IRIDOCYCLITIS—PAROTITIS—POLYNEURITIS : A NEW CLINICAL SYNDROME.

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THE case to be described is an example of a clinical picture which appears to be very little known, but is nevertheless one which has been recognized and described in medical literature. Briefly, the syndrome consists of iridocyclitis and parotitis, with or without involvement of either cranial or peripheral nerves, and occasionally accompanied by cutaneous lesions. Whether this syndrome is produced by any specific infection or may occur as the result of several different infective agents is still an open question. In this particular case no definite infective agent could be isolated.

Ida C., a waitress, was admitted to the Hospital for Epilepsy and Paralysis, Maida Vale, on March 23, 1921, complaining of feeling tired and drowsy all day, of a dull aching pain in the back, occasional neuralgic pains in the jaws and neck, and mistiness of vision in both eyes.

History.—She had felt seedy and run down since the beginning of February, 1921. During the first two days of March she felt particularly ill and drowsy, and her back ached. On the evening of March 3 she noticed that her mouth was drawn over to the left side. Two days later mistiness of vision in the right eye was noted, which became progressively worse. About March 12 the left eye became affected as well. On March 10 the mouth was said to have returned to the middle line, but it was felt that the left side of the face was now weak as well as the right. Her sister had noticed a slight swelling in front of the right ear about two days after the paralysis of the right side of the face had occurred; a few days later a similar swelling appeared on the left side of the face. No diplopia nor any weakness of the limbs had been noticed.

On Examination.—The patient appeared a well-nourished and healthy young woman, though somewhat pale.

Eyes.—Both showed much ciliary injection. *Corneæ*: haziness of the deep layers with numerous 'K.P.' Pupils widely dilated, reacting very faintly to strong illumination, inactive to accommodation. No synechiæ. Vitreous full of fine floating opacities. No optic

neuritis. Fundus details not visible. *Tension*: full to + 1 each eye. *Acuity of Vision*: counted figures at 2 metres right eye, at 4 metres left eye.

Both parotid glands were swollen, but somewhat hard, and without any of the usual evidences of active inflammation (pain, tenderness, redness, etc.). A bilateral facial paralysis of the peripheral type was present, more marked on the right side; no affection of taste could be detected. The functions of the other cranial nerves appeared normal. Examination of the rest of the nervous system was negative except that the tendon reflexes in the arms were only obtained with great difficulty, the knee-jerks were very sluggish, and both ankle-jerks were absent. On the sensory side she complained of some numbness in the fingers of both hands, but no actual loss of sensation could be demonstrated. Physical examination of the heart, lungs, and abdomen was quite negative. The urine contained no abnormal constituent.

A definite rash was found on the skin. This was seen on the anterior aspects of both legs and the lower parts of the thighs. The lesions consisted of small erythematous patches, reddish to purple in colour, varying in size from a sixpence to a shilling, not unlike small lesions of erythema nodosum but without the same amount of induration. There was no fever.

An examination of the blood showed the leucocytes to be 5500, with the differential count as follows: polymorphonuclears 65·5 per cent, lymphocytes 28 per cent, large mononuclears 3·2 per cent, transitionals 8 per cent, eosinophiles 3·2 per cent. The cerebrospinal fluid appeared normal to the naked eye; it contained only 15 cells per c.mm., of which all were lymphocytes; the Nonne-Apelt test for globulin was negative, and the percentage of albumin was only 0·015. The Wassermann test was negative in both the blood and the cerebrospinal fluid.

A careful examination was made for any source of infection in the mouth, teeth, throat, ears, nose, and urinary tract, but without any definite result. All the teeth had been removed some years before. The bowels were open regularly, and the stools appeared normal.

On April 4 the condition of the eyes had become worse. The ciliary injection was more marked; 'K.P.' were more numerous. The sclerotic in the ciliary region for a distance of 3 to 4 mm. from the corneal margin was acutely inflamed and raised, forming a ring surrounding the cornea; this was more obvious in the right than the left eye. *Tension* + 1 each eye.

