Progressive Lenticular Degeneration.

Under the title "Progressive Lenticular Degeneration: a Familial Disease Associated with Cirrhosis of the Liver," S. A. K. Wilson (Brain, 1912, p. 295) describes a new disease characterised by strikingly distinctive clinical and pathological features. The picture presented in this elaborate paper has been drawn from six cases collected by the author from the literature and reported by Gowers, Ormerod, and Homén respectively from four cases personally observed by him, three of which were examined pathologically in an exhaustive manner, and from the unpublished records of two further cases.

Progressive lenticular degeneration is defined as "a disease which occurs in young people which is often familial but not congenital or hereditary. It is essentially and chiefly a disease of the extra-pyramidal motor system, and is characterised by involuntary movements usually of the nature of tremor, dysarthria, dysphagia, muscular weakness, spasticity and contractures, with progressive emaciation. With these may be associated emotionalism and symptoms of a mental nature. It is progressive, and after a longer or shorter period fatal. Pathologically it is characterised predominantly by bilateral degeneration of the lenticular nucleus, and in addition cirrhosis of the liver is constantly found, the latter morbid condition rarely, if ever, giving rise to symptoms during the life of the patient."

The following is a more detailed account of the salient features of the disease as depicted by Wilson in his admirable monograph from a study of the twelve cases above referred to:

Etiology.—The average age at onset was 15, the youngest patient being 10, the eldest 26. Seven of the cases were males and 5 females. In no case was there a definite history of a neuropathic heredity, although 8 were familial cases—in two instances three, in one other
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two—belonging to the same fraternity. Race appears to be of no importance. There was no evidence of syphilis, alcohol, or other predisposing cause, while exciting causes were uniformly absent.

Symptomatology.—Three cases may be referred to as acute or subacute, terminating fatally in from four to thirteen months with a high irregular temperature and rapid emaciation. The remaining nine ran a more chronic course.

Involuntary movements and tremor were striking features. The tremor, which was one of the earliest symptoms, and was usually found at the outset, was rhythmical, and varied in rate from five to eight per second. It was increased by excitement and by voluntary effort. In some cases tonic or clonic spasms were observed. Spasticity was noted in all the cases, the only muscles escaping being the intrinsic muscles of the eye. The face was fixed and expressionless. In most cases the flexor muscles were predominantly hypertrophied. Contractures were a constant feature. Dysarthria and dysphagia were present in every case, and in the advanced stages there was more or less complete anarthria. Muscular weakness was present, but even in the later stages a considerable degree of voluntary power remained, e.g. a patient whose appearance suggested complete spastic paralysis was able to walk. Sensory symptoms, apart from occasional pains, were absent. The tendon jerks were unaltered. The plantar reflexes were of the flexor type. Impairment of the organic reflexes sometimes occurred towards the end, but it is doubtful to what extent this may not have been attributable to the patient’s mental state. Mental impairment was noted in at least 8 of the 12 cases. This would appear to take the form of an inability on the part of the patient to “add much to his store of mental images, while his capacity for retaining impressions is impaired.” A degree of facility, docility, and childishness was noted in advancing cases. The optic discs were normal, as were the pupillary reactions. There was no nystagmus. The palate moved on phonation and reflexly. There was no fibrillation or localised amyotrophy; there were no cerebellar symptoms; there was no impairment of sensibility. The reflexes were not those of pyramidal disease.

Pathology.—Ten of the cases were examined after death, and in seven of these symmetrical bilateral lenticular degeneration was found. Of the other three cases one was not examined microscopically, while in the remaining two the condition of the corpus striatum was not specially referred to. The striking feature is the limitation of the changes to the lenticular nuclei. In the majority of cases there have been changes obvious to the naked eye. Thus the lenticular nucleus may appear discoloured or spongy, atrophied or completely disintegrated and excavated. Microscopically the change is seen to consist of glial overgrowth, the glia tending to disintegrate and break down.
There are no signs of obliterator endarteritis or other pathological processes in the walls of the blood-vessels. The liver is always in an advanced state of cirrhosis. The cirrhosis is more or less multilobular, but in some places monolobular. In several of the cases the spleen is reported to have been enlarged, while regressive and proliferative changes have been found in the thyroid gland.

