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RESEARCH ARTICLE

PROBABLE VOGT-KOYANAGI-HARADA DISEASE: A CASE REPORT

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Abstract

Vogt-Koyanagi-Harada(VKH)diseaseisabilateralgranulomatouspanuveitis affecting young adults, withor without systemic manifestations. The prognosis of VKH disease is generally favorable, however the development of complicationscanleadto blindnessanddeafness.(1)
In this case, we report a 22-year-old man who developed probable Vogt–Koyanagi–Haradadisease without any systemic symptoms, but with several risk factors for poor visual prognosisbeforetreatment,andwhowassuccessfullytreatedwithintensivecorticosteroidtherapy.
Our aim is to describe a clinical case with bilateral serous retinal detachment that presumed asprobableVKHdisease.

Introduction:-

Vogt-Koyanagi-Harada (VKH) syndrome is a multisystemic granulomatous autoimmunedisease affecting organs with high melanocyte concentrations, including the eye,the central nervous system, auditory and integumentary systems, it is characterized by ocular, neurologic, and dermatologic signs.(2)

Theinternationalrevised criteriaofVKHdevise the disease to completeVKH,incompleteVKH, andprobableVKH. (3)

Casereport:
The case of a 22-year-old patient without any pathological or traumatic history. He presented himself at the ophthalmological department for a decrease in visual acuity in both eyes evolving for one week, without anyextraocular signs.

In the ophthalmologic evaluation his best corrected visual acuity was 1/10 in both eyes. The examination under a slit lamp revealed a quiet anterior segment in both eyes, without keraticprecipitatesorflareintheanteriorchamber. Theintraocularpressurewasnormal.

The fundus examination after dilatation revealed a macular serous detachment in both eyes, without inflammation signs in the posteriorpole (figure1).

ENTexamination,dermatologicalandneurologicalexaminationwerenormal.

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Afluoresceinangiographyrevealedserousdetachmentswithlatepoolingofdye,multifocal areas of pinpoint leakage, andleakageof dye in thesubretinal space(figure1).

Optical coherence tomography (OCT) confirmed marked serous retinal detachment at the macular area in botheyes.

Markers of autoimmunity and inflammation were negative as were the serological testing which eliminate others infectious diseases.

ourpatienthad no neurologicalorauditorysigns,hewasdiagnosedwith probableVKHdisease based onthe international criteria.(4)

Treatmentwasstartedwithbolusesofmethylprednisolone(1 g/dayfor4days),followedbyaschemeof prednisolone(60 mg/day)withprogressive decrease.

After12monthsoftreatment, the visual evolution was favorable andthebestcorrectedvisualacuitykepton9/10withcompletereabsorptionofsubretinalfluid,normalretinalthicknessesconfirmedbyOCT(figure3).

Discusion:-

Vogt-Koyanagi-Haradaisasystemicautoimmunediseasecharacterizedbythepresence of ocular signs, associated with extraocularchanges; neurologic, auditoryand integumentary, without ocular trauma or surgery.(2) (3)

The American Uveitis Society diagnostic criteria for VKH disease include no history ofocular trauma or surgery, and at least one finding in three, out of the following fourcategories:1)bilateralchroniciridocyclitis;2)posterioruveitis,includingexudativeretinaldetachment, disk hyperemia or oedema and sunset glow fundus; 3) neurologic signs: tinnitus,meningismus;4) cutaneousfindings ofalopecia,poliosis and or /vitiligo.(4)

The First International Workshop on VKH disease had revised the diagnostic criteria asfollows; complete VKH is defined as bilateral involvement with neurologic or auditory findingsand integumentary findings; incomplete VKH is defined as bilateral involvement with neurologicor auditory findings or integumentary findings; and probable VKH is defined as bilateral ocularinvolvementonly, like in our case. (2)

VKH disease can lead to significant visual loss, but the right diagnosis and the treatment can minimize ocular morbidity. (5) The treatment is based on early start of systemic corticosteroids at high doses ranging between 200 mg/day and one gram/dayfor 3–5 days, followed by a progressive decrease for 4 to 9 months, to suppress the acute choroidal inflammation and avoid recurrence. while other immunosuppressive and cytotoxic agents are reserved for resistant cases. (6

There are recent studies which support the evidence that first-line use of corticosteroid combined with immunosuppressive agents decreases the development of late complications and recurrence of the disease, improves long-term vision and facilitates more rapid tapering of steroids. Cyclosporine seems to be a better immunosuppressive agent than azathioprine, with good efficacy and safety. (7) (8)

Our patient received systemic corticosteroid therapy without immunosuppressive agents. No resistance or recurrence was noticed, and one year after the initial clinical presentation he wasasymptomatic.

Our study presents a rare clinical variant of the VKH disease with complete recovery after treatment and enhances the fact that probable VKH disease is rare but should be promptly diagnosed and treated. (8) (9)

Conclusion:-

VKH is a multisystem pathological disorder. The diagnosis is mainly clinical. The mainstay of treatment isaggressive corticosteroid therapy in the acute phase and eventually an additional immunosuppressive agent.Thereisapotentialforsignificantvisualloss,buttimelyidentificationandtreatmentcanminimizeocularmorbidity.

Figure:-

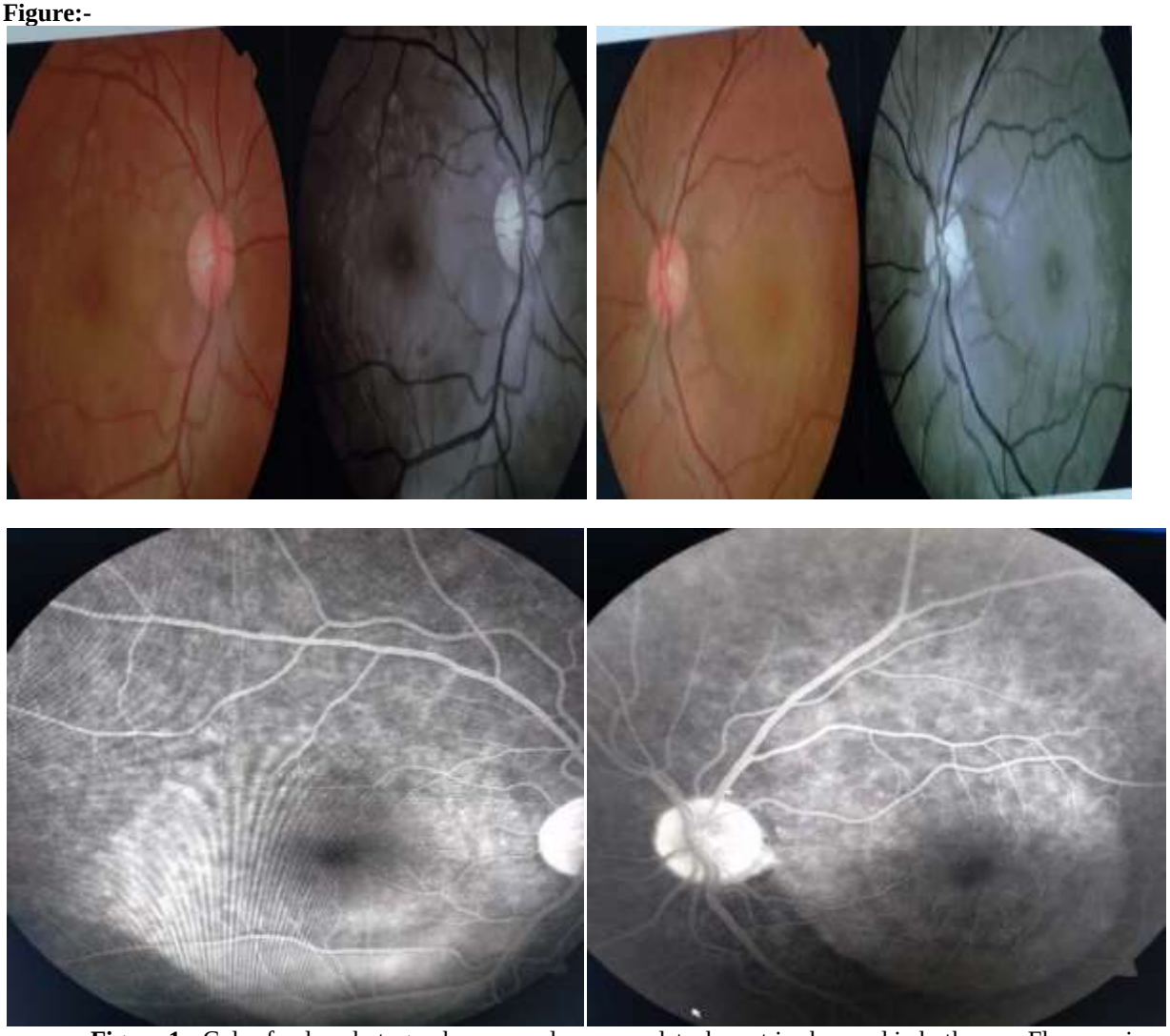

Figure 1:- Color fundus photographs: a macular serous detachment is observed in both eyes. Fluorescein angiographyshowmultiplehyperfluorescentdefectsduetodyeleakage,suggestive of serous retinal detachments, punctiform hyperfluorescent defects are also shown inthemaculararea.

