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Ocular Manifestations Of Cogan Syndrome - A Case Report

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I. Introduction

Cogan's syndrome is a rare autoimmune systemic vasculitis characterized by intraocular inflammation and vestibulo-auditory dysfunction (usually neurosensory deafness, but also tinnitus and vertigo). Cogan's syndrome is a rare disease, which primarily affects young adults; reports that establish the age of onset as ranging from 3 to 50 years have been published [1]. The ocular involvement in Cogan's syndrome is variable; the most common is interstitial keratitis, but it can present itself in other ways too: scleritis, episcleritis, retinal vascular disease, uveitis, iritis, conjunctivitis, papilloedema, exophthalmos, tendonitis [2].Rarely the interstitial keratitis is asymptomatic and it is discovered at an ophthalmological examination of a patient with audiovestibular symptoms [3]. The most common clinical findings are ocular redness (74%), photophobia with tearing (50%), ocular pain (50%) and transitory diminution of visual acuity (42%) [4].

II. Case Report

A 18 year old girl came with history of diminution of vision in her right eye from the last 1 month. On examination, UCVA was RE 5/60 and LE 6/60, anterior segment finding suggestive of R/E 3x3 mm corneal desmetocoele with iris plugging, anterior chamber was irregular with corneal thinning and L/E had signs of interstitial keratitis with stromal edema. Systemic evaluation was suggestive of sensorineural hearing when patient underwent pure tone audiometry. The diagnostic work up in this case also included complete blood picture, chest X-ray, ESR, CRP where everything came out to be within normal limits. Viral markers, VDRL, Mantoux were negative, but the only complaints associated were decreased hearing with vertigo which made us to get patient examined from a ENT surgeon too, which brought us to a diagnosis of cogan syndrome as it is a syndrome of exclusions.

In this case, other signs of scleritis or episcleritis were not seen rather it was a very quiet eye, with mild congestion in left eye. The treatment plan in this case was that the patient was then taken up for corneal patch grafting and was further put on topical steroid 1% Prednisolone 8t/day alongwith topical cyclosporine twice a day. Oral systemic steroids were also added to the dose of 1 mg/kg/day. As cogans syndrome is always a diagnosis of exclusions, the other differentials which were ruled out were Meniere's disease, any cardiovascular, genitourinary tract vasculitis. There was no raised ANA antibody titres or any other associated syphilis.

Rather, in this case decreased hearing was associated an year back before the keratitic picture was set in which eventually led to a perforation in right eye and a corneal patch graft thereof.

III. Discussion

General symptoms can appear such as fever (up to 39 degrees C) and weight loss (up to 10 kg) [**5**]

Grasland et al [**6**] published a study that concluded the disease remains restricted to the ear and eye in 17/ 52 (33%) patients with typical CS and 7/ 59 (12%) patients with atypical CS. General symptoms can appear such as fever (up to 39 degrees C) and weight loss (up to 10 kg)

IV. Conclusion

Cogan's syndrome is a rare presumed autoimmune disorder characterized by non-syphilitic interstitial keratitis and audiovestibular symptoms that resemble Meniere's syndrome, sometimes associated with systemic manifestations, especially cardiac complications.The diagnosis should be suspected whenever there are ocular abnormalities closely followed or preceded by audiovestibular symptoms. No serological marker for the disease has been found, so the diagnosis is an exclusion one. The evolution of Cogan's syndrome is unpredictable. Patients with ear and eye involvement alone have a good prognosis and an average life expectancy. The corneal disease may improve even without therapy, while prognosis for hearing is usually poor on the long-term,

deafness being often irreversible [7]. Patient is doing well at the end of 1 month with quiet corneal patch graft and decresed stromal haze in the other eye with BCVA 5/60 and 6/12 respectively.

