Vogt -Koyanagi -Harada syndrome - A case report

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Summary:

A 30 year old Malay male developed bilateral choroiditis followed by vitiligo, poliosis and canities. CT scan showed nodular thickening of the optic nerves. Characterisitic abnormalities were seen in visual and brainstem auditory evoked potentials and fluorescein angiography. These findings were in keeping with the diagnosis of Vogt-Koyanagi-Harada syndrome. Treatment with dexamethasone resulted in complete recovery of vision and partial improvement in skin and hair lesions.

Key words: Uveitis, optic nerve, poliosis, Vogt-Koyanagi-Harada syndrome

Introduction:

Vogt-Koyanagi-Harada syndrome (VKHS) is a disease entity consisting of an inflammatory reaction of the uvea, the retinal pigment epithelium and the meninges, variably accompanied by involvement of brain substance, pathology of the cranial nerves, hair and skin changes in the form of poliosis, canities, alopecia and vitiligo [1,2]. The etiology of VKHS is considered to be autoimmune and both cell mediated and humoral immune arms may play roles in the pathogenesis[3]. We report a case of VKHS.

Case Report:

A 30 year old Malay male was hospitalised because of headache and sudden onset of blurred vision of two days duration. Examination revealed visual acuity of 6/18 in both eyes with reactive pupils. Fundoscopy revealed large areas of bilateral serous detachment and hyperemic swollen optic discs. All other organ systems were normal on examination. He was diagnosed as having optic neuritis with chorioretinitis. A month later he noticed patchy whitening of hairs on his scalp (canities), eyebrows (poliosis), moustache and patchy whitish pigment around the mouth, the limbs and body

which was non-pruritic and non-anaesthetic (figure 1). With the above combination of symptoms, a diagnosis of VKHS was made.

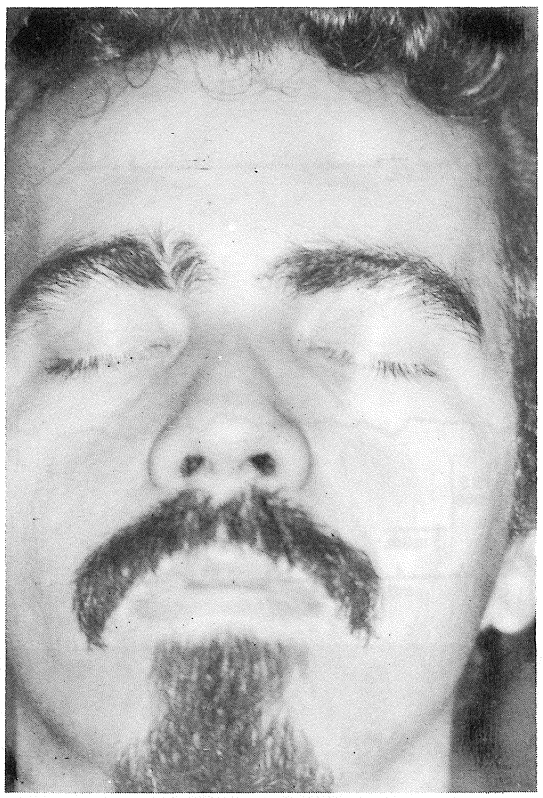


Fig. 1 Photograph of the patient's face showing white hairs in his eyebrows, eyelashes and moustache.

Investigations revealed normal haemogram. Anti-nuclear antibody, serology for syphilis, LE cells were negative and C_3, C_4 levels were normal. Fluorescein angiography revealed a number of areas of massive retinal leak visible around the posterior poles in addition to large pale choroidal areas. The large choroidal areas showed slow filling and leakage. Visual evoked potential (VEP) showed normal P_1 latency but broadened waveforms and brainstem auditory evoked potential (BAEP) showed absent response from left ear and low amplitude of the waves from the right ear (figure 2). Repeat VEP and BAEP were done four months later which showed similar pattern. CT scan showed nodular swellings on optic nerves (figure 3).

He was treated initially with oral Prednisolone 60mg daily with tapering dose for one month. He did not respond well to this regime. The treatment was changed to oral Dexamethasone one mg QID for three months and then tapered off over another two months. After one month of treatment, he responded very well. His visual acuity improved to 6/5 bilaterally and his poliosis and canities disappeared. On review one year later, his accomodation and fundi were quite normal. The optic discs were pink with clear margins. His hair and eye brows were dark. There was slight improvement in his vitiligo.

