Short Notes and Clinical Cases.

UVEO-PAROTITIC PARALYSIS.

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THE case described below in detail is one of a rare disease in which an inflammatory process of the anterior segments of the uvea, and of the parotid, is associated with peripheral neuritis, the latter involving the cranial nerves, usually the facial, and occasionally the nerves of the limbs and trunk.

This rare syndrome was apparently first recognized as a definite clinical entity in 1909 by Heerfordt,¹ who described three cases in his article entitled "On a Subchronic Uveo-parotid Fever, localized in the Parotid Gland and the Uvea of the Eye, and specially complicated with Paresis of Cerebrospinal Nerves." Heerfordt refers to other cases which, in his view, belong to the same syndrome. In a recent number of this JOURNAL, Feiling and Viner² again brought the syndrome into prominence by the description of a typical case under the almost equally elaborate title of "Iridocyclitis-Parotitis-Polyneuritis: a New Clinical Syndrome."

The recording of another case in this article gives me an opportunity for suggesting that the disease might be described by the shorter yet appropriate name of "Uveo-Parotitic Paralysis."

DESCRIPTION OF CASE.

The case is that of a married woman of forty-three, who was first seen in the out-patient department of the National Hospital, Queen Square, by Dr. Kinnier Wilson, and was later admitted under his care on August 27, 1923. Its salient features are summarized in the following account.

The illness began in November, 1922, when the whole of the right side of the patient's face and forehead had become "itchy"; at the same time her skin all over took on a yellowish tinge. This lasted for about two or three weeks, but was not accompanied by fever. About the same time a pain in the right side of the abdomen, of which she had complained at intervals for nearly two years, greatly increased in intensity and led to the operation of appendicectomy in December. Three weeks later the patient suddenly began to have pains in both legs, with a "pins-and-needles" sensation, which extended from the feet to the hips. When she attempted to walk, her legs

gave way and she fell. The weakness of the legs continued for three weeks, as also did the paræsthesiæ.

In the middle of January, 1923, while the legs were still affected, "lumps" appeared on both cheeks, just in front of the ear and below the mandible on both sides. These "lumps" were larger and more painful on the left than the right side, and were associated with mild fever. With the appearance of the swellings both eyes became painful, and, in addition, the conjunctiva of the right eye was injected, while the left was normal in this respect. Vision became misty in both eyes, but with the subsidence of the swellings this improved slightly. By the middle of February the swellings had entirely disappeared and only slight mistiness of vision in the right eye remained. The patient noted, however, that a circular shadow about the size of a penny was present in front of the right eye. The pain at the back of the right eye continued.

On account of pyorrhœa, some teeth were extracted under gas in the middle of February, some more two weeks later, and the remainder three weeks after that. After the second lot were extracted the patient was fevered and had a sore throat. Three weeks after the final extraction, on April 6, stiffness was felt on the right side of the face and with it pain of a "red-hot needle" type, extending all over the head. On April 7 the left side of the mouth was pulled up, and she could not close the right eye properly; in other words, the right side of the face was paralysed. There was also slight deafness of the right ear. The facial palsy improved and became practically well in July. With this improvement the pain at the back of the right eye disappeared, as did to a great extent the mistiness of vision and the shadow.

About the middle of August, however, the left side of the face suddenly became paralysed, with drawing up of the right corner of the mouth. The left eye could not be closed, and there was slight deafness of the left ear. Accompanying the left facial palsy was pain at the back of the left eye, and very bad dysphagia. This dysphagia had not been present with the right facial palsy. A shadow, similar to that seen with the right eye, was now seen with the left. Two days after the facial paralysis began the patient felt generally weak.

Just before admission to hospital, on August 27, she became unsteady on her feet, was fevered, and vomited undigested food. This only lasted one day.

Ever since the patient had her teeth extracted she has suffered with pains between the shoulders and in the pit of the stomach, but she only vomited on the occasion already mentioned. Headaches have been more or less constant since the right facial palsy commenced. Diplopia has never been noted.

