A CASE OF ERB'S JUVENILE DYSTROPHY ASSOCIATED WITH BILATERAL ENLARGEMENT OF THE PAROTID AND SUBMAXILLARY GLANDS.

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THE following case is remarkable for the association with fairly general and pronounced muscular atrophy of an enlargement of the parotid and, to a less degree, of the submaxillary glands. The muscular atrophy, though somewhat anomalous in type, conforms most nearly to Erb's juvenile muscular dystrophy. The swelling of the parotid appears to be a simple glandular enlargement.

The onset of both atrophy and swelling was simultaneous, without any definite cause. The swelling of the parotids has long remained stationary, and the muscular atrophy has during the last year or two apparently ceased to extend. It is difficult to see what connection there can be between the two phenomena, or whether there is any at all; possibly it is a mere coincidence in the same patient of two rare symptoms, but if so I think it is a sufficiently curious one to merit a brief record. In favour of some common cause for the parotid swelling and muscular dystrophy is, of course, the coincidence in the onset of both affections.

Frederick E., aged 18 years, complains of difficulty in walking and getting about, and of wasting of muscles.

Family history good; no other case of the same kind in it, nor of any other form of chronic paralysis. The patient is the eldest of five children, the others are healthy; I have examined them, and found no signs of muscular atrophy or hypertrophy in any.

The patient was born naturally and was quite healthy until 13 years of age. He was an active and very intelligent boy. He

FIG. 1.

FIG. 2.

FIG. 3.

had had no previous illness; the present one began insidiously, without assignable cause. The first symptom noticed was weakness of the legs, which would give way suddenly and cause him to fall down. The muscular wasting did not attract attention until he was first brought to the Bristol General Hospital at the age of 15 years; it had then the distribution described below, and from the fact that weakness began in the legs, and that when he fell he could at first pull himself up by his arms, but later became unable to do this, it is inferred that the wasting began in the lower and subsequently spread to the upper extremities. With the onset of these symptoms came simultaneously swelling of both parotid glands; this took some weeks or (?) months to develop to the present size, and then remained stationary. There was not at any time pain, redness, or tenderness in them, and no intermittent attacks of pain or increased swelling occurred. He has never had mumps. The muscular weakness progressively increased, so that he could hardly get about and could not get up again when he fell. He has never had any pain in the limbs, nor any sensory disturbance, subjective or objective. From the onset of the illness he has ceased to grow in size.

When he first came to the Hospital in 1900, he was just able to stand and to walk, his gait was somewhat tottering; he did not drag his feet. He could get out of bed, but could not get back into it again, and could not go upstairs. When placed on his back on the floor he could turn over, and get on to his hands and knees, but no further. The special senses were normal except that he was deaf; watch was heard in contact (left ear) at one inch; (right ear) he was deaf to the high notes of Galton's whistle. When admitted in 1903 the muscular wasting and general condition appeared to have practically remained stationary, and is described below. The atrophy was perhaps more marked in some of the muscles, but on the other hand he walked better, and did not fall about so much, he was now able to go up a few stairs provided the steps were low, and he was also better able to button and unbutton his clothes.

Present State.—The lad presents a remarkable appearance from the relatively large size of the head, the increased breadth of the facial aspect due to the parotid swellings, and the stunted growth of the trunk and limbs with the pronounced muscular atrophy. The head measures 22 inches in circumference, the parotid swellings give a peculiar broad appearance to the face, and the head is actually broad (14 inches between the mastoids).

The upper lip is large and long, the lower end of the nose broad, and alæ nasi wide. There is some nasal obstruction due to chronic hypertrophic rhinitis, but no adenoid growths are present.

The ears are large and prominent. There is a symmetrical enlargement of each parotid gland, to about the size of a small orange; the swellings are painless, moderately soft, elastic and lobulated. The submaxillary glands are also enlarged, but not to the same extent; the right is the larger. The parotid ducts are patent, the saliva contains KCNS and turns starch into sugar; it is normal in amount, and he does not suffer from dry mouth. *There are no enlarged lymphatic glands.* The eyeballs are somewhat prominent, the upper lids full, especially on the right side, where fulness is due to the enlarged lachrymal gland, which can be plainly felt.

The facial muscles are not wasted, the eye-movements are normal; the pupils equal and act well; the tongue, soft palate, and larynx are normal, and speech unaffected. In the face Chvostek's sign can generally be obtained.

The muscular atrophy in the trunk and limbs is very pronounced and affects in most marked degree the following muscles on both sides: deltoids, triceps, pectoralis major, sterno-mastoid, latissimus dorsi (left more than right), supinator longus (right more than left); in rather less degree, trapezius (right more than left), biceps, long flexors and extensors of wrist and fingers. In the pectorals the lower part of the right and the upper part of the left muscle are chiefly affected; the external and internal rotators of the humerus act fairly well; the supraspinati and infraspinati are not wasted and of good size, the right supraspinatus especially so.

The abductors and adductors of wrists are weak. In the hands the long extensors of the thumb are affected, especially extensor ossis metacarpi, and the thumb is adducted towards the palm; it can be opposed to the little finger. The left abductor indicis, and the hypothenar eminence are wasted. The fingers, owing to predominant action of interossei, are slightly flexed at proximal, extended at distal, phalangeal articulations.

It should be added that though the small muscles of the hands are relatively spared, yet like all the other muscles of the body they are small and poorly developed.

The serratus magnus is small, but its action is normal. The intercostal muscles are much wasted. The action of the diaphragm is normal. The erector spinæ is unaffected.

Of the abdominal muscles atrophy affects the lower two

thirds of each rectus, whilst the upper third is spared; the interdigitations between the sections of the muscle are well shown; the external oblique muscles are much atrophied, the right having almost entirely gone. There is an interval between the recti when they contract, and from the weakness of the obliques the abdominal wall above Poupart's ligament is protruded on coughing.

In the glutei there is marked wasting of the right, slight of the left. The action of the psoas-iliacus is very feeble. In the thighs the quadriceps extensor is much atrophied on each side, especially on the left, the adductors are also somewhat wasted, but the hamstring muscles appear to have escaped. Below the knee there is slight wasting of the anterior tibials and peronei, but the calf muscles have escaped. The feet are flat, with prominent internal malleoli of the ankle, and the feet cannot be dorsi-flexed beyond a right angle. The muscles show no fibrillary tremors.

On electrical stimulation the affected muscles show a diminished response, in proportion to the degree of atrophy present, to both forms of current; the most affected do not respond at all. There is no qualitative change, nor R.D.

Reflexes.—Tendon reflexes all absent. Superficial reflexes, epigastric, abdominal and cremasteric present; plantar difficult to obtain, of flexor type.¹ No alteration of sensation; no affection of special senses except that he is now totally deaf. No affection of bladder or rectum. Thoracic and abdominal organs normal.

The arrested growth of the boy has been noticed above, it affects all parts of the body, and the small size of the bones, which is, of course, most obvious in the long bones, is perhaps especially worthy of remark, as exemplifying the general arrest of growth which seems to have been coincident with the onset of muscular atrophy. The lad of 18 looks like a boy of 14 or 15. Intelligence is good.

Mr. C. H. Walker kindly took skiagrams of the long bones; nothing abnormal was observed in the process of ossification, and the skiagrams are therefore not reproduced. I am indebted to Dr. Newman Neild for the photographs.

¹ When the patient was in London, Dr. Batten, on examining him after a bath, found an extensor plantar reflex. On subsequent examination this has been noted once or twice since, but sometimes the reflex is not obtained at all.