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Coexistant Retinitis pigmentosa(RP) and Vogt Koyanagi Haradas syndrome(VKH)- a case report of management crisis

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Abstract: Abstract The following article is a case report of a patient who presented with two uncommon degenerative conditions together. He was a diagnosed case of retinitis pigmentosa, with all its manifestations but had ambulatory visual potential. When this condition was overwhelmed with another significant vision threatening condition like Vogt Koyanagi Haradas syndrome, treatment had to be early, quick and aggressive to safeguard the macula to preserve central visual acuity as much as possible

Keyword :retinitis pigmentosa- vogt koyanagi harada's syndrome- macula- available vision-management crisis

Introduction :

Retinitis pigmentosa is a devastating hereditary condition causing progressive atrophy of the retinal photoreceptors leading to defective night vision, peripheral visual fields and subsequently central visual acuity ¹². On the other hand Vogt Koyanagi Harada's syndrome is an acquired autoimmune condition affecting the choroidal melanocytes ⁶, although the acute attacks respond well to steroids, it usually has a stormy course with multiple recurrences and chronicity ultimately leading on to retinal degenerative changes. Both these conditions are potentially blinding in their own course and therefore when both of them occured together in the same patient, management had to be very aggressive and was challenging to safeguard the precious available useful vision. case report:

A 32 year old male patient from Kerala, came to us with complaints of sudden onset of redness and pain with associated drop in available vision for last 10 days. He was a diagnosed case of typical retinitis pigmentosa with documented flat (scotopic and photopic) ERG tracings done one month back. On examination his BCVA in right eye was PL+(accurate PR) and in the left eye was HM+ (the visual acuity recorded one month back was about 4/60 in both eyes). Anterior segment examination revealed a picture of bilateral granulomatous anterior uveitis with large pigmented keratic precipitates, anterior chamber reaction of grade-III, koeppe nodules, posterior synechiae and festooned pupil. Lens showed cataractous changes attributing to nuclear

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University University Journal of Surgery and Surgical Specialities sclerosis grade I and posterior subcapsular cataract(fig 1 and 2) Posterior segment evaluation revealed vitritis with grade II vitreous haze and bilateral multiple serous retinal detatchments involving the macula(fig3 and 4). Discs were pale with attenuated vessels . No obvious skin lesions, hearing deficits or neurological abnormalities were noted. A probable diagnosis of VKH¹ was made and the same was confirmed on fundus florescein angiography which revealed early hypoflorescence with multiple leaking hyperflorescence spots in the late phase. These findings were further attributed by the presence of multiple serous detatchments seen in ultrasound Bscan of both eyes with associated serous elevation seen in foveal and parafoveal regions of both eyes by OCT(fig5,6).He was treated with three doses of intravenous Methyl Prednisolone 1g in 100ml

NS for three days³ followed by three shots of periocular(posterior subtenons) steroid injections (0.5 ml of 20mg of Triamcinolone Acetonide) and was started on high dose oral steroids(60 mg of oral Methyl Prednisolone/ day and tapered gradually). The serous detatchments regressed over a period of one week but the visual acuity remained the same. The patient was then started on Azathioprine 50mg BD⁵ and was also maintained on low doses of oral steroids. After three months his visual acuity improved to 4/60 in both the eyes. He is currently on Azathioprine , low dose steroids and close monitoring of his ocular and systemic parameters(especially liver function tests) on a monthly follow up basis.during his last visit, anterior segment of both eyes(fig 7 &8) revealed resolved uveitis fundi(fig 9&10) showed waxy pale discs , attenuated vessels and bony spicules, corroborating well with a prior diagnosis of retinitis pigmentosa, along with sunset glow of resolved VKH

Fig 1 & 2 : slit lamp photos taken during first visit(showing bilateral granulomatous anterior uveitis)







fig2-left eye Fig 3 & 4 : fundus photos showing multiple serous detatchments involving the macula fig 3-Right eye fig4-left eye Fig4 & 5: OCT pictures showing serous elevation in foveal and parafoveal regions





fig5-Right eye fig6-left eye Fig 7 & 8: slit lamp photos during last visit showing resolved anterior uveitis

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fig 7-Right eye



fig 8-left eye Fig 9 & 10 : fundus photos showing taken during the last visit showing resolved vitritis and settled retinal detatchments, revealing pale discs, attenuated vessels and bony spicules corresponding to underlying retinitis pigmentosa



fig 9-Right eye



fig 10-left eye

Discussion:

Retinitis pigmentosa (RP) is a slowly progressive retinal dystrophy progressive damage to rod and causing cone photoreceptors, causing defective dark adaptation, progressing as a defective night vision with constricted visual fields and finally affecting the central vision by involving the macula. In many cases of retinitis pigmentosa there is relative sparing of posterior pole till the end stages of the disease and the patient maintains reasonably good central vision inspite of significant field constiction. This emphasises the importance of the macular sparing effect in a patient with RP.VKH or Vogt Koyanagi Harada's syndrome is an autoimmune disorder affecting the melanocyte rich tissues in the body, in the ocular system the uveal melanocytes⁹, presenting with recurrent attacks of bilateral granulomatous anterior uveitis with multiple serous detatchments usually involving the macula .VKH is also known to have a stormy course with multiple recurrences and chronicity, ultimately leading to irreversible degenerative changes in the fundus.

Therefore, in such a situation when two vision threatening conditions prersented toghether in the same patient, it was extremely important to reattatch the retina at the macula, to restore the only available central vision. Thus this case proved to be a classic example of management crisis where treatment had to be both aggressive and quick knowing well that the results are not rewarding. *Improvement in two to three metres from just perception of light made a lot of difference to a patient whose peripheral, colour vision and dark adaptation were already compromised.*

This emphasises the importance of aggressive systemic steroid administration and immunosuppressant usage ^{3,2,5} in such cases.In our patient though the retinal detatchments settled in response to IV Methyl Prednisolone, the visual recovery was slow. This necessitated maintainance therapy with either high dose steroids or additional immunosuppressants. As apposed to side effects of long term high dose steroids, Azathioprine was found to safe and effective and was started. At present both eyes are quiet revealing signs of resolved disease. Such patients would certainly require long termimmunosuppression and thereby the risks are to be weighed carefully against the benefits to achieve reasonable results. The need to restore the macular functions as early as possible, (for the reasons stated above) further justified early immunosuppression in our patient.

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