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Brain Imaging in the Vogt-Koyanagi-Harada Disease: A Case Report

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Abstract **Case Report**

Vogt-Koyanagi-Harada disease is an uncommon multisystem inflammatory disorder characterized by panuveitis with serous retinal detachment and is often associated with neurologic and cutaneous manifestations including headache, hearing loss, vitiligo, and poliosis. While ocular and meningeal signs are typically observed in the acute stage, the associated classic tegumentary findings are observed subsequently. This case report a 66 year female patient with a history of VKHD confirmed, present a sever decreased visual acuity associated with progressive, the brain IRM prsente a leptomeningeal enhancement, diffuse white matter abnormalities and labyrinth enhancement. The role of brain MRI in early disease detection, which allows for prompt treatment initiation and better disease outcome.

Keywords: vogt-koyanagi-harada disease, leptomeningeal enhancement, IRM.

INTRODUCTION

Vogt-Koyanagi-Harada disease (VKHD) is a rare autoimmune disease that targets the ocular melanocyte-rich tissues inducing granulomatous panuveitis and diffuse T-cell lymphocytic infiltration of the choroid. Additional sites of disease involvement include the inner ears, meninges, hairs, and skin [1, 2]. VKHD typically manifests during the third or fourth decade of life and is more prevalent in certain ethnic groups with greater skin pigmentation, such as Native Americans, Hispanics, and Middle Easterns [3].

It is important to differentiate VKHD from other disease entities that manifest as uveitis such as sarcoidosis, tuberculosis, and primary intraocular B-cell lymphoma. Sympathetic ophthalmia and bilateral posterior scleritis due to rheumatoid disease are also part of the differential diagnosis [2]. Although the diagnosis VKHD relies heavily on ophthalmologic examination and investigations, the classic associated brain MRI findings can serve as a useful adjunct tool to confirm the diagnosis and exclude other differential diagnoses. This case report serves to familiarize the radiologists with the classic imaging findings associated with VKHD depicted on contrast-enhanced brain MRI.

CLINICAL CASE

This is the case of a 66-year-old woman with to our emergency department with low-grade fever associated with progressive headache, dizziness, blurry vision, and sever decreased visual acuity (3/10 both eyes), ophthalmologic examination revealed ocular congestion in both eyes and signs of bilateral anterior and posterior uveitis associated with an inflammatory retinal detachment.

The fundus exam revealed an inferior retinal detachment in both eyes predominant in the right eye (Figure 1).

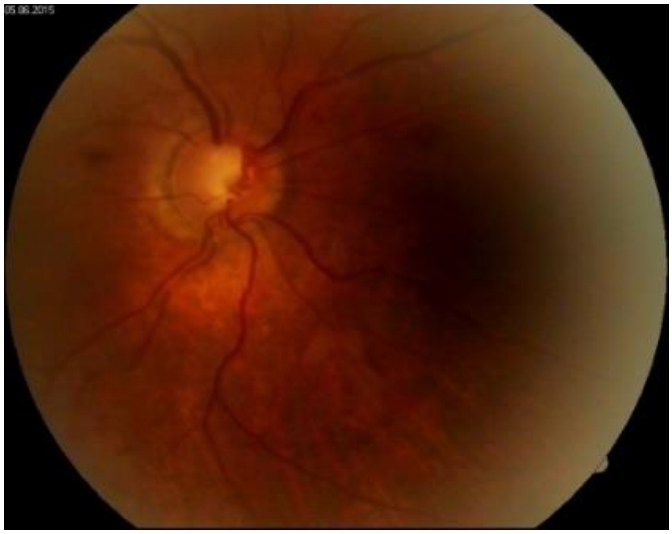

Figure 1: Right eye fundus

The paraclinical examinations revealed:

- Laboratory examinations: high inflammatory markers (CRP, fibrinogen), high glycemia, anemia (HB=10)
- Rheumatological examination showed the presence of vasculitis, arthritis.
- ENT examination revealed a bilateral neurosensitive hypoacusis.
- Dermatological examination revealed alopecia and no signs of vitiligo or poliosis
- MRI examination revealed a optical nerve thickening, discrete leptomeningeal enhancement, while brain findings included diffuse white matter abnormalities T2/FLAIR on periventricular, bilateral semi oval centers and labyrinth enhancement.

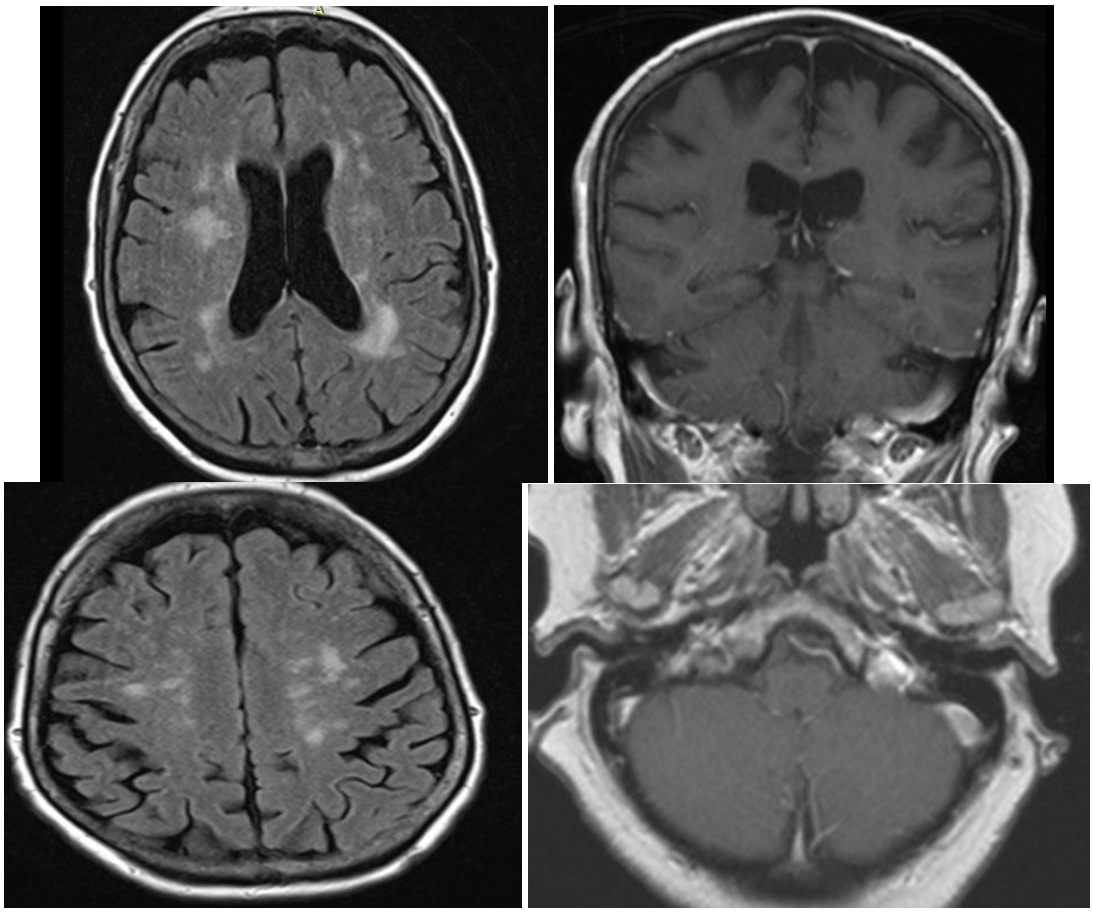

Figure 2: Brain IRM finding

DISCUSSION

Given the rarity of VKHD, its diagnostic criteria continue to change to reflect our evolving understanding of this entity. In 2001, the International Committee on Nomenclature proposed classifying VKHD into three categories based on the extent of extra-ocular findings: complete, incomplete, and probable [4]. Complete VKHD requires the presence of all five criteria: (1) absent history of penetrating ocular trauma or surgery, (2) no clinical or laboratory evidence suggestive of other ocular disease entities, (3) bilateral ocular involvement, (4) neurological or auditory signs, and (5) integumentary findings. On the other hand, probable VHKD is considered when only the first three criteria are fulfilled.

Ophthalmologic evaluation in the form of fundoscopy, fluorescein angiography, OCT, or ultrasound is the cornerstone to detect VHKD. However, MRI is useful not only because of its ability to detect choroidal lesion, but also can also show labyrinthine, meningeal, and brain parenchymal abnormalities. Reported brain imaging findings associated with VKHD include scattered periventricular white matter lesions as well as brain stem and peduncular lesions.

Pachymeningeal enhancement is more common in the context of VKHD; however, leptomeningeal enhancement is also encountered [1, 5]. In addition, MRI can show the abnormal enhancement of the membranous labyrinth bilaterally. Choroid involvement of VKHD typically affects both globes, however, near a quarter of patients starting with unilateral involvement proceeding to bilateral involvement within 15 days [6, 7]. Isolated posterior pole choroidal thickening is observed in about 78.6% of cases, whereas diffuse thickening with posterior pole predominance is observed in about 21.4% of patients. Therefore, uveitis favoring the anterior pole should alert the radiologist to consider a different pathologic entity [1].

Immunosuppressive therapy for six to nine months is the mainstay of VKHD treatment to prevent disease relapse. High-dose steroids are primarily used, while other immunosuppressive and cytotoxic agents are reserved for resistant cases [2].

CONCLUSION

Brain MRI is a useful adjunct tool to diagnose VKHD. Distinct enhancement patterns on brain MRI reflect the inflammatory changes affecting the choroids, cochlea, and meninges.

Pachymeningeal pattern of enhancement is more common in cases of VKHD; however, leptomeningeal and labyrinthine enhancement also occurs as depicted in our case.

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