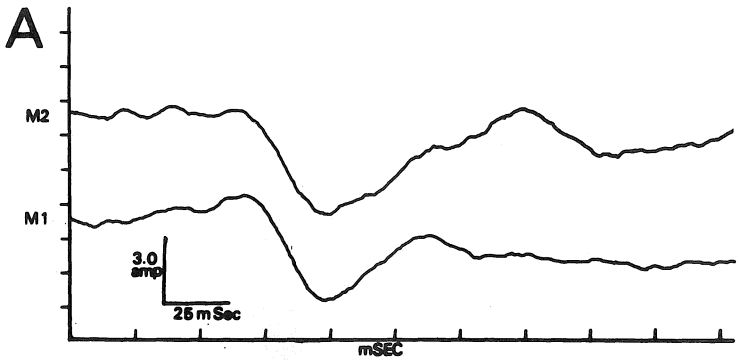
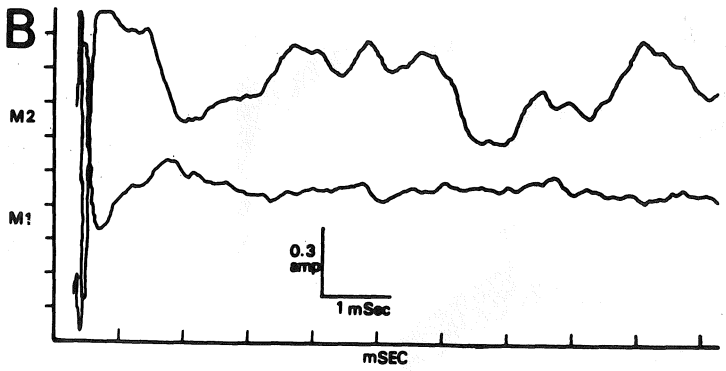


Fig.2
A
Visual evoked potential showing broadened wave forms.



B
Brainstem auditory evoked potential showing absent response on the left and low amplitude waves on the right.

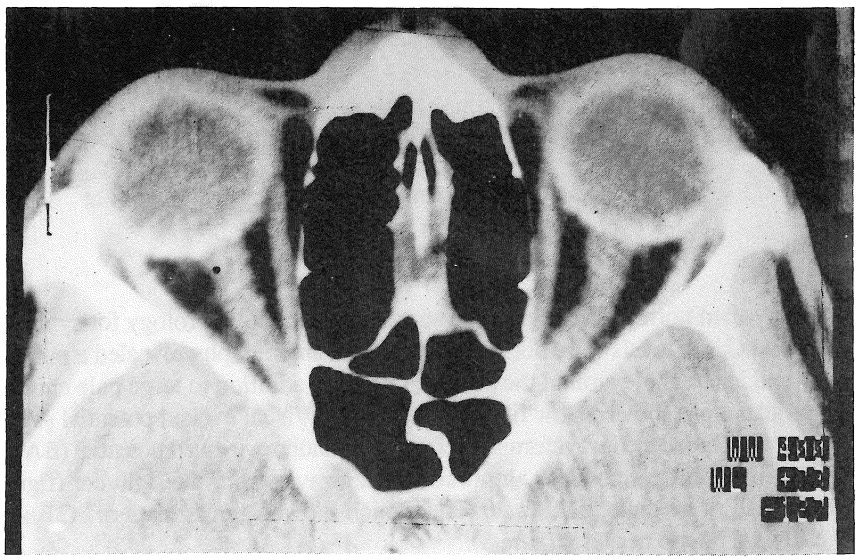


Fig.3 CT scan of the orbit showing irregular thickening of the optic nerves.

Discussion:

The VKHS occurs mainly in the third and fourth decades of life. Orientals, Negroes and darkly pigmented Caucasians are more susceptible to the disease, thus explaining the markedly greater frequency of VKHS in the Far East (especially in Japan), in Latin America and in the Mediterranean area. The most common presenting symptoms are ocular[4]. Our patient is a man in his third

decade of life whose initial presentation was with posterior uveitis and headache. These findings were similar to that described by Manor[4].The neurological features in VKHS are quite variable.The neurological picture in our patient was more of cranial nerve involvement notably the second and eight nerves, even though the patient did not complain of hearing loss. Fixed dilated pupils and impaired accomodation can occasionally be found as reported by Levy et al.[5] Our patient did have dilated pupils, though there was no impairment of accomodation. The delay in appearance of skin and hair changes has been well described in VKHS. Their occurence concomitant with the opthalmic or neurologic manifestations may hasten considerable the identification of the disease. In the 48 cases reviewed by Manor, poliosis was present in 39%, alopecia in 33% and vitiligo in 31%[4]. The time interval between the onset of the disease and the skin and hair changes ranged from two weeks to four years. But the interval was commonly between four weeks and eight weeks. In our case the time period was four weeks.

The cerebrospinal fluid (CSF) examination could not be done as lumbar puncture was not consented to CSF findings reported in VKHS include pleocytosis of four to 700 cells per cubic millimetre mostly lymphocytes and elevated CSF gammaglobulin concentration. The VEP findings was consistent with that found by Manor where the P_1 latency was normal but the waveforms were broadened[4]. BAEP confirmed the eight nerve involvement with absent response from the left ear and low amplitude response from the right ear. This showed that there was bilateral acoustic nerve involvement. The other interesting feature in this patient is the CT finding of irregular optic nerve swellings. The nodular swellings could be due to the inflammation of the optic nerve.

Our patient was treated with oral Prednisolone for a period of one month, with no objective improvement in his visual acuity. The treatment was changed to oral Dexamethasone. He responded to this, within one month of treatment, with an increase in visual acuity from 6/18 to 6/5. There were no neurological sequelae in our patient, except for the dilated non-reactive pupils. His visual acuity returned to normal. This was in contrast to the findings of Manor where visual acuity was lost in 17 out of 48 cases he reviewed[4]. In conclusion we report a case of VKHS with good response to Dexamethasone.

Referances:

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