SOME UNUSUAL CASES OF SJÖGREN'S SYNDROME

BY

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SJÖGREN'S syndrome indicates a general constitutional or systemic disturbance of unknown origin. Very complete investigations of typical cases have been made by Sjögren, Bruce, Lutman and Favata and Stenstam without finding any common aetiological factor, and all that can be said is that most cases appear in women at or after the menopause without any history of previous disturbance of the menstrual cycle.

In this regard it is well to bear in mind the extreme chronicity of the complaint and the possibility of the menopause being only an aggravating factor in a condition already established. The most striking impression gained from reading the details of published cases is the freedom of the patients from any disorder not referable to the syndrome described by Sjögren.

It is of interest, therefore, to record cases with unusual constitutional features, and numbers one and two described below are of a man and his daughter, the latter with a lifelong history of dry eyes and showing signs of pituitary dysfunction. The third and fourth are of a woman and her daughter, the latter having pulmonary tuberculosis. The fifth case is of a young woman whose symptoms seemed to originate from a fracture of the base of the skull.

CASE 1.—Because of a feeling of grittiness in his eyes and some hypersensitivity to light of many years duration, Mr. A., aged 68 years, had had a habit of sitting with his eyes closed when not at work. During this time he had also had slight dryness of the mouth. He made no complaint of these disabilities until a few months ago when he had a cerebral haemorrhage with hemiplegia and his eyes became worse.

He was found to have typical filamentary keratitis with a Schirmer's test of 5 mm. in 5 minutes (control, 25 mm.). The filaments and much of the discomfort disappeared in a few weeks with no more than the application of vaseline to the lid margins, though the deficient lacrimal secretion remained unchanged. The corneal sensitivity was normal. There was no history of enlargement of the parotid glands, and any rheumatism had been slight.

A diagnosis of keratitis sicca may be preferred to that of Sjögren's syndrome, and there is only his previous tendency to sit with the eyes closed to indicate that the condition preceded the cerebral haemorrhage.

CASE 2.—Miss G.A., aged 36 years, the daughter of Mr. A., above, said emphatically that even when crying she had never known a tear to come from either eye. Any irritation such as from foreign bodies had caused only a stinging sensation without lacrimation. Her mother stated that even as a child her daughter's eyes had been chronically sore and intolerant of light, and no tears had formed.

She came under observation 10 years ago at the age of 26 with chronic conjunctivitis, mucoid threads and myopia. A year later, in 1939, she consulted her physician, Dr. E. J. Fischmann, for amenorrhoea and a very large number of boils. She had been having severe headaches, sometimes with vomiting, and her size of shoes had changed from 5 to 7 with a proportionate enlargement of the hands. A roentgenogram of the sella turcica was thought to show some enlargement, but the fields of vision, blind spots and discs revealed no abnormality.

At this time Dr. Fischmann demonstrated the pituitary symptoms and signs to the Auckland Clinical Society. These consisted of obesity of a typical hypopituitary type, amenorrhoea, loss of hair on the scalp, growth of hair on the face, a tendency to kyphosis, striae distensae, slight exophthalmos, polydipsia, albuminuria and increased blood pressure. In addition and in contradiction to this picture of pituitary basophilism, she also had acromegaly.

Three months later, a small loss of peripheral field was detected down and out in the right field and up and out in the left. The blind spots and discs remained normal. The pituitary gland was then explored by Sir Carrick Robertson, but no tumour was found.

Shortly after the operation, the eyes became gritty and inflamed and showed fine superficial keratitis, chiefly of the upper half of each cornea, with slight pitting and fine scattered epithelial spots, many of which stained with fluorescein. Mucoid threads were present. Sensitivity was normal. About this time the mouth began to feel dry, and swelling of the parotid glands was first noticed. During the next eight months she was examined on several occasions without any material variation in the signs, though the degree of discomfort was very variable.

Service with the New Zealand Division abroad prevented further examination by the writer for some years, but, after the war, there was no doubt that she presented the typical features of Sjögren's syndrome. The corneae showed the same changes as before with, in addition, slight limbal vascularisation and some small filaments. Mucoid threads were present and Shimmer's test showed no absorption beyond the lid margin from either eye in 5 minutes (control 30 mm.). The discs were normal and the only field defect was a partial right centro-caecal scotoma. The mouth was dry and the parotid glands were considerably enlarged and tender. There was a history of rheumatism and the menstrual periods had not returned. She stated that she had seldom perspired and then only slightly.

