UVEOPAROTITIS; A REPORT OF TWO CASES*

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DURING recent years there has been a decided increase in the number of reported cases of uveoparotitis. A rather comprehensive review of the literature reveals that approximately 100 such cases have been reported, less than 20 of which come from America and the remainder from foreign countries, chiefly Scandinavia, England and Germany. The rarity of this syndrome warrants the mention of additional cases and the careful scrutiny of these entities with regard to etiology.

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Parotitis, uveitis, and a low grade fever are the essential features of this condition. Prodromal symptoms, lasting for several weeks or months, include general malaise, drowsiness, weakness, nausea, vomiting, and loss of weight. Other signs and symptoms which are often encountered during the course of the illness include paralyses of the cranial nerve (particularly the seventh), a slightly elevated, discrete, erythematous rash occurring chiefly on the extensor surfaces of the forearms and legs, dryness of the mouth, various aching pains, polyuria and night sweats. Occasionally the submaxillary, sublingual or lacrimal glands are enlarged and also the cervical lymph glands.

The disease runs a chronic course, the symptoms lasting for months or for several years, and one of its features is the irregularity in the onset of symptoms. It may occur at any age, but is more common in the second and third decades and is slightly more prevalent in females. The morbidity is high whereas the mortality is low, only five deaths having been recorded.

The parotid glands are usually involved early in the course of the disease. They become enlarged and hard, and although not painful may be slightly tender. The glands are not adherent to the skin and never suppurate. The swelling, which is usually bilateral, lasts for months or years.

Inflammation in the uveal tract is present in all cases of uveoparotitis. The first ocular symptom is usually dimness of vision, associated with circumscribed corneal injection and narrowing of the palpebral fissures. Occasionally there is pain. Although in the beginning the involvement may be unilateral, bilateral uveitis usually follows. The pupils are often sluggish, dilated, or irregular, with a tendency to develop posterior synechiae. The other important eye findings which may be observed are fatty deposits on the posterior surface of the cornea, nodules in the iris, vitreous opacities, keratitis, optic neuritis and atrophy.

Reference is made to several excellent reviews of the literature on this subject, and no attempt will be made to duplicate such reports. The first description of this syndrome was given by Heerfordt [1] in 1909. Garland and Thompson,[2] Savin,[3] and Folger [4] have made comprehensive and critical surveys of the literature on this subject, while Levin [5] has reported in detail on the neurological aspect of the syndrome.

Case Reports

Case 1. A 35 year old negro male came to the Harbin Hospital on May 17, 1937, with a chief complaint of aching pain throughout the chest and abdomen associated with nausea and vomiting. On several occasions during the previous year he had had severe gastrointestinal "upsets" associated with aching pain in the chest and abdomen. His temperature had been moderately elevated for several days with these attacks. His employer stated that at times he was rather drowsy and acted as if he might be drugged, enough so that it was questionable whether he should continue his work as a chauffeur.

About three years prior to the time he was first seen he had consulted Dr. George B. Smith (ophthalmologist) because of dimness of vision, and in July 1934 a diagnosis of uveitis was made.

The patient's first wife had died of pulmonary tuberculosis in 1930, and one child by this wife was confined to a tuberculosis sanatorium at the time of his examination. His second wife was healthy. He used tobacco excessively and during each winter had frequent head colds with a slight cough. At the age of 20 he had had a gonorrheal infection, but no history of a chancre could be obtained.

Physical examination revealed a well developed, well nourished negro male not acutely sick. The lacrimal glands were swollen; considerable bilateral circumcorneal injection was present; the pupils reacted sluggishly to light and were irregular. Moderate dental caries and pyorrhea were present. Both parotid glands were enlarged, firm, smooth and slightly tender without any fixation of the skin. Upon being questioned, the patient stated that his wife had commented upon the appearance of each side of his face several times in the four or five months prior to the time of his examination. The submaxillary glands were not enlarged but several cervical lymph glands were palpable. Physical examination of the heart and lungs was not remarkable. The blood pressure was 120 mm. Hg systolic and 78 mm. diastolic. There was slight tenderness in the region of his umbilicus; otherwise his abdomen was negative. The biceps and patellar reflexes were normal. Dr. Smith reported that the uveitis had improved, that no posterior synechiae were present, and that his visual acuity was normal.

The first specimen of urine examined contained a small amount of albumin and a moderate amount of pus. Urinalyses at later dates, after the administration of urinary antiseptics, were negative. The blood findings were as follows: hemoglobin 92 per cent (Sahli), red blood cells 4,010,000, leukocytes 9,650, polymorphonuclears 60 per cent, eosinophiles 2 per cent, mononuclears 3 per cent, small lymphocytes 31 per cent, large lymphocytes 4 per cent. The blood Wassermann and blood Kahn reactions were negative. No malarial parasites were found. Examination of the spinal fluid revealed a cell count of 6, very small amounts of globulin and sugar, and a negative Kahn reaction. Colloidal gold showed no color change. A chest roentgenogram revealed normal lung fields and a cardiac outline within normal limits. The injection intradermally of 0.01 mg. of Old Tuberculin gave a positive reaction.