Paræsthesiæ were present, as already described, at the beginning of her illness. Two or three weeks before her admission to hospital there was numbness at the tips of the middle three fingers of the left hand, which has never quite disappeared. During these last few weeks there has been a sharp pain from the tip of the fourth finger of the left hand to the shoulder. This pain lasted only a few minutes, but came on several times in the day.

Constipation has always been a source of trouble to the patient.

The previous history revealed nothing bearing on her present illness

except that she had mumps as a child. The family history was similarly unimportant.

STATE ON EXAMINATION.

On admission the patient was seen to be small and poorly nourished, though the abdomen was fairly well covered. Her complexion and skin generally were of a yellowish colour, but she stated that her mother's and father's people all had this sallowness. On the right side of the abdomen was the linear scar of the appendicectomy operation. No enlargement of any glands could be found. Tenderness was discovered only in the left mastoid region. There was no sign of any ductless gland disturbance. The patient's intelligence and memory were normal, and there were no delusions or hallucinations, or emotional overaction. The respiratory, cardio-vascular, alimentary and genito-urinary systems were normal.

Cranial Nerves.—Visual acuity was R. 6/9, L. 6/12. The fields and fundi were normal in all respects. The pupils were equal and central, though not quite circular. The pupillary edge was slightly irregular, and on ophthalmological examination small opacities were found well forward in the vitreous. Free action of the pupils to light and accommodation was present. The other cranial nerves were unaffected, except for the slightest diminution to cotton wool stimuli over the second and third divisions of the left fifth, and the marked weakness of the left side of the face of the peripheral type. Slight weakness remained in the right side of the face, with a tendency to contracture. Taste and smell were unaffected.

The *sensory system* revealed no abnormality except that mentioned in connection with the fifth cranial nerve.

The *motor system* was unimpaired, and the reflexes were normal.

The *cerebrospinal fluid* was clear and colourless, with 8 cells per c mm.; large mononuclears about 5 per cent.; total protein 0.03 per cent.; Nonne-Apelt test, a faint haze; Lange test, no change in any tube. The Wassermann reaction was negative in the fluid and the blood.

The blood culture was sterile after seventy-two hours, as was also the culture from the conjunctiva. A fairly long chained streptococcus was obtained from a throat swab.

Blood Count.

Reds	.	.	.	.	.	.	3,950,000 per c.mm.
W.B.C.s	.	.	.	.	.	.	11,730 per c.mm.
Hb.	.	.	.	.	.	.	85 per cent.
C.I.	.	.	.	.	.	.	1.
Polymorphs	.	.	.	.	.	.	74 per cent.
Lymphocytes	.	.	.	.	.	.	20 per cent.
Hyalines	.	.	.	.	.	.	6 per cent.

Figs. 1 and 2 show the appearance of the patient's face at the time of admission to the hospital. It will be seen that the left side of the face was severely paralysed. The weakness still remaining in the right side of the face can also be observed. Incidentally, it may be remarked that when an attempt

was made to close the eyes, they deviated upward and to the right, instead of straight upward.

During the patient's stay in the hospital considerable further improvement has taken place and no fresh symptoms have appeared.

DISCUSSION.

The cardinal features of the case have been :—

1. Bilateral but asymmetrical inflammation of the eyes in the form of uveitis and conjunctivitis, coupled with the subjective appearance of

FIG. 1.

FIG. 2.

a shadow, first in the right and subsequently in the left eye, in the latter instance coinciding in time more or less with the facial paralysis.

2. Bilateral, symmetrical and synchronous parotitis.

3. Involvement of certain cranial nerves producing right, and later left, facial paralysis ; in addition, there has been slight involvement of the fifth and tenth cranial nerves ; and, finally, it may be hazarded that the slight deafness may have been due to eighth nerve affection, though it is possible that a degree of Eustachian obstruction may have accounted for this.

Negative features of the case have been the absence of definite peripheral neuritis, apart from the cranial nerve involvement, although the paræsthesiæ in the limbs mentioned above may fairly be taken to suggest that the peripheral nerves have not entirely escaped.