There was no radiographic evidence of salivary calculus and the sella turcica was of normal size with intact bony outlines. The Kahn test was one plus positive and the Wassermann reaction doubtful on two occasions. At a third examination the Wassermann reaction was one plus positive. The blood count and sedimentation rate showed no abnormality.

In a personal communication, Dr. Fischmann gives the following additional information:—

"Involvement of the joints was observed on two occasions. In 1945, moderate pain and swelling of the proximal interphalangeal joint of the right middle finger appeared, accompanied by ache in the calf muscles, especially during the night. X-ray of the involved joint proved negative. In 1947, pain appeared in the knees, wrists and ankles with slight swelling of the ankles and the right thumb base joint.

The joint involvement does not conform with any of the three classical patterns of chronic joint disease (rheumatoid arthritis, osteo-arthritis and gout). It resembles the palindromic rheumatism of Mayo Clinic investigators more closely than any other joint disease known to me.

To test pituitary function, an insulin tolerance test, the basal metabolic rate and a water concentration and dilution test were done in 1945. These tests as well as an electrocardiogram were normal.

Would you think that the early cessation of ovarian function due to absence of the two gonadotrophic pituitary hormones may have played a rôle in the early onset of keratoconjunctivitis sicca?"

As the patient is intelligent and there is no reason to doubt the accuracy of the history it is probable that this is a case of Sjögren's

syndrome of congenital origin analogous to Duke-Elder's case of congenital keratoconjunctivitis sicca, and a case recorded by Hamilton. In the present state of our knowledge it is impossible to say what significance, if any, can be given to the pituitary dysfunction or the doubtful Wassermann reaction.

CASE 3.—Mrs. S., aged 58 years, complained of a varying degree of soreness of the eyes of about four year's duration. Various kinds of drops had been prescribed by her physician, but they had all been irritating and she had found by experience that the best remedy was frequent bathings with water. On being questioned, she stated that the mouth had been dry, the saliva frothy and the tongue at times inflamed for about eight years. The inflammation of the tongue had responded to treatment with vitamins. The parotid glands had been swollen and tender on three occasions in 1946. She had had her teeth removed about fifteen years ago and had not had any rheumatism.

On examination, the eyes showed the characteristic changes of keratoconjunctivitis sicca with a few fine filaments and a mild degree of staining with fluorescein. Shirmer's test showed only 2 mm. of moistening in each eye on two occasions.

The patient lived in a remote town and it was not possible to have any pathological investigations made.

CASE 4.—Miss M.S., aged 26 years, daughter of Mrs. S., case 3, was seen for the first time this year. She complained that her eyes had been chronically inflamed and sore in varying degrees for about four years. When questioned, she said that she could not remember ever having noticed any tears, that her voice had been husky for a few years and that, during this time, she had had slight dryness of the mouth in the mornings and a tender swelling of the parotid glands every month or two. This swelling had usually lasted only a few days and had twice been diagnosed as mumps. Her teeth had decayed at an early age and had all been removed when she was 17. Perspiration had always been free on the hands and feet, but not elsewhere. Her menstrual periods had been and still were quite normal.

As a child, she had been suspected of having pulmonary tuberculosis. This was confirmed in 1944 and she was admitted to a sanatorium, and, although the infection was under control, she still had a pneumothorax. Her health otherwise had been good except for slight rheumatism in the arms and legs. Her father had died of tuberculosis and her two sisters were affected. Two brothers were healthy.

She was a tall, round shouldered girl of unemotional temperament whose only obvious disabilities were her chest and her inflamed eyes. The characteristic mucoid threads of keratoconjunctivitis sicca were present, the corneae showed superficial keratitis chiefly in the upper halves and, when stained with fluorescein, showed much superficial punctate staining and many fine mucoid filaments. The vision was reduced by the keratitis to 6/9 in the right eye and 6/24 in the left. There was no intra-ocular abnormality. Shirmer's test showed an absorption of 4 mm. in the right eye and 3 mm. in the left in 5 minutes (control 30 mm.). The corneal sensation was normal.