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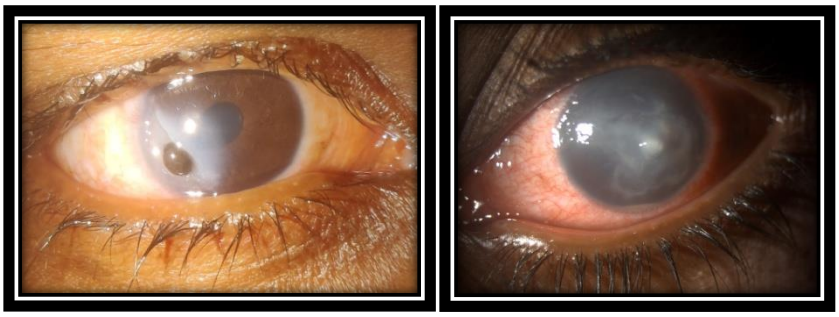

Fig-1: Presentation showing R/E corneal perforation L/E showing stromal keratitis with inflammatory signs

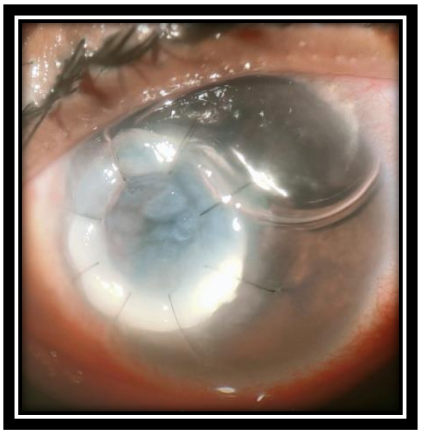

Fig:2- Post op corneal patch graft

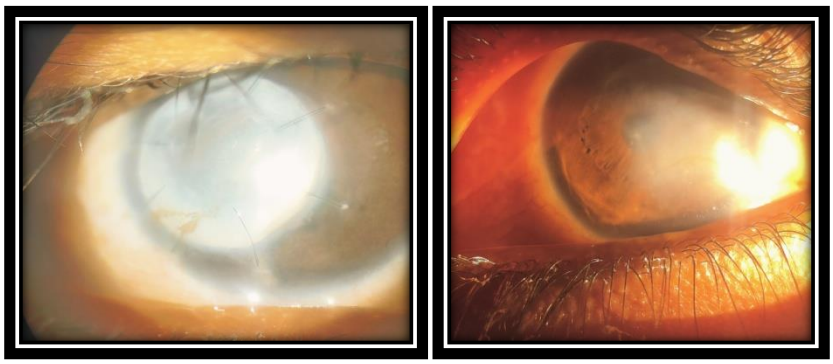

Fig:3- POD-30 days
Stable Corneal Patch graft R/E and L/E Stromal scarring has reduced

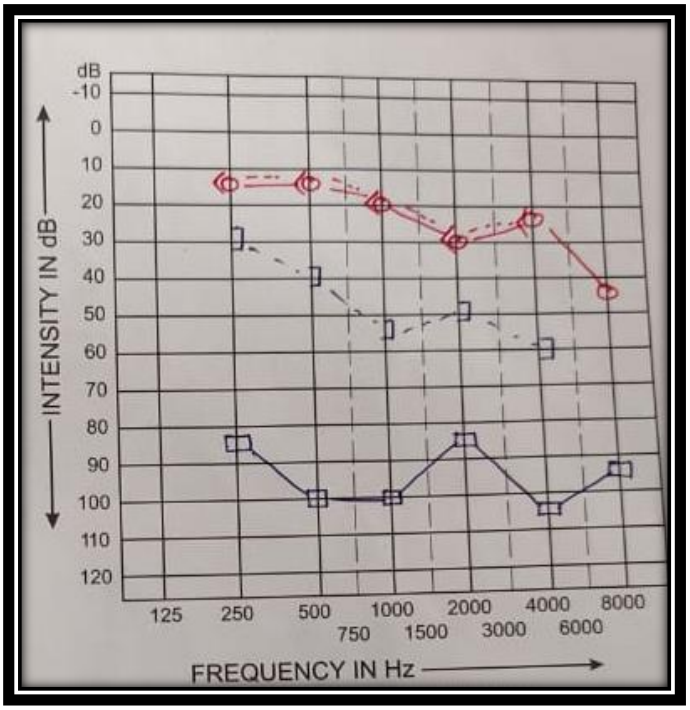

Fig 4: shows Sensorineural Hearing loss, Rt Ear 21.6dB, Lt Ear 95dB S/O Rt Ear Minimal Hearing Loss, Lt Ear Profound hearing Loss