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RESEARCH ARTICLE

VOGT-KOYANAGI-HARADA SYNDROME: CASE REPORT AND LITERATURE REVIEW.

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Abstract

Introduction: Vogt-Koyanagi-Harada Syndrome is a cell-mediated autoimmune disease directed against melanocytes Leading to chronic multisystemic disease affecting the eye, inner ear, skin and meninges. VKH is a common disease in Kingdom of Saudi Arabia, accounting for 2.5–19.4% of all uveitis cases.

Case report: A 24 years old male patient presented to our ophthalmology clinic with bilateral painless loss of vision for 3 days. With signs of bilateral conjunctival hyperemia, anisocoria, anterior chamber reaction, multiple focal exudative retinal detachments bilaterally associated with fundus edema.

Discussion: VKH is a syndrome diagnosed clinically. It is unlikely presents with typical finding. Laboratory evidence must be involved to identify the disease and it's possible causes.

Introduction:-

Vogt-Koyanagi-Harada (VKH) Syndrome, also called uveomeningioencephalitis, is a chronic multisystemic disease affects the meninges, eye, skin and inner ear .it's considered to be a cell-mediated autoimmune disease that directed against melanocytes. It's characterized by serous retinal detachments, panuveitis with iridocyclitis, diffuse choroidal swelling and optic disc hyperemia. [(1)] It may be associated with neurologic and skin manifestations. Typically the findings are mostly bilateral, with possible asymmetric severity.

VKH disease has a range of onset from 3-89 years, reaching its maximum in the third decades. Females are more affected than males; the female-to-male ratio mostly is 2:1 [(2)] . VKH disease is not common, but it may be seen in Asian, Middle Eastern, Hispanic, and Native American populations [(3)]. And represents 7-8% of all patients with uveitis [(4)] .

VKH appears to be common in the kingdom of Saudi Arabia, accounting for 2.5–19.4% of all uveitis cases in various studies. It's mostly affect young females. Usually Bilateral panuveitis is the commonest eye manifestation and near 50% of eyes present with exudative retinal detachment [(5)].

Case report:-

A 24 years old male patient came to our ophthalmology clinic with bilateral painless loss of vision for 3 days associated with frontal headache for 1 week. There was no history of vertigo, tinnitus, alopecia, neck stiffness, penetrating ocular surgery or trauma. He had no previous similar attacks. On examination there was bilateral conjunctival hyperemia, anisocoria, anterior chamber reaction, multiple focal exudative retinal detachments with

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associated evident fundus edema (**Figure-1A**). The best corrected visual acuity (BCVA) was 20/40 for the left eye (OS) and 20/20 for the right eye (OD).

Slit-lamp examination (SLE) showed +1 aqueous flare in both eyes. There were no vitreous cells and the intraocular pressure (IOP) was 16OS, 18OD. Optical coherence tomography (OCT) report revealed sub-retinal fluid accumulation in both eyes (OU) (**Figure 1B**).

Pupillary function and biomicroscopic examination were normal. Autoimmune and inflammatory diseases markers were negative and the serology test for the infectious pathologies diagnosis were negative. systemic review was normal. A series of blood tests (e.g. CBC, ACEI, ANA, enzyme-linked immunosorbent assay) were done to rule out other causes of uveitis.

The patient was referred to the rheumatology clinic, There were no rheumatological manifestations pointing toward a specific systemic rheumatic disease.

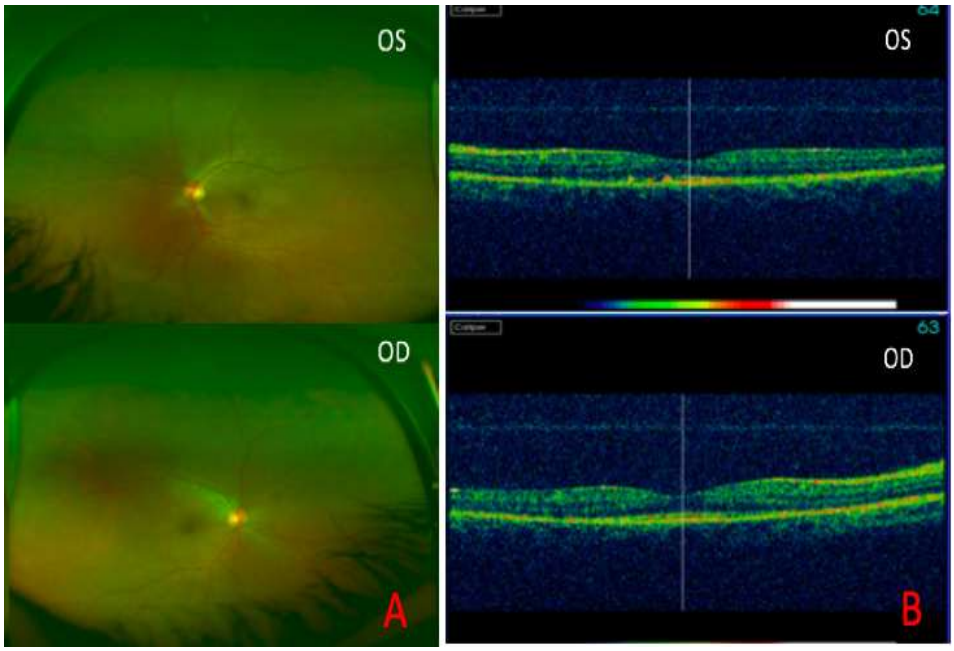

Figure 1-A: *Fundus examination, B: OCT*

The patient was diagnosed with VKH disease, he received oral prednisolone 50 mg OD tapering 5 mg every 5 days with follow up, and Mycophenolate mofetil 500mg BID for 1 week then 1g BID. The patient had adverse reactions manifested by abdominal pain and palpitation, so Mycophenolate mofetil was withdrawn and changed to Azathioprine 50mg OD for 7 days then100 mg OD. Omeprazole 20 mg OD and Alendronate 70 mg once weekly. Calcium 500mg BID and Vitamin D 1000 IU OD as osteoporosis prophylaxis.