The swellings of the parotids were still present, but were smaller and certainly harder. No other glandular enlargement could be found. The paralysis of the right side of the face was still complete, but on

the left side a certain amount of recovery had taken place ; the left eye could be closed nearly completely, and on voluntary movement the mouth was definitely drawn to the left side ; wrinkling of the forehead was absent on both sides. The signs of polyneuritis in the limbs were more definite. There were sensations of numbness and tingling in both hands, and some unsteadiness in the performance of the finer muscular movements of the fingers. Both supinator- and both triceps-jerks were lost ; the knee-jerks and the ankle-jerks on both sides were lost also. There was no definite atrophy of muscles, no paralysis in the limbs, and no objective disturbance of sensation.

On April 23, and again on April 29, a paracentesis of the anterior chamber of the eye was performed on each eye, to relieve the intra-ocular tension.

By May 9 the general condition had improved, though the eyes remained about the same. The swelling of the parotid glands had disappeared, the paresis of the left side of the face was clearing up, but the paralysis of the right side of the face was still nearly complete ; the tendon-jerks in the arms were still absent, but the knee-jerk on the right side could be obtained. The left knee-jerk and both ankle-jerks were still lost.

By May 30 both knee-jerks had returned, but the supinator-jerks were still absent, as well as the triceps-jerk on the left side. The rash had also disappeared, gradually fading away without undergoing any material change. Steady improvement in the eye was now apparent. A blood-count taken at this date showed : red blood-cells 4,920,000 per c.mm., white cells 9000, hæmoglobin 90 per cent, colour index 0.9 ; so that the slight leucopenia present at the first had disappeared.

Steady improvement in all the symptoms now followed, and on June 20 the following note was made : " The patient is much stronger, walks well, and takes her food well. Eyes : the pupils are unequal, right being dilated and larger than the left. The right pupil does not react to light at all, while the left does so sluggishly ; the intra-ocular tension has diminished. There is still considerable ciliary congestion, especially in the right eye. In the arms the tendon-reflexes are now active. The knee-jerks are both present ; but both ankle-jerks are still lost. There is no subjective or objective disturbance of sensation." On July 1 the patient was able to leave the hospital.

The case may be summed up as showing the following features : (1) Double iridocyclitis of severe degree with cycloplegia ; (2) Double parotitis ; (3) Double facial paralysis ; (4) Some signs of a generalized polyneuritis ; (5) The presence of a rash on the skin.

It is interesting and important to note that the first symptom to attract any serious notice was the facial paralysis on the right side,

which preceded both the inflammation in the eyes and the swelling of the parotid glands. Indeed, the patient was seen by one of us as an out-patient on March 3 and regarded as an ordinary case of peripheral facial paralysis (Bell's palsy). Mistiness of vision in the right eye and some swelling of the right parotid gland were not noticed till at least two days later.

The first question that naturally arises is, Was the case not one of mumps with complications in the shape of polyneuritis and iridocyclitis? We are emphatically of the opinion that it was not a case of mumps, for the following reasons: (1) The appearance of the facial paralysis on the right side, which was severe from the onset, definitely preceded the swelling of the parotid glands by at least two days. (2) The swellings of the glands themselves were quite unlike that of mumps. Instead of the usual rather diffuse swelling of the gland, which is soft, and fills up the hollow between the ear and the angle of the jaw, producing the characteristic facial appearance, this case presented a swelling confined for the most part to the pre-auricular portion of the gland, harder and less noticeable than the ordinary swelling of mumps. The swellings further persisted for much longer than is usually the case in mumps. Other points which render the diagnosis very unlikely are the presence of a leucopenia at the onset of the disease, the absence of a marked lymphocytosis in the cerebrospinal fluid in a case exhibiting such marked signs of nervous involvement, the presence of a double and very persistent facial paresis, and the signs of a polyneuritis.