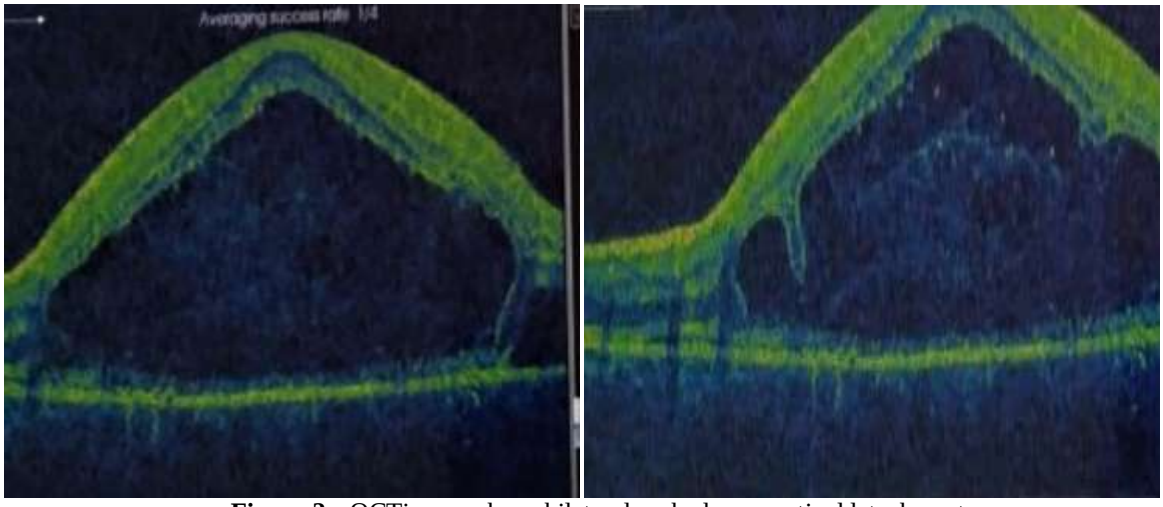

Figure 2:- OCTimagesshowabilateralmarkedserousretinaldetachment.

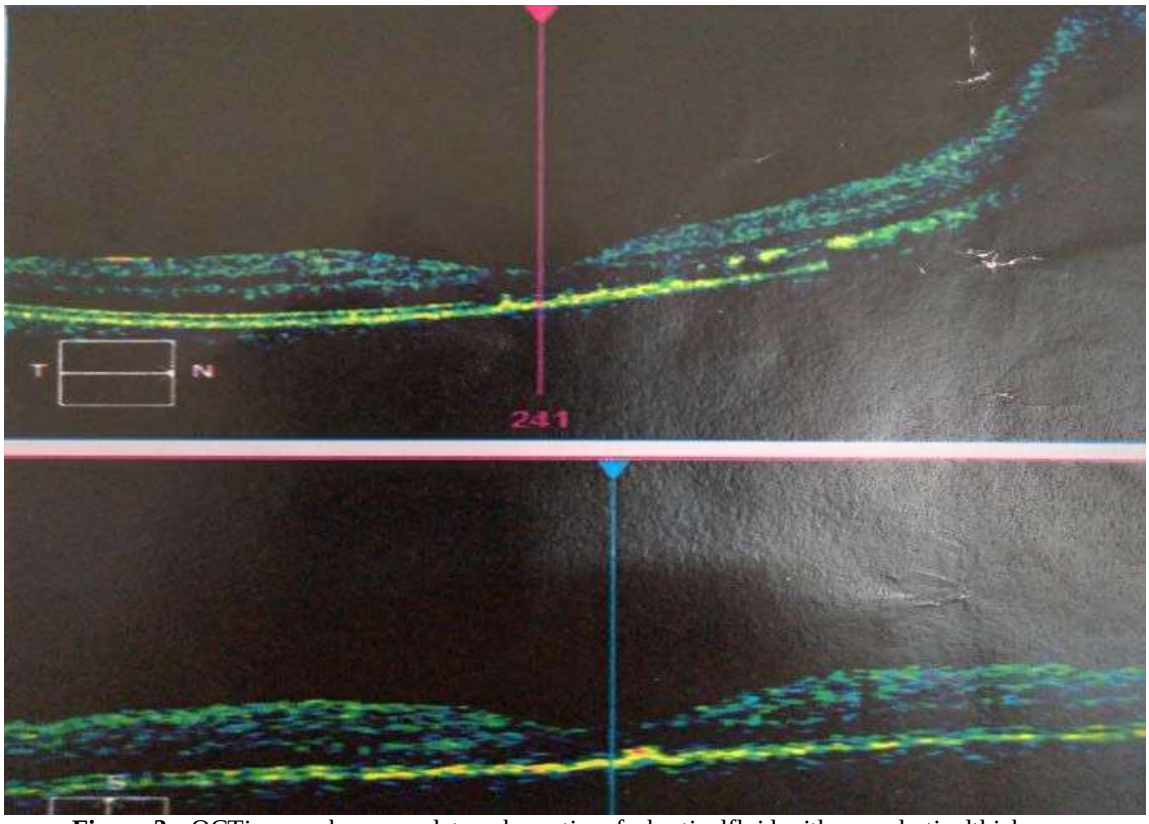

Figure 3:- OCTimagesshowcompletereabsorptionofsubretinalfluid,withnormalretinalthickness.

Competinginterests
Authorshavedeclaredthatnocompetinginterestsexist.

Authors'contributions
Allauthorsreadandapprovedthefinalmanuscript.

Consent (Where Everapplicable)
Thepatienthasgivenitsinformedconsentforthecasereporttobepublished.

Abreviations:-

1. VKH:Vogt-Koyanagi-Harada
2. ENT: Ear, nose and throat examination.

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