Vogt-Koyanagi Syndrome

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THE ENTITY known as Vogt-Koyanagi syndrome is familiar to most ophthalmologists. It is very similar to (or perhaps even a form of) other reported disease entities such as Harada's disease, Cogan's syndrome and forms of uveo-encephalitis. All the diseases and syndromes mentioned have variations as to symptomatology but common to them are bilateral inflammation of the uveal tract, of varying degrees, and auditory abnormality. Also, there may be varying degrees of labyrinthine involvement. It is because of the symptom of hearing loss that the condition becomes of interest to an otologist. Sudden deafness was the first symptom in the case herein reported.

The disease complex was first described in this country by Parker in 1931 and the term Vogt-Koyanagi syndrome was applied by Carrasquillo in 1942.[4] The etiologic factors are not well understood but the disease is considered in some cases to be a form of sympathetic ophthalmia. The consensus is that a virus is the principal cause, but some investigators believe it due to hypersensitivity. Recently, in the case of Cogan's syndrome, there have been suggestions of similarity of the syndrome to periarteritis nodosa, as was emphasized by a report by Boyd in 1956.[1] Although the disease was considered rare, in that there were only 70 cases reported up to 1953, more and more have been observed and five cases were reported in San Francisco alone in one year.[2]

The points of similarity and differences between the entities described are shown in Table 1. Vertigo, tinnitus, bilateral nerve deafness, pain in the eyes and engorgement of the ciliary vessels of the eye are common to all three syndromes. Also, labyrinthine function is depressed to a variable degree in all three. Only as to uveitis, alopecia and vitiligo is there a definite distinction between the disease entities. Hence the conjecture that they are probably variants of a similar pathologic process.

Because of the nature of the disease, it is of interest to note the possible effect on it of the administration of corticotropin.

REPORT OF A CASE

A 21-year-old woman student had diarrhea. Then acute infection of the upper respiratory tract, with sore throat and earache, developed within a few days. Upon treatment by a physician, the earache subsided but dizziness and impairment of hearing then developed and the patient was referred to an otologist.

The patient complained of pronounced impairment of hearing, pulsating tinnitus, dizziness and unsteadiness of gait, moderate coryza and a slight change in the voice. Upon examination, the presence of serous fluid in the right middle ear was noted and there was considerable engorgement of vessels in the left drum membrane. Audiograms showed profound bilateral loss of hearing of a nerve impairment type at all frequencies between 50 and 60 decibels. No responses to bone conduction tests were obtainable. Right myringotomy and aspiration was done

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TABLE 1.—*Comparison of Symptomatology of Vogt-Koyanagi Syndrome with that of similar conditions*

	Vogt-Koyanagi	Harada's	Cogan's
Vertigo	±	±	+
Tinnitus	+	+	+
Nerve deafness (bilateral)	+	+	+
Pain in eyes	+	+	+
Engorgement of ciliary vessels of the eye	+	+	+
Labyrinthine function	±	±	Depressed
Uveitis	Anterior segment	Posterior segment	—
Alopecia	+	+	—
Vitiligo	+	+	—

but there was very little improvement. Audiograms about a week later showed no improvement in hearing.

Edema of the eyelids having developed during this period, the patient was referred to an ophthalmologist, who noted pronounced photophobia and slight bilateral loss of vision with considerable edema of the left lids and conjunctiva. Mild iritis was present. No disease of the retina was observed. Hydrocortisone drops were used topically and Meticorten® was given by mouth (0.5 mgm. tablets, four times a day for three days). Three days later, the swelling was less than before in the left eye but greater in the right. Photophobia persisted.

The patient was later referred to a neurologist, who reported no neurological evidence that the impairment of hearing might be owing to a space-occupying lesion in the central nervous system.

In the course of extensive examination three weeks later, the patient was seen by three otologists and by three ophthalmologists. In addition, several neurologists were called in consultation. As part of the examination, she had testing of the labyrinthine mechanism using caloric tests and there was no response to this testing in either ear. During the three weeks of hospitalization, the following were carried out:[3] An electroencephalogram, hemogram and sedimentation rate, a fasting blood sugar and nonprotein nitrogen determinations, a Kolmer test, and determination of the protein content of the spinal fluid. None showed evidence of deviation from normal. Virus studies were negative for the entire virus panel.

The thresholds obtained by audiometry were checked by psychogalvanic skin response and were found to be consistent and speech test results were consistent with pure tone test results.

During the three weeks the patient was in hospital, both topical and systemic steroid therapy were continued. The ocular symptoms improved dramatically but the hearing only slightly.

The improvement which was noted almost immediately in the ocular symptoms was probably owing to the fact that Meticorten therapy was begun almost at the onset of this particular case. It is interesting to note that the symptoms involving the eighth nerve were not appreciably affected by the oral cortisone therapy.

When last examined some six months after the onset of the disease, the patient was considerably improved with regard to general condition. The symptoms referable to the eyes had completely subsided and the hearing was slightly improved.

SUMMARY

In a case, which suggested a possible variant of Vogt-Koyanagi syndrome, points of similarity between that condition, Harada's disease, Cogan's syndrome and uveo-encephalitis were noted, and the suggestion was reaffirmed that these are probably a variance of a similar disease. The exact etiologic process is not known but the possibility of a virus origin plus a hypersensitive state is suggested. Steroid therapy was employed throughout this case. Pronounced improvement and ultimate cure of ocular symptoms occurred but symptoms relative to the eighth nerve were not much improved.

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Auto-Sensitization to Erythrocytes

A Report of a Case

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THE CLINICAL SYNDROME characterized by painful ecchymosis with auto erythrocyte sensitization was described by Gardner and Diamond[1] in 1955. We recently observed a case which corresponded with their description. With the exception of two possible cases reported by Whitfield[2] in 1921, the four cases reported by Gardner and Diamond are the only cases of this syndrome in the literature.*

CASE HISTORY

A 40-year-old woman, first observed March 5, 1956, for treatment of a large, painful area of ecchymosis in the right popliteal area, gave a history of occasional development of ecchymotic lesions, mostly in the extremities and sometimes across the shoulders. Pain deep in the muscles or subcutaneous pain usually preceded appearance of the lesions, and little or no trauma was associated with them.

In 1947 pain developed in the right shoulder. It was diagnosed as arthritic and was treated with heat. In December 1950, painful areas developed in the shins and there was a recurrence of the shoulder pain. In March and April 1951, during an episode of "flu," the patient noted generalized aches and especially pain in the wrists and forearms. A purpuric eruption over the right wrist ensued. She also noted profuse bleeding from even slight cuts. Biopsy specimens of skeletal muscle and skin were removed and when examined interstitial myositis, periangiitis, edema of the dermis and endothelial proliferation

*Since this report was prepared, additional cases have been reported (Gottlieb, P. M., Stupniker, S., Sandberg, H., and Woldow: Erythrocyte auto sensitization, Am. J. Med. Sci., 233:196-203, 1957.

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