VOGT-KOYANAGI DISEASE*

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ROBINSON (1954) described a case of uveitis with deafness "in order to stimulate interest in the early recognition of such cases, so that investigations of the cause may be carried out, and perhaps a more specific method of treatment found". While there is no known effective treatment for the condition (Reed, Lindsay, Silversides, Speakman, Monckton, and Rees, 1958), it has been suggested that prolonged maximal dosage of corticosteroids might be of value (Crawford, 1953). The following case may therefore, be of interest.

Case Report

A housewife, aged 36 years, attended the Eye Out-Patient Department, on April 28, 1960.

History.—For 7 years, the patient had attended an ophthalmologist in another area with repeated attacks of hazy vision, for which she had been treated with local atropine and cortisone, and a prolonged course of oral cortisone during the year 1958. Recently, however, she thought that her sight between attacks was becoming more blurred, and for 2 weeks before her attendance, there had been a dense haze in front of both eyes. She had noticed white right upper eyelashes for one year. There was no history of headache, alopecia, hearing difficulty or ocular trauma. The previous history included a full medical investigation as an in-patient in 1959, for a thyroid swelling which was diagnosed as a simple cystic adenoma of the thyroid. At that time, the only unusual finding was one white eyelash in the right upper lid, and the blood picture showed a relative lymphocytosis (53 per cent.).

Examination (April, 28 1960).—The visual acuity was 6/36 in the right eye and 6/60 in the left, not improved with correction. Both eyes were white. The irides were atrophic, both pupils being small and irregular with multiple posterior synechiae: there were many large keratic precipitates in both eyes. A red reflex could be made out only with the ophthalmoscope, because of the small pupils and vitreous opacity. The ocular tension was not palpably abnormal. There were several white lashes among the black lashes of the right upper lid (Fig. 1).

FIG. 1.—Poliosis of right upper lashes.

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The patient was admitted to hospital for further examination. She was healthy-looking, 5' 6½" tall, and weighing 10st. 12lb. General examination revealed no abnormality other than a palpable right upper lobe of the thyroid. The blood pressure was 130/90, and the pulse rate 72 per min. The urine contained no sugar no albumen. The erythrocyte sedimentation rate was 10 mm. in the first hour. Mantoux test 1: 10,000+. Wassermann reaction and Kahn test negative. Agglutinins to typhoid, paratyphoid A and B, Salmonella, and *B. abortus* were negative. X rays of chest and hands were normal.

A blood count showed no abnormality.

There was slight upper tone loss in the hearing of the left ear. While in the ward between May 15 and 19, the patient developed a patch of alopecia in the occipital area. (Fig. 2). In view of the lack of meningeal symptoms, it was felt that lumbar puncture was not justifiable.

FIG. 2.—Alopecia of scalp hair.

Treatment.—On admission, the patient was started on a regime of isoniazid 50 mg. three times a day, para-amino salicylic acid 3 g. four times a day orally, Dimycin 1 g. intramuscularly on alternate days, and local 1 per cent. Antrenyl and Prednisolone 21-phosphate drops, 4-hrly to each eye. 3 per cent. iodine in CCl₄ was painted on to the bald area of the scalp after the alopecia developed. After 25 injections of Dimycin, the patient developed an allergic skin reaction; this part of the treatment was stopped and the rash cleared.

Progress.—On June 2, 1960, the visual acuity was 6/12 in each eye, a few old keratic precipitates were present, and the pupils were still small with many adhesions. In October, 1960, the visual acuity remained the same, and a fair view of the fundi failed to reveal any posterior lesion. In December, 1960, the patient developed a "cold", with a recurrence of hazy vision, and the visual acuity fell to 6/60 in the right eye and 6/18 in the left, with marked vitreous opacity in both eyes. She was re-admitted to hospital and a

course of Medrone was started, in addition to the existing treatment. The loading dose was 30 mg. and this was gradually reduced to a maintenance dose of 4 mg. twice daily.

Within 3 weeks, the visual acuity had returned to 6/12 in each eye, with much clearing of the vitreous. The Medrone dosage was then cut to 2 mg. daily, for 3 months, at the end of which time, the visual acuity was 6/9 (partly) in each eye. The scalp hair had now fully regrown. Medrone dosage was then cut to 2 mg. every second day, and this dosage was maintained until July, 1961, when the patient again reported that she had had a cold, with a persistent headache for 2 weeks, at which time her eyes had been hazy.

The visual acuity then was down to 6/60 in the right eye, and 6/24 in the left. There were many large keratic precipitates in both eyes, and the vitreous had again become cloudy, though no abnormality of tension was noted. The Medrone was then stepped up to 4 mg. daily for 2 weeks, and 2 mg. daily for a further 2 weeks. In August the patient reported again. The visual acuity in the right eye was 6/24, and in the left 6/12. Some old keratic precipitates remained in the right eye, but there was no flare. Some vitreous opacity was present in both eyes.