An interesting point in the case has been the curious variations of body temperature. Weekly rises took place, the highest of these being 101°F . The temperature charts, over a period of about six weeks, showed a mild resemblance to the charts of a case of Hodgkins' disease published by the late Sir James Galloway.³

There can, in my opinion, be no doubt that this is a case of uveo-parotitic paralysis, falling into the same group as the cases described by Heerfordt and by Feiling and Viner.

The order of appearance of the signs of the disease varied in the cases recorded. The eyes were the first to be affected in Heerfordt's first case, and in the one described by Mackay,⁴ but these two did not go so far as to have peripheral nerve involvement. The facial paralysis appeared to precede the parotitis and uveitis in the case of Feiling and Viner, while, in my case, there were signs of slight peripheral neuritis in the legs before the appearance of the parotid and eye disturbance, although the real nerve involvement took place some considerable time after the parotitis. Other cases had parotitis as their first sign. Thus, in the case of Daireaux and Pechin⁵ the eye affection appeared after the parotitis and two months before the nerves were affected. It can be seen that the cardinal signs of this disease may take place in any order. The occurrence of the facial paralysis three months after the parotitis resembles Dopter's⁶ case, in which the facial paralysis developed two months after the parotitis. Heerfordt mentions a definite prodromal period of general malaise. In my case the period between November, 1922, and January, 1923, might justifiably be included as the prodromal period. In Heerfordt's first case it lasted as long as three months.

The case described resembles that of Feiling and Viner in the involvement of both sides of the face, one side shortly after the other, and in the occurrence of paræsthesiæ. Two of Heerfordt's cases showed dysphagia, as did this case. In one of his, paralysis of the recurrent laryngeal nerve was found on the left side. No rash was seen in this case, as described by Feiling and Viner and by Brewerton.⁷ In Heerfordt's first case optic neuritis was found, but there was no sign of this in mine, nor was there any disturbance of the oculomotor nerves.

Additional features exhibited in the present instance are the slight involvement of the fifth and eighth cranial nerves and the peculiar temperature variations. Heerfordt in one of his cases describes periodic rises of temperature, but does not give any details.

The ætiological factor of this disease must now be considered. We are not dealing with a case of mumps, for the following reasons: the patient had mumps as a child, and, according to a critical review by Feiling,⁸ one attack confers lifelong immunity, inasmuch as he was unable to find an authentic case of a second infection. Again, the patient was not aware of any cases of mumps in the neighbourhood when

her illness began. Further, the interval between the parotitis and the facial paralysis in this instance was considerably longer than what obtains in the case of mumps.*

Mention has been made of the slight similarity between the temperature charts in this case and those of Hodgkins' disease. Certainly several glands were affected, viz., the appendix, tonsils, parotid and ciliary body, but the writer is far from suggesting that this disease has any relation to Hodgkins' disease.

There can be no doubt that uveo-parotitic paralysis is of a toxic or toxi-infective nature, but its cause has not yet been discovered. Bacteriological examinations, in my case, revealed nothing except a long-chained streptococcus in the throat. One cannot accept this streptococcus as responsible for the disease, as it is acknowledged as a fairly common inhabitant of the throat in healthy people.

I wish to express my indebtedness to Dr. Kinnier Wilson for permitting me to publish this case and for many kind suggestions.

* Coureaud and Petges⁹ described facial paralysis in association with mumps, but the face was affected a very short time after the parotid.

REFERENCES.

- ¹ HEERFORDT, *Archiv f. Ophthalmol.*, 1909, lxx, 254.
- ² FEILING and VINER, *Jour. of Neur. and Psychopath.*, 1922, ii, 353.
- ³ GALLOWAY, *Brit. Med. Jour.*, December 23, 1922.
- ⁴ MACKAY, *Trans. Ophthalmol. Soc.*, 1917, xxxvii, 208.
- ⁵ DAIREAUX and PECHIN (quoted by Heerfordt).
- ⁶ DOPTER, *Gaz. des hôp.*, 1904.
- ⁷ BREWERTON, *Trans. Ophthalmol. Soc.*, 1910, xxx, 148.
- ⁸ FEILING, *Quart. Jour. Med.*, 1915, viii, 257.
- ⁹ COUREAUD and PETGES (quoted by Heerfordt).