CASE NOTES

AN UNUSUAL CASE OF THE VOGT-KOYANAGI SYNDROME*

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THE Vogt-Koyanagi syndrome is a rare form of bilateral uveitis, with associated disturbances involving the skin, hair, and ears. Bilateral uveitis is present in all cases, appearing in both eyes at the same time, usually without other subjective symptoms. Alopecia and poliosis (bleaching of the eyelashes) are reported to occur in about 90 per cent. of cases. The aural symptoms coincide more or less with the eye symptoms, the causative lesion being apparently in the labyrinth or central nervous system. Occasionally there may be general symptoms indicating meningeal involvement.

A patient with the Vogt-Koyanagi syndrome recently attended the uveitis clinic at the Institute of Ophthalmology, London. The case is of interest as the initial symptoms pointed to a diagnosis of intracranial tumour.

Case Report

A married woman aged 47 was referred to the Uveitis Clinic on November 13, 1956, for investigation.

History.—The patient had attended Moorfields Eye Hospital on September 19, 1956, complaining of intermittent blurring of vision and headaches for the preceding fortnight. Records show that the visual acuity at the time was 6/9 in the right eye and 6/36 in the left. The media were clear but the fundi showed "gross papilloedema". The peripheral visual fields were full but the central fields showed enlargement of the blind spot. The patient was accordingly referred to the National Hospital for Nervous Diseases, Queen Square, with a suggested diagnosis of cerebral tumour. The presence of severe bilateral papilloedema was confirmed when she attended the Out-Patients Department there one week later, and admission for further investigation was arranged. At this time the eyes were slightly congested and watering.

On October 9, the patient was admitted to hospital, but by this time her chief complaint was of deteriorating vision, and she was found to have developed a severe bilateral uveitis with posterior synechiae. The visual acuity had fallen to 3/60 in either eye. The uveitis was treated with atropine and cortisone drops. Investigations, which included electro-encephalograms and skull and chest x rays, showed no evidence of an intracranial tumour. The cerebrospinal fluid values at this time were as follows:

Pressure: 100 mm. Hg. clear.
Cells: 139 lymphocytes per c.m.
Proteins: 55 mg./100 ml.
Pandy: Weak positive.
Lange: No change.
Wassermann reaction: Negative.

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The overall picture was of an increase in the protein content and number of cells. The albumin/globulin ratio was normal.

On November 2 she was discharged from the National Hospital, and she was subsequently admitted to Moorfields where, in addition to the local treatment of the eye, systemic steroid therapy was given.

Examination.—When the patient was seen at the uveitis clinic for the first time, the sight had deteriorated to “hand movements” only in both eyes. Marked ciliary engorgement was evident and the slit lamp showed endothelial oedema of both corneae, numerous fresh keratic precipitates, a heavy flare, and numerous cells, together with exudate on the anterior lens capsule and posterior synechiae. No fundus details could be seen, as the opacities in the media obscured any red reflex. The tension, to the fingers, was low in both eyes.

When the patient was questioned regarding the onset of the disease, it was disclosed that at the beginning she had experienced buzzing in the ears and some slight deafness, while voices seemed distorted. The medical history-taking elicited nothing apart from information concerning recurrent attacks of cystitis. The routine medical examination of the patient at the uveitis clinic showed no evidence of systemic disorder apart from lumbo-sacral osteo-arthritis. No ankylosing spondylitis was present.

Extensive investigations were then undertaken by the Department of Pathology. Swabs were taken from the conjunctival sacs but no organisms were grown on culture. On November 13, 1956, the blood count showed: Hb 98 per cent.; white blood cells 7,500 per c.mm. with a normal differential count; erythrocyte sedimentation rate 9 mm. in the first hour; blood Wassermann reaction, Kahn test, and gonococcal complement-fixation test negative. Brucellosis and toxoplasmosis tests were negative. The urine contained no abnormal constituents. The Mantoux test was negative to a dilution of 1/100.

Diagnosis.—The association of *dysacusia* with the onset of the condition was thought to be highly significant, and a provisional diagnosis of an incomplete Vogt-Koyanagi syndrome was made. When the patient was re-examined at the uveitis clinic in June, 1957, she had developed poliosis, and furthermore, she stated that she had experienced considerable loss of scalp hair some 6 months previously. No vitiligo of the skin could be seen. The development of poliosis, and the history of alopecia, confirmed the diagnosis. The ocular findings at that time were as follows:

Right Eye.—Visual acuity 3/60; eye white; tension normal to fingers; pupil well dilated, but numerous posterior synechiae present and fibrous tissue on lens capsule; keratic precipitates, cells, and flare still present, together with numerous vitreous opacities.

Left Eye.—Visual acuity, “counting fingers”; eye white; tension normal to fingers; *iris bombé* present with fine deposits on posterior corneal surface and moderate flare.

Treatment.—In July, 1957, a broad iridectomy *ab externo* was carried out on the left eye by Mr. S. J. H. Miller. The portion of excised iris was cultured in the hope that a virus might be recovered but none was grown.

Progress.—In September, 1957, both eyes still showed active uveitis; the visual acuity in the right eye was 6/60, but in the left eye it had not improved beyond “counting fingers”.

Neurological investigation of the eighth nerve was carried out on April 1, 1958, by Dr. Hallpike at Queen Square, and the report was as follows:

“Cochlear function—normal. Vestibular function normal apart from some abnormality in the pattern of caloric responses. This is probably attributable to difficulty with fixation, and may therefore be insignificant”.

Discussion

The aetiology of this disease is uncertain. The widespread nature of the lesions suggests a toxic cause, while the changes in pigmentation resemble the skin changes noted in certain cases of sympathetic ophthalmitis. The condition has been ascribed to a virus infection, but none has so far been isolated. An alternative theory attributes the condition to an anaphylactic effect of the uveal pigment on the skin and hair.

This patient presented with typical signs and symptoms of papilloedema. The nature of the papilloedema, which was such a marked feature in this case, is a matter for speculation. In view of the CSF findings, meningeal involvement seems the most probable explanation. Another possibility is that the disc swelling was associated with the onset of uveitis in the posterior segment.

Summary

A case of the Vogt-Koyanagi syndrome, presenting initially with papilloedema, is recorded. Iris tissue, removed at operation, 9 months after the onset of the uveitis, was unsuccessfully cultured for a virus.

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