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Radiodiagnosis

CASE REPORT ON VOGT KOYANAGI HARADA SYNDROME

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ABSTRACT Vogt-Koyanagi-Harada syndrome (VKHS), initially described as an uveo-meningo-encephalitic syndrome, is a rare systemic granulomatous autoimmune disease that targets melanocyte-rich tissues, such as the eye, inner ear, meninges, skin and hair.[1] VKHD is often associated with neurologic and cutaneous manifestations, including headache, hearing loss, vitiligo and poliosis. This disease is mainly a Th1 lymphocyte mediated aggression to melanocytes after a viral trigger in the presence of HLA-DRB1*0405 allele [2]. The absence of ocular trauma or previous intraocular surgery sets VKHD apart from sympathetic ophthalmic, its main differential diagnosis. Clinical diagnosis done by using revised diagnostic criteria Vogt-Koyanagi-Harada syndrome proposed by international nomenclature committee. Here presenting a case report of VKHS.
In our patient all 1-5 criteria present and confirmed by imaging.

KEYWORDS : Vogt Koyanagi Harada Syndrome

CASE REPORT:

HISTORY:-

27 year old female came with complains of loss vision in both eyes with history of fever, headache and diminished vision since 7 year and no history of trauma. On general examination vitiligo spot present over back of chest. Routine blood examination are normal.

On ophthalmic examination reveals no perception of light in both eyes. Bilateral eye lagopthalmos, clear conjunctiva, keratopathy, shallow anterior chamber and exudative membrane over pupils in both the eyes.

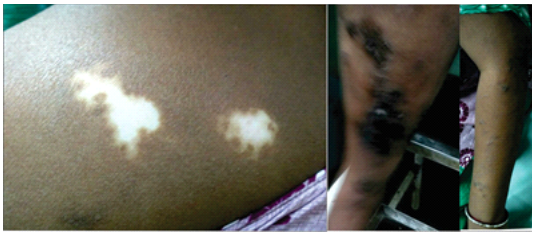

FIG.1 shows vitiligo spot at back, Cutaneous Findings

IMAGING FINDING

On USG bilateral small eye, retinal detachment, calcified lens seen. There is diffuse thickening of choroid s/o diffuse choroiditis. On CT scan diffuse thickening of bilateral eye wall, calcified lens and bilateral small eye wall. On MRI there is diffuse thickening of choroid of both eye wall which appear hyper intense on T2 and showing diffuse enhancement on post contrast scan.

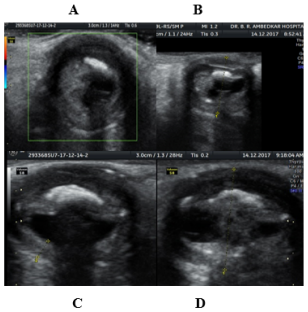

FIG.2 :-A,B) right eye B-scan shows reduced axial length , thickened choroid and retinal detachment with calcified lens. C,D) left eye B-scan shows reduced axial length , thickened choroid and retinal detachment with calcified lens

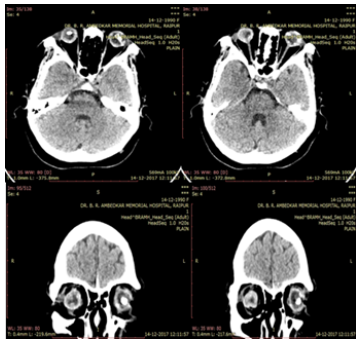

FIG .3:- A,B,C,D) axial and coronal NCCT orbit shows diffuse thickening of choroid of both eye.bilateral eye appear small in size with bilateral calcified lens and foci of calcification noted in left eye wall.

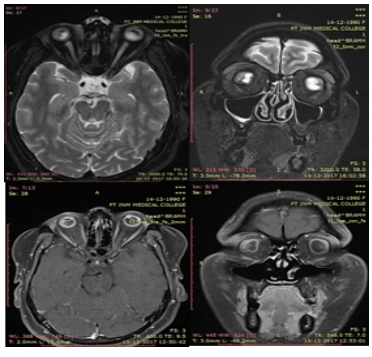

FIG.4 A,B) T2W FS Axial and coronal image shows diffuse thickening of bilateral choroid with small eye wall. C, D) T1W+C axial and coronal image shows diffuse enhancement of choroid of both eye.

DISCUSSION

Vogt-Koyanagi-Harada syndrome is a bilateral, chronic, diffuse granulomatous pan-uveitis frequently associated with neurological, auditory, and integumentary manifestations more commonly involve individuals of pigmented skin, such as Asians and Native Americans. It is very infrequent among persons of African descent[3].

The pathophysiology of this disorder has not clear but is thought to be related to T-lymphocyte-mediated autoimmunity against melanocyte

tyrosinase–related proteins, these are found in uvea, retina, and the leptomeninges[4]. A genetic basis is likely the predisposing factor because there is an increased incidence with darker pigmented skin; however, a particular inheritance pattern has yet to be identified.

VKHS is divided into four stages: prodromic, acute uveitic, convalescent and chronic or recurrent [1]. To "stage" the disease may enable exchange information rapidly between care-givers as to chronology of the disease which implies in treatment strategies.

There has yet to be a consensus regarding a definitive diagnostic test for VKH syndrome, which consequently limits the diagnosis to a combination of clinical and ancillary test findings. Thus, there are 3 distinct categories, classified as complete, incomplete, and probable VKH syndrome based on the following criteria: 1) uveitis without a history of ocular trauma or surgery, 2) uveitis without clinical or laboratory evidence of other ocular disease, 3) bilaterality of the uveitis with retinal detachment, 4) the presence of auditory or other neurologic (nonvisual) findings, and 5) integumentary (skin) findings[5]. Complete VKH syndrome manifests all 5 criteria, whereas incomplete VKH syndrome manifests criteria 1–3 along with either 4 or 5. Probable VKH syndrome (as in our patient) represents isolated ocular disease clinically, with criteria 1–3. Thus, the extraocular clinical manifestations of complete VKH syndrome may not be fulfilled until months or years following the initial presentation of ocular disease[3].

B-mode ocular ultrasound features of Vogt-Koyanagi-Harada syndrome are: low to medium reflective thickening of the posterior choroid, serous retinal detachments, mild thickening of the sclera and/or episclera adjacent to areas of choroidal thickening, vitreous opacities and sub-retinal septations may be seen[6]

MR imaging may detect early CNS involvement by Vogt-Koyanagi-Harada syndrome before the onset of neurologic symptoms which include: typical bilaterality of ocular findings, scattered periventricular white matter lesions on T2-weighted imaging/FLAIR, and pachymeningeal enhancement. Bilateral contrast enhancement of the choroid is seen along diffuse choroidal thickening with scleral sparing and retinal detachment [6]. Complications include glaucoma, subretinal fibrosis, choroidal neovascularization and cataract.

Treatment of iridocyclitis should be performed according to the intensity of anterior segment inflammation. Topical corticosteroids (e.g., dexamethasone 0.1 % or prednisolone acetate 1 % eyedrops) in combination with mydriatics/cycloplegics (e.g., tropicamide 1.0 % eyedrops) to reduce ciliary spasm and prevent posterior synechiae, are most frequently used [1].

CONCLUSION:-

Diagnosis of rare VKH syndrome can be made by high suspicion on clinical finding and should be confirmed by imaging.

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