On June 15, 1937, the patient stated that he felt much better, that his drowsiness was less, and that no attacks of vomiting and abdominal pain had occurred. The physical findings on that date were unchanged. By request he returned on March 28, 1938 for a routine examination. Since last seen he had had two "slight attacks" of fever and general aching in the chest and abdomen, accompanied by vomiting. He stated that the enlargement of the parotid glands had varied in size, and they were found to be definitely smaller than at the time of his first examination. His sight had been good, and only a slight ciliary injection was noticed. He continued to be drowsy at times. The examination of his urine was negative.

He was seen again on June 24, 1938, at which time he had no complaint. The swelling of the lacrimal glands had disappeared and the eyes were grossly normal. The parotid glands continued to be slightly enlarged, chiefly on the right side. He had no nausea, vomiting, or aching pains, and his urine was negative. On March 2, 1940, he stated that his general health had been good since June of 1938. Very slight enlargement of both parotid glands was present, and the ophthalmologist reported normal eye findings.

This patient was not observed when he first had symptoms probably associated with uveoparotitis, as he reported dimness of vision almost three years before he was examined. As might be expected, in view of the intellect of the patient, the history regarding the duration of his general symptoms was vague. At no time did he take his condition seriously, and he was examined at the insistence of his employer.

Case 2. A 21 year old white American female came to the Harbin Hospital March 26, 1938 with a chief complaint of blurred vision in the right eye. For several days her right eye had been red, and the visual disturbance had been noticed for about two weeks. The patient stated that she had been a nervous person all her life but more so recently. At the age of 13 she was taken out of school for one year upon the advice of her physician because of a "rapid heart rate." Several years later she was told by another physician that she had a "leaking heart." It was not possible to elicit a history of any other cardiac disturbance. Two years prior to the time of her

examination she had had slight contact with a person who had active pulmonary tuberculosis. Otherwise her past history was not remarkable, and a review of her systems was non-contributory.

Physical examination revealed a poorly nourished, fairly well developed woman, 5 feet 4½ inches tall, weighing 108 pounds. Her temperature was 99.2° F. The pupils were equal, regular, and reacted to light and accommodation. A slight epicorneal injection was present in the right eye. The mouth was not remarkable, with the exception of slight dental caries. The parotid and submaxillary glands were not enlarged, neither were the cervical lymph glands. Examination of the heart and lungs did not reveal any positive findings, and the blood pressure was 118 mm. Hg systolic and 82 mm. diastolic. The abdomen was negative and the reflexes physiological. The blood count was as follows: hemoglobin 75 per cent (Sahli), red blood cells 3,760,000, leukocytes 7,500, polymorphonuclears 55 per cent, monocytes 2 per cent, eosinophiles 3 per cent, small lymphocytes 34 per cent, large lymphocytes 6 per cent. The urine was found to be normal, and the blood Kahn test was negative. A roentgenogram of her chest revealed normal lung fields and a cardiac outline within normal limits. A tuberculin test using 0.005 mg. of purified protein derivative tuberculin was negative.

During the following weeks her temperature ranged from normal to 100.5° F., and on April 22, 1938, a slightly elevated macular erythematous rash appeared on the arms, lower legs and feet. On April 29, 1938, an ophthalmological examination was made by Dr. F. P. Calhoun, and his report was as follows: "Vision right and left 15/15. The near points of accommodation were 10 cm.; there was no muscle imbalance and the intraocular tension was normal. The right eye showed a slight pericorneal injection, and the left eye showed a moderate pericorneal injection. Dilating the pupils for a fundus and slit lamp examination, many cells were found on the posterior surface of the right cornea; none was found on the left cornea although the aqueous beam was cloudy. With the ophthalmoscope vitreous cells were seen in the right eye but no other pathology, and the fundus of the left eye was normal. Diagnosis, bilateral cyclitis."

When the patient was seen on May 5, 1938, both submaxillary glands were enlarged and slightly tender. No enlargement of the parotid glands could be detected; the skin rash was still present and had not changed. After having been on a high caloric diet for several weeks she had gained about two pounds in weight. At that time she complained of a dry sensation in her mouth. On May 12, 1938 she stated that she had been nauseated for 18 hours and had vomited several times. During the few days preceding that date she had had various aching pains throughout her body which at times were severe. She continued to have a slight elevation in temperature, and her skin rash was fading. The ophthalmologist reported that her eyes were beginning to improve.

During the next few months she improved in general, the skin rash disappeared, her temperature was lower, and her eyes approached normal. The precipitates on the posterior surface of the cornea disappeared. On September 9, 1938, the left submaxillary salivary gland was not palpable, but the right continued to be slightly enlarged. By November 8, 1938, the patient was feeling very well and weighed 122 pounds. About one year later, in December of 1939, she complained that her vision was not quite normal, although it was fairly good. Dr. Calhoun reported as follows: "the vision was slightly improved and the slit lamp and fundus examinations were normal." Her general condition at that time was excellent.

During the early course of her illness therapeutic measures were instituted to improve her general condition. She was on a high caloric diet; ferrous sulphate was administered for the secondary anemia; she was in bed for several weeks; and efforts were made to relieve her nausea, aches and pains. At the suggestion of the ophthalmologist her pupils were dilated with atropine for several months until the cyclitis had completely subsided.

Comment

The first case presents typical findings of uveoparotitis. It is of interest to note that this patient had had much contact with active pulmonary tuberculosis and that his tuberculin test was positive. However, he did not have any clinical or roentgen-ray evidence of pulmonary tuberculosis. He improved considerably during the first few months he was under observation and after almost three years his condition continued to be good and his vision normal. The parotid glands were known to have been enlarged for about three years and were still enlarged at the time of his last examination in March 1940. No serological evidence of lues could be obtained.

In the second case the following characteristic findings of the uveoparotid syndrome are present: fever, bilateral cyclitis, skin rash, dryness of the mouth, nausea, vomiting, aching pains, and enlargement of the submaxillary glands. The fact that no enlargement of the parotid glands was detected makes a diagnosis of uveoparotitis questionable. In quite a number of the cases reported in the literature there was enlargement of the submaxillary or sublingual glands in addition to parotid enlargement. The authors feel that the evidence presented favors the diagnosis of uveoparotitis and would like to suggest that this syndrome may occur with enlargement of one of the salivary glands other than the parotids. Of interest is the fact that epidemic parotitis is known to occur with involvement of the submaxillary or sublingual glands, without any enlargement of the parotids.

The recognition of this condition is important from the point of view of prognosis and treatment. As a rule this disease is a self-limited one and treatment should be directed chiefly toward improvement of the patient's general condition, the alleviation of distressing symptoms, prevention of complications, and reassurance as to the probable outcome. One important therapeutic measure is the maintenance of mydriasis in an effort to avoid the formation of posterior synechia with permanent impairment of vision. Failing to diagnose this syndrome correctly may result in a faulty prognosis being given to its various manifestations, and may lead to unjustifiable treatment.

It has been established beyond any doubt that this syndrome is a well defined clinical entity. There is, however, a great diversity of opinion as to the etiology of this disease and a variety of causes have been suggested by authors who have reported cases. It is frequently referred to as uveoparotid tuberculosis, and in several cases evidence of tuberculosis has been obtained. There are those who believe that all such cases are tuberculous in origin but no sound proof has been offered to substantiate this theory. In the majority of cases tuberculosis has not been demonstrated, and the tuberculin reactions have been negative. Referring to this disease, Cohen and Rabinowitz [6] made the following statement: "the tuberculous patient is tuberculin sensitive except under conditions of massive tuberculous infection, but it must be conceded that the patients in this series had no massive infection, if at all infected, it was mildly, and yet the tuberculin reactions were negative in the majority of instances; in the cases in which the tubercle bacillus was demonstrated, the reaction was positive." It has been suggested by Lewis, Raines and Stewart [7] that this disease may not be due to any one specific infection but that it is a manifestation of local tissue hypersensitivity to any one of a variety of allergens, and that although tuberculin may

often be the allergen responsible, it is unwise to regard all cases of uveoparotitis as tuberculous. At the present time this appears to be the logical explanation of the cause of uveoparotitis. That it may be due to an organism which has not yet been identified must be considered. If the latter should be the case, this unknown organism apparently produces a low grade infection in a sensitized individual, the course of which is chronic with a spontaneous termination.

As it was not possible to demonstrate any primary tuberculous focus in the first case one cannot say that it was caused by tuberculosis, even though there was much contact with active pulmonary tuberculosis and the tuberculin test was positive. It must be remembered, however, that an infection of this type might have been present but not detected. In the second case the tuberculin reaction was negative, no causative agent was found, and its etiology remains unknown.

Summary and Conclusions

1. The findings in a typical case of uveoparotitis are described.

2. The possibility of this syndrome's occurring with involvement of one of the salivary glands other than the parotids is suggested and such a case is reported.

3. The importance of the recognition of this clinical entity is stressed.

4. The need of further knowledge of the etiology of uveoparotitis is pointed out. It is suggested that the syndrome may arise from hypersensitivity to a number of allergens.

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