The Wassermann reaction was negative and the blood examination revealed an increased sedimentation rate and such other changes as would be expected in a case of tuberculosis.

CASE 5.—Miss M.W., aged 33 years, met with an accident at the age of 21 in which the base of her skull was fractured, with consequential paralysis of the right side of the face, paralysis of the right external rectus muscle and deafness of the right ear. Except for diplopia, the vision was unaffected. When she recovered full consciousness she found that her mouth was dry, and she has been unable to eat dry foods ever since.

The facial paralysis recovered in about six weeks, but the external rectus paralysis, the deafness and the dry mouth have persisted.

A few months after the accident her teeth began to decay, her nose and throat became dry so that she had to gargle every morning, the saliva became thick, white

and scanty, and the tears did not form normally. The eyes, however, remained comfortable and free of inflammation.

About four years later the left eye became chronically inflamed and gritty and tended to close, especially in a strong light. When irritated, both eyes smarted but no tears formed. Mucous threads were sometimes present in the mornings.

Except for some slight enlargement of the thyroid gland at about the age of 17 her general health has been good, her menstrual periods have been regular and she has had no abnormalities of the skin and no rheumatism. The parotid glands have not been enlarged. Since the accident, however, her memory has been poor and she has had a persistent feeling of drowsiness.

On examination, she was found to be of healthy appearance and of unemotional temperament. The teeth showed very extensive repair. There was some chronic conjunctivitis, chiefly of the left eye, and both corneae showed fine superficial erosions staining with fluorescein. Shirmer's test revealed:—Right eye, 10 mm. of moistening, left eye, 6 mm., normal control, 30 mm. The differential blood count, sedimentation rate and Kahn test were normal.

The sella turcica was normal in shape and size and its bony outlines were well defined, regular and intact. There was no radiographic evidence of the old fracture of the skull.

COMMENT.

Some of these cases illustrate a common and understandable vagueness about the time of onset. The progress is usually extremely slow and there is a considerable difference between the result of Shirmer's test in the normal subject and in the patient with signs and symptoms of keratoconjunctivitis sicca. A patient may show the syndrome in a fairly advanced state without being greatly inconvenienced and it is usually impossible even to infer when secretion began to fail. Sjögren has pointed out that the organic changes in the gland appear earlier than the symptoms of diminished secretion.

Case 5 may be analogous to Wagenmann's mentioned by Duke-Elder. The history of xerostomia soon after the fracture of the skull is unusually definite, but it is noteworthy that the symptoms in case 2 increased after the craniotomy for pituitary tumour.

In speculation as to the cause of the syndrome, allowance must be made for the possibility of commencement of the pathological process long before the first complaint of symptoms.

The doubtfully positive Wasserman reaction in case 2 and the tuberculosis in case 4 suggest infection as a causative factor. This is supported by the case of a man of 60 recently referred to the writer by Dr. A. N. Talbot. He had keratoconjunctivitis sicca with slight aqueous flare in both eyes and a posterior synechia. His Wassermann reaction and chest roentgenogram were negative. These, however, are isolated instances the significance of which must not be overestimated. It is more reasonable to suppose a congenital defect which may be made manifest at any period of life by some aggravating circumstance. This suggestion is supported by Lisch's series in which twelve people in three generations of one family showed some evidence of the syndrome.

Lisch states that the majority were of asthenic physique and looked ill, and Lutman and Favata mention the slender build and nervous temperaments of their cases. All those mentioned in this paper and four others under treatment at the present time are phlegmatic rather than excitable and, excepting case 2 with pituitary dysfunction, cannot be said either to be of any constitutional type or to look ill.

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 ADAPTATION TO ENVIRONMENT

BY

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It has been a practice, especially during the war years, to analyse the job, to analyse the men, and to attempt to fit the right man into the right job. It is thus assumed that certain people have characteristics which enable them to develop along one particular line, and that along this line they will do better than along any other. Newton has taught that action and reaction are equal and opposite in the physical sphere, and to a limited extent the same may be true in the psychological. In other words, although the man has an effect on his work, his work also has an effect on him.

The majority of us become ophthalmic surgeons in rather a haphazard fashion; in fact, the same may be true of the whole medical profession, and it is more than likely that its members represent a random sample of the upper middle classes, it being only occasionally true that anyone goes into the medical profession because he has a particular flair for it, or thinks that he has.