At this point, the patient was beginning to show some increase in weight, and the Medrone was cut to 2 mg. every second day for a week, followed by 2 mg. bi-weekly for a week.

On September 15, 1961, the visual acuity was 6/18 in the right eye, and 6/12 in the left. Many old keratic precipitates were present in the right eye, and a fair amount of vitreous opacity was present in both eyes.

In view of the patient having put on 16 lb. since beginning treatment, it was decided to discontinue the systemic corticosteroids, and 4 mg. of Depo-medrone was given subconjunctivally into the right eye.

Comment

This patient now presents the classical picture of anterior uveitis, poliosis, alopecia, and deafness generally known as the Vogt-Koyanagi syndrome. There has been, as yet, little to indicate a posterior uveitis, and for this reason, the name Harada has not been attached, although many consider the diseases to be identical.

Cowper (1957) attempted to classify the disease into three phases:

- (a) Meningeal of abrupt onset, 2-4 weeks in duration, with low fever and headache;
- (b) Ophthalmic, which may accompany or follow (a), with bilateral photophobia, lacrimation, loss of vision, and the typical picture of anterior and/or posterior uveitis, which may last from 3-5 months;
- (c) Convalescent, 6-12 months, during which the alopecia and poliosis may appear.

Duke-Elder (1947), however, states that the skin and hair changes appear 3 weeks to 3 months after the onset of ocular symptoms.

In this case, the history of uveitis is long standing. There has not, until one month ago, been any history of headache, nor has there yet been any complaint of deafness. The integumentary changes did not become apparent until 6 years after the onset of the ocular symptoms, and the latter have been atypical, in that the patient has suffered neither photophobia, lacrimation, nor congestion, the eyes remaining white, even at the height of an attack.

This patient, therefore, demonstrates the difficulty in coming to an early diagnosis.

From the point of view of treatment, the Dimycin, para-amino salicylic acid and isoniazid were given empirically. As the natural history of the disease is characterized by remissions, it is difficult to assess the success (or otherwise) of any treatment.

Rosen (1945) found, in a group of 47 patients, that thirty had vision of 3/200 in each eye, and only seven had vision better than 20/50 in one eye. In two known cases, deafness has persisted.

Reed and others (1958) stated that "less than 30 per cent. of patients regain useful vision", and this opinion is further elaborated by Böke (1959), who found that, when the disease involved both the anterior and posterior segments, the eye eventually progressed to complete blindness, in spite of the use of corticosteroids, whereas when the lesion was limited to the posterior segment, oral corticosteroids produced an immediate response, with return of full function of the eye.

In this case, the patient has had one recurrence while on oral corticosteroid treatment, and two recurrences while on the anti-tuberculous régime. It was considered fitting to treat the patient with systemic drugs on account of the generalized symptomatology. The scalp hair, however, has now regrown, and there is no apparent alteration in the lash or hearing condition. The fact that the visual acuity between attacks is being fairly well maintained, encourages one to continue with corticosteroid therapy, and as the patient is now beginning to show corticosteroid side-effects (moon-face and weight increase), it is proposed to continue therapy by means of subconjunctival injections of 4 mg. Depo-medrone, this treatment having been instituted one week ago.

As far as aetiology is concerned, there is nothing in this particular case to support to a greater extent, either of the commonly-held theories of allergic or virus origin. Although it was (inadvertently) sought in 1959, there was no demonstrable endocrine abnormality, a finding which supports the theory of Jaffé (1950) that the associated endocrine disorder sometimes found in these cases is fortuitous rather than causative.

The similarity between this condition and sympathetic ophthalmitis has already been stressed on many occasions, but this has not helped to resolve the aetiological nature of either disease.

Summary

A case of Vogt-Koyanagi disease is described. The difficulty of making an early diagnosis is demonstrated. It is proposed to continue corticosteroid therapy by subconjunctival injection, in view of the side-effects which accompanied oral therapy.

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REFERENCES

- BOKE, W. (1959). *Klin. Mbl. Augenheilk.*, **154**, 407 (cited by Kimuna, 1960).
COWPER, A. R. (1951). *A.M.A. Arch. Ophthalm.*, **45**, 367.
CRAWFORD, H. E. (1953). *Hawaii med. J.*, **13**, 26.
DUKE-ELDER, S. (1947). "Text-book of Ophthalmology", vol. 3, p. 2364. Kimpton, London.
JAFFÉ, N. (1950). *Amer. J. Ophthalm.*, **33**, 571.
KIMURA, J. (1960). *A.M.A. Arch. Ophthalm.*, **63**, 577.
REED, H., LINDSAY, A., SILVERSIDES, J. L., SPEAKMAN, J., MONCKTON, G., and REES, D. L. (1958).
Canad. med. Ass. J., **79**, 451.
ROBINSON, P. K. (1954). *Brit. med. J.*, **2**, 792.
ROSEN, E. (1945). *A.M.A. Arch. Ophthalm.*, **33**, 281.