keratitis develops only rarely in acquired syphilis, if it does,is typically unilateral(60%).Ocular findings similar to those seen in congenital syphilitic keratitis. Uveitis and retinitis are much more common manifestations of acquired syphilis than keratitis. .

**INVESTIGATIONS**:

Diagnosis is usually based on history, clinical presentation and supported by serologic testing. T.pallidum cannot be detected by culture. Dark field microscopy and direct immunofluorescentantibody tests used to identify spirochete from lesion exudates or tissue. Serodiagnosis based on non treponemalantigen tests like Venereal Disease Research Laboratory(VDRL) and rapid plasma reagin(RPR)tests and treponemal antigen tests like fluorescent treponemal antibody absorption(FT-ABS)assay and microhemagglutination assay for T.pallidum antibodies(MHA-TP).A lumbar puncture is needed in every case of syphilitic uveitis.Positive CSF VDRLis diagnostic of neurosyphilis.HIV testing is performedin all patients.

**TREATMENT:**

In the acute stages,ocular inflammation is treated with cycloplegic agents and topical corticosteroids to limit stromal inflammation and scarring. Systemic syphilis is treated with penicillin according to the stage of the disease. Late syphilis in the absence of neurosyphilis requires Benzathine penicillin G 1.2-2.4 millionunitsim weekly,total 3 doses.Patients allergic to penicillin treated with Doxycycline 100mg orally bid for 4 weeks or Tetracycline 500mg orallyqid for 4 weeks.

**FOLLOW UP:**

Patients must be monitored for Jarisch-Herxheimer reaction, hypersensitivity response to treponemal antigens released when large number of spirochetes are killed during the first 24 hours of treatment. It presents with constitutional symptoms and sometimes with increased ocular inflammation. Supportive care, observation and rarely corticosteroids are sufficient.

**DIFFERENTIAL DIAGNOSIS:**

IK associated with pulmonary tuberculosis is usually unilateral. Lepromatous interstitial keratitis is usually bilateral, the organisms have been found throughout the stroma, implying that a direct infectious etiology may occur, in contrast to an immunologic etiology. Lyme disease does not commonly feature stromal edema, can appear as large nummular infiltrates in various levels of the stroma without corneal neovascularization. Cogan syndrome is triad of nonsyphilitic interstitial keratitis, vestibuloauditory disease, and associated autoimmune vasculitis; in contrast to the deafness associated with syphilitic interstitial keratitis, the hearing loss in Cogan syndrome also has the vestibular symptoms

**CONCLUSION:**

Interstitial keratitis, though a rare entity, should be the first differential diagnosis in a non-ulcerative stromal inflammation in the younger age group and subsequently screened for syphilitic etiology.

**REFERENCE**:

1.AAO cornea pg207-209,intraocular inflammatory disease

2.CorneaFundamentals,Diagnosis& Management - Krachmer,Mannis& Holland

3.Smolin and Thoft's The Cornea: Scientific foundation and clinical practice

4.Kanski Jack J, Bowling Brad .Clinical Ophthalmology-A Systematic Approach. Ed 7, cornea: Elsevier:2011.

5.Harrison's principles of internal medicine 18th edition, infectious diseases

6.Interstitial Keratitis Overview of Interstitial Keratitis Author: Parag A Majmudar, MD; Chief Editor: Hampton Roy Sr, MD

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities