Unusual association of diseases/symptoms

Is there an association between Vogt–Koyonagi–Harada syndrome and intracranial meningioma?

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The authors report a case of Vogt–Koyonagi–Harada syndrome associated with intracranial meningioma. The authors discuss the HLA-DR4 gene as a common and causative factor in both conditions. This case report draws attention to the possibility of a causative association between intracranial meningioma and Vogt–Koyonagi–Harada syndrome, a rare but potentially blinding condition if not detected and treated early.

BACKGROUND

Vogt–Koyonagi–Harada syndrome (VKH) is a rare, multi-system disorder that typically presents with ocular signs (reduced vision, bilateral panuveitis and exudative retinal detachment), and signs of meningeal involvement (headache, confusion, dizziness, meningism, dysacusia), with or without integumentary signs (vitiligo, poliosis, alopecia).^{1 2} The diagnosis is clinical. It is more common in females and usually presents in the 2nd to 5th decade. Prompt diagnosis and systemic immunosuppression may prevent severe visual loss. It is important for ophthalmologists, physicians and neurosurgeons to be aware of the possible association between intracranial meningioma and VKH syndrome, a rare but potentially blinding condition if not detected and treated early.

CASE PRESENTATION

A 63-year-old Caucasian female was admitted to the medical assessment unit with bilateral visual loss noted 4 days before presentation. She complained of occasional headache. Three months before presentation, she had undergone craniotomy and excision of a right sphenoid wing meningioma grade II. This tumour was discovered on radiological investigation (figure 1) of her deteriorating mental state (anxiety, depression and confusion) of 3 months duration. She had no history of ocular trauma or surgery and no significant medical history.

The best corrected visual acuity was hand movements in both eyes with poor light projection. There was bilateral anterior and posterior segment inflammation, and exudative retinal detachments with subretinal fibrosis confined posterior to the equator, (figure 2). Her blood pressure was normal and there was no vitiligo or alopecia, or focal neurological deficit. Ocular B scan excluded intraocular tumour and scleritis.

INVESTIGATIONS

Haematologic investigations including full blood count, antinuclear antibody, antineutrophil cytoplasmic antibodies, ACE, erythrocyte sedimentation rate, C reactive protein, venereal disease research laboratory, human leucocyte

Figure 1 CT scan of the head showing right sphenoid wing meningioma.

antigen, treponema and toxoplasma antibodies were normal. Chest radiograph was normal. CT scan of the brain found no new pathology. Fluorescein angiography was not done as the patient was not physically able to undergo this investigation.

DIFFERENTIAL DIAGNOSIS

In a patient with bilateral exudative retinal detachment, one has to consider uveal effusion syndrome, choroidal tumours (unlikely to be bilateral), inflammatory causes such as posterior scleritis and vasculitis, and infective causes such as syphilis and toxoplasma.

TREATMENT

A clinical diagnosis of VKH was made. She was treated with intravenous methylprednisolone for 3 days, followed

Figure 2 Colour fundal photographs showing right and left exudative retinal detachment.

by a tapering dose of oral prednisolone, topical dexamethasone and cyclopentolate for 3 months.

OUTCOME AND FOLLOW-UP

At 2 weeks, three right tractional retinal breaks were noted and treated with vitrectomy, cryotherapy and silicone oil. After 6 weeks of treatment the visual acuities had shown no improvement, and the right exudative retinal detachment had recurred.

DISCUSSION

VKH is a multi-system disorder, believed to be caused by an autoimmune disorder of T cells affecting melanocytes. It is most frequent in individuals with a genetic predilection (HLA-DR4 or DR1).³ Prompt diagnosis and systemic immunosuppression may prevent severe visual loss. Due to the late presentation and the severity of the disease, this patient's visual acuities did not improve despite treatment. The presence of subretinal fibrosis suggests that she had developed reduced vision and exudative retinal detachments months before presenting to ophthalmology. The patient may not have complained of reduced vision in the presence of her altered mental state, or this complaint may have been missed.

To our knowledge, this is the second case report associating VKH and meningioma. Kumar *et al*⁴ described a female patient with Harada disease and intracranial meningioma. The authors noted an increased incidence of meningioma in patients with abnormalities on chromosomes 6q, 1p and 22.8, and that the gene for HLA DR4 was also located on chromosome 6q. Studies have shown that meningioma is associated with HLA DRB1 04,^{5 6} the same gene associated with VKH.⁷ Our patient was positive for HLA DRB1 and DRB4, DR53, DQ5 and DQ7. Although this could be purely a chance finding, it is quite interesting that the HLA DR4 gene is responsible for both meningioma and VKH.

This case of VKH with intracranial meningioma should raise an awareness of the possibility of an association between the two. Patients with intracranial meningioma

and altered mental state should be carefully assessed for alterations in vision. Perhaps patients with intracranial meningioma may also need HLA testing. Further studies are needed to determine if there is a definite association between the two conditions.

Learning points

- Patients with altered mental state with intracranial meningioma need to be assessed carefully for reduction in vision.
- Intracranial meningioma may be associated with the rare but potentially blinding VKH syndrome.
- Further study is needed to establish if such an association between these two conditions is more prevalent and the role of the HLA DR4 gene in the aetiology.

Competing interests None.

Patient consent Obtained.

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