

Abstracts of British and Foreign Journals.

Landouzy and Déjérine on Progressive Atrophic Myopathy involving the Face. (*Revue de Médecine*, Dec. 1886.)—The authors have devoted much time and trouble to the very careful and minute investigation of the various types which are to be found in the disease which is commonly called Progressive Muscular Atrophy (*Comptes-rendus de l'Académie des Sciences*, Paris, 7 Janvier 1884. *Revue de Méd.*, Feb.-April, 1885). They consider that the types, at least which they describe, belong to a disease beginning "in" the muscle and not in the spinal cord, a conclusion they wish to indicate by calling the disease a progressive atrophic myopathy, instead of a progressive muscular atrophy. The types which they claim previously to have established are: (1) the facio-scapulo-humeral type, and (2) the scapulo-humeral; of which the second was believed to be much the more common, and the first was considered to belong to only a very few cases originating in childhood, such cases, in fact, as Duchenne had called Progressive Muscular Atrophy of Children. The chief points they wish to add to what they have said before are, that the affection of some facial muscles is much commoner than was supposed, that it consists chiefly in thickened lips, the upper generally overhanging, along with weak and unequal movements of the corners of the mouth and of the cheeks, and an incapacity to shut the eyelids completely, which gives this *facies myopathica* a heavy look and clumsy movements more easily recognised than described. The pronunciation is very indistinct, especially of the labials. And this *facies* they regard as a factor, which may come first in the symptoms of the disease either in children or adults, or may come on gradually late in the disease, or may possibly never enter into it at all. The muscular lesion which manifests itself by atrophy they consider as of the same nature as that which manifests itself by pseudo-hypertrophy; they both originate, as the most modern researches are taken to show, in a muscular irritation. That the clinical results of atrophy and pseudo-hypertrophy should be so distinct is a parallel to the cases of interstitial hepatitis, which are sometimes from beginning to end atrophic, and sometimes from

beginning to end hypertrophic. They bring forward six new cases in full detail, and in one of these was an autopsy. The first subject was a man of 36, whose grandmother, mother, and brother were atrophic, and in whom the inherited *facies myotrophica* was plain from childhood. His playmates called him a "Chinaman." He could not keep his place in a military band, because there was no chance of his ever learning to play the flute—his lips were too clumsy and hypertrophied. His scapulo-humeral symptoms on the left side were well developed at the age of 15. He was allowed to stay in the army till after the Franco-German war, when he was 21, though he only pretended to shoot from the left shoulder, and after that could do very little. He was treated in the Charité in 1885. By good fortune there were photographs of his family and himself, which showed the same type of face and atrophy in them, and the progress of the same disease. The photographs of his own case are admirably reproduced in the *Revue*. The second case was in a woman æt. 27, who came at first to the Hospital, not imagining she was ill herself, but entirely as interpreter for her paralysed mother, and in her the disease was recognised from her face only. She seemed entirely unconscious of any abnormality either in the face or the arms, but on examination the ordinary scapulo-humeral type of atrophy was found; and photographs showed that her facial symptoms had existed from childhood. She could not whistle, or blow out a candle, or frown, or shut her eyes completely. Her lips were thick, and moved very little in speech or laughter; there was reaction of degeneration in the *orbicularis oris*.

The third subject was a man, born in 1856, in whom some affection of the face was noticed when he was three years old. Some atrophy of the muscles of the trunk and legs came on two years later, and very gradually spread to the arms. When he was nine he was taken to see Duchenne at Boulogne, who was very much interested in the boy, as illustrating that rare form of disease which he had then (1868) named atrophic paralysis of children. He published an account of the case (*De l'Electrisation localisée*, 3^{me} éd. 1872, p. 1098), fully describing the facial symptoms mentioned in the other cases, except the inability to close the eyes completely; he illustrated them by photographs, and found similar symptoms in his mother and his brother. A year or two later the flexors of both legs began to contract, and he became a hopeless cripple. He could use his hands, however, and made a scanty living by being dragged about France in a go-cart by an idiot, and selling little

books of instruction in shorthand writing. At last the idiot died and he came into Hospital under the care of M. Landouzy in 1886, showing the same *facies myopathica*, of which Duchenne had published a description 18 years before, with the additional inability to close the eyes completely which had not been noticed previously. The atrophy in the arms and neck were much more advanced. In the 4th case there was no history of heredity; atrophy of a scapulo-humeral type came on *æt.* 40; and no affection of the face was noticed till he was 45. In the 5th case there was probably some complication with lead-poisoning. He was a man of 40, who had had dropped wrist in 1872, and in consequence given up his trade as painter; but in 1885 was found to have progressive atrophy of scapulo-humeral type, with the addition of facial symptoms in 1886 and notable atrophy of the tongue. The 6th and last case, in a man of 66, was fatal, and was examined very minutely. At the age of 20 he began to notice atrophy about the shoulder, which spread very slowly into the arms and legs. He was a hawker, and could go on with his trade till he was 63. There was no visible affection of the facial muscles whatever; no fibrillar contractions; no reaction of degeneration; no abnormality of knee-jerk and no hereditary history. He died of pulmonary-tuberculosis, and an autopsy showed no abnormality of the spinal cord, of the cervical sympathetic, of the anterior roots of the peripheral nerves; but simple atrophy of the muscular fibres with great multiplication of the nuclei. In some muscles a few fibres were hypertrophied, and in the most atrophied muscles there was some interstitial fat. The point on which the authors lay most stress is that, though they could not detect abnormality of the facial muscles in life, they found after death that they were degenerate or beginning to degenerate, a point they consider of importance as showing that, sooner or later, the face becomes involved in this progressive atrophy. Westphal has also published three cases of progressive muscular atrophy with affection of the ocular muscles (*Charité-Annalen*, Berlin, 1886), and he notices the thick, overhanging upper lip, "like a tapir's," in one of them.

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