Eight weeks after the first presentation, the BCVA of his left eye increased to 20/20 and retinal thickness of 191 µm, with no posterior choroidal thickening

Twelve months after the first clinical presentation, the BCVA of both eyes continued to be 20/20 with complete subretinal fluid reabsorption, normal retinal and choroidal thicknesses confirmed by OCT characteristic of VKH. Normal Fundus appearance in both eyes.

The patient transferred to Jeddah for follow up to be closer to his work. In Jeddah, he was found to have VA 6/6 OU, IOP 15 OD ,16 OS, SLE was found to be normal (Figure2). management was continued to be the same.

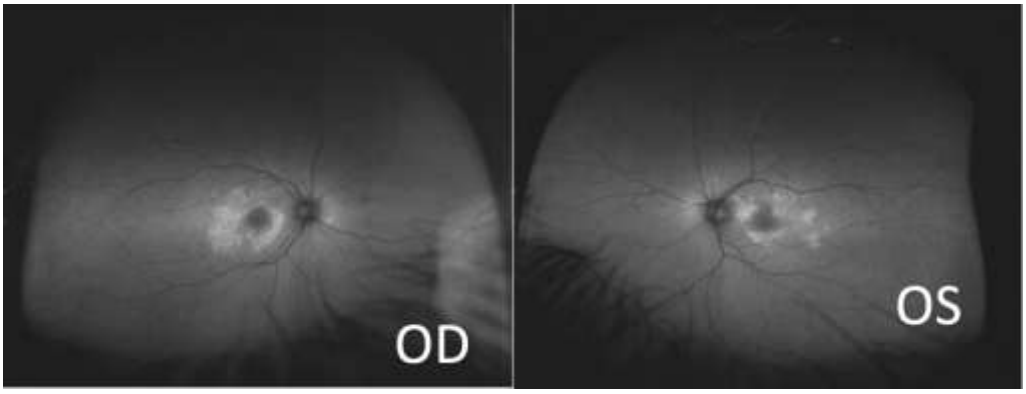

Figure 2:- Fundus exam

Discussion:-

Vogt-Koyanagi-Harada (VKH) disease is a bilateral chronic multisystemic inflammatory disorder, predominantly affecting the eye in conjunction with neurological and dermatological manifestations.[1] it's diagnosed clinically. it is uncommon to have typical findings. The International Committee on Nomenclature published revised criteria for the diagnosis of VKH disorder, [6] usually it is classified as: complete, incomplete, and probable. Ancillary tests, e.g. ultrasonography, fundal fluorescein angiography may be helpful. Patients with VKH present in stages [1], either as acute (neurological followed by ocular and dermatological manifestation), or as convalescent (for those who are treated) or as chronic recurrent disease with ocular complications.

Its pathogenesis is unknown, but it showed a strong association with autoimmune, and infectious basis. [7,8] There is a high incidence among those with HLA-DR4, and other human leukocyte antigens. [9] Several studies described its occurrence following a closed head trauma, following surgery for metastatic malignant melanoma, and after cutaneous injury. [10,11,12] Specific racial and ethnic groups are linked with incidence of VKH. It is seen among those races with dark pigmented skin, like Asians, Hispanic, Middle Eastern, and Native Americans. [1,3] It accounts for 7-8% of cases of uveitis in Japan, while 1-4% of cases in USA [4].

Although there are few studies about VKH in Saudi Arabia found in the literature compared to other regions, VKH is considered a common disease accounting for 2.5–19.4% of all cases of uveitis in various studies, [13,14,15,16] where it is predominantly found in young females.[5] This proves a female predilection matching most studies worldwide, which confirms that gender is an important factor in the differential diagnosis of uveitis. Al Mezaine et.al[13] found VKH is the most common cause of uveitis among their female patients. In another study,[14] Nizamuddin et.al observed that it is the second cause. Hamade et.al. also had the same result.[15] Similarly, Islam et.al had VKH as the sixth cause.[16] While VKH was at the bottom of the list in Finland and Boston.[17,18] VKH is the most common cause of panuveitis according to Nizamuddin et.al.[14]

Coming to presentation, Bawazeer et.al[19] found most of the cases had incomplete disease (56.2%), while probable disease was the least (15.6%). They also detected that extra-ocular symptoms were quite common, where neurological symptoms was the commonest (59%), and then cutaneous symptoms (53%). Furthermore, they found that patients in the acute stage came with panuveitis and bullous retinal detachment, while the majority of those in the convalescent stage came with sunset glow fundus and choreo-retinal scars.

Currently, treatment of VKH consists of systemic corticosteroids for acute disease, and immunosuppressive drugs for chronic and recurrent cases. [1] Prognosis is generally favorable among many cases found in the literature, depending on age at onset, complications, recurrence, time to initiate treatment, and visual acuity at presentation, where better VA at the presentation most likely will improve. [6] Most common post-treatment complications recorded were cataract and glaucoma in most studies.

Conclusions:-

VKH syndrome is a common disease in the kingdom of Saudi Arabia. It is the most common cause of uveitis among their female patients. it's diagnosed clinically. it is uncommon to have typical findings. early diagnosis and treatment is strongly required.

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