Facial paralysis has been reported in mumps, but has not been known to precede the swelling of the parotid glands. Couraud and Petges saw 7 cases of facial paralysis in an epidemic of 60 cases. It appeared at a time varying from three to nine days after the onset of the disease, was always unilateral, and lasted from six to ten days only, disappearing completely without treatment: a totally different picture from that presented by the case under discussion. Though a persistence of the swelling of the parotid glands on one or both sides has been reported as a sequel to mumps, it is in the highest degree unusual and may justly be regarded as clinical evidence against the disease. Feiling,¹ in a previous paper, has reviewed the various complications of mumps, and a reference to his paper will confirm the additional reasons given above for regarding the present case as being some infection other than mumps. Mackay² in 1917 published a paper entitled "A case of uveoparotitis with iridocycloplegia", in which he records in detail a case similar in many respects to that here reported, and further summarizes the literature up to that date. Mackay's case was that of a woman, age 30, who first presented herself on March 10 with iridocyclitis and cycloplegia; about a week

later swelling of the pre-auricular parts of both parotid glands appeared; on April 24 it was noticed that the parotid glands were still slightly indurated. His case appears to have shown no nervous complications. He quotes a valuable paper of Heerfordt's entitled, "On a subchronic uveoparotid fever localized in the parotid gland and the uvea of the eye, and especially complicated with paresis of cerebrospinal nerves". Heerfordt³ records three cases of his own which, in addition to iridocyclitis and parotitis, showed signs of involvement of the cerebrospinal nerves in the shape of optic neuritis in one case, transient facial paralysis and some dysphagia in a second, and in the third a right-sided facial paralysis with disturbance of sensation in the skin of the abdomen and hands. His cases did not present cycloplegia. Heerfordt quotes two other cases which he considered to belong to the same group.

Mackay was able to find in the literature since Heerfordt's paper only six cases of dilated pupils and paralysis of accommodation with parotitis; of these it is probable that three followed mumps. Brewerton,⁴ however, in 1910, reported a case which is very similar in many respects to that of the writers, and which is particularly interesting in having also shown a skin rash. It was that of a boy, age 17, whose illness began on Nov. 18 with a rash, supposed to be nettle-rash, on the face and arms, lasting five days; when the rash was subsiding a small lump was noticed in the right side of the face, and on the next day on the left side also. When examined six days later both parotid glands were swollen and very hard; both eyes showed iridocyclitis with keratitis punctata and posterior synechiæ; vision was reduced to counting fingers at two feet. In January he developed a rash which was considered to be an atypical form of erythema nodosum. The affection of the eyes in the writers' case was one of acute cyclitis and anterior scleritis complicated by secondary glaucoma of marked degree, which necessitated frequent paracentesis of the anterior chamber of the eye to relieve the tension. In addition there was almost complete cycloplegia. The condition was probably caused by a generalized toxæmia, and consequently the eye affection was primarily vascular and localized in the ciliary body. In spite of the severity of the inflammation, recovery of sight has been nearly complete and the cycloplegia has entirely disappeared.

In comparing the case with those quoted from the literature it is impossible not to be struck with the similarity presented. The two symptoms common to all are the inflammatory lesions in the eyes (with or without a paralysis of the pupils) and the parotitis. These would seem to be an essential feature of the disease if this syndrome can be raised to the position of a clinical entity. Other symptoms are involvement of either the cerebral or spinal nerves or both in

what must be regarded as a toxic neuritis; in one case at least (the writers') symptoms of this neuritis in the shape of a peripheral facial paralysis preceded all other objective manifestations of the disease. In two cases rashes on the skin have been observed.

REFERENCES.

- ¹ FEILING, *Quart. Jour. Med.*, 1915, viii, No. 31.
- ² MACKAY, *Trans. Ophthalmol. Soc.*, 1917, xxxvii, 208.
- ³ HEERFORDT, *Archiv f. Ophthalmol.*, 1909, lxx, 254.
- ⁴ BREWERTON, *Trans. Ophthalmol. Soc.*, 1910, xxx, 148.