The nature and pathogenesis of the disease offer problems of engrossing interest. Wilson is of opinion that the morbid agent is probably of the nature of a toxin, which toxin is probably associated with the hepatic cirrhosis and generated in connection with it. The toxin must have a selective action.

Diagnosis.—This seems to present no difficulty to one who is familiar with the clinical picture. Pseudobulbar palsy alone perhaps calls for special notice. It is indeed conceivable that the pathological process may actually implicate the genu of the internal capsule on either side, and so produce pseudobulbar symptoms. In the typical pseudobulbar cases the palate and tongue are paralysed, whereas in this affection movement occurs. Again, in pseudobulbar palsy there are usually symptoms indicative of a double hemiplegia, with the alterations in the reflexes met with in affections of the pyramidal tracts.

Duration and Progress.—The disease runs a progressive course. In 3 cases death occurred in four, six, and fifteen months respectively. The shortest of the chronic cases lasted two and a half years, and the longest seven years.

Myotonia.

Myotonia congenita, a very rare disease, was first described by Thomsen, himself a sufferer from it, in 1876. Little has been added to our knowledge of Thomsen’s disease, as it is now generally termed, during the past thirty-five years. The only cases reported in this country are those recorded by Buzzard, Hale White, Dreschfeld, and Risien Russell. Wardrop Griffith (Quart. Journ. of Med., January 1912, p. 229) describes in detail the clinical histories of two new cases, and refers to some interesting observations he has made in relation to the myotonic condition of the muscles. As is well known, the striking clinical feature of the disease is this myotonic state, which is characterised by a tonic contraction which prohibits the relaxation of the voluntary muscles when they are first brought into action, the retarding influence soon passing off if the movements are persisted in. Griffith found that when the patient’s muscles were in a state of very complete relaxation, secured by lengthened repose, passive movements of the limbs carried out with great gentleness did not result in the muscles becoming rigid. Again he observed that there was a distinct difference as regards the myotonia, according as the voluntary effort was prolonged or brief. When, for instance, the patient closed his
hand firmly, no matter how short that voluntary effort might be, it required as a rule fifteen seconds for extension of the fingers and hand to be completely effected. After the first closure of the hand succeeding closures were followed by more and more rapidly carried out acts of extension, until the movements became normal in every respect. When, however, the patient was directed to grasp with his hand firmly and keep it tightly closed for some time it was found that extension could then be effected as rapidly as in a normal individual. As the result of many observations it was found that if the firm contraction was kept up for fifteen seconds the act of extension could be carried out normally. The same observations were noted in the case of most of the muscles and movements of the body, and, generally speaking, with similar results. The finer movements of the hands and fingers and the interosseus muscles were shown to be affected contrary to the statements of some writers. The muscles of the eyeballs were not involved, but the levator palpebræ presented the myotonic phenomenon. Thus when the patient was asked to look forcibly upwards and then downwards it was noticed that the upper lid did not follow the eyeball in its descent. It is of interest to note that although the latissimus dorsi muscle was thrown into brief but strong contraction by getting the patient forcibly to adduct the arm it became stiff like the other muscles of the body, but when the patient coughed it was noticed that the contraction associated with this act was not followed by any undue prominence of the muscle.

"Myotonia Atrophica and Hypertrophica" is the descriptive title which Griffith applies to a third case reported in this communication.

The patient, a man of 48, had a vacant expression. He had complained recently of diplopia, although no weakness of the ocular muscles was detected at the time of examination. The levator palpebræ exhibited the myotonic phenomenon above referred to. Mastication was weak, and the relaxation of the masseters and temporals slow. There was some difficulty in swallowing, and the patient could not protrude the tongue beyond the teeth. Speech was slow and indistinct. The vocal cords were in the cadaveric position. Both sternomastoids were greatly wasted. There was no wasting of the other muscles of the shoulder or upper arm. There was well-marked wrist-drop, with wasting of muscles on the back of the forearm. On grasping with the hand there was a general contraction of all the muscles of the forearm, with slow relaxation, which appeared to be more rapid when the patient ceased to grasp than when he exerted a powerful muscular effort in so doing. In some of the muscles the faradic excitability was diminished, while in all the contractions obtained on faradic stimulation remained for some few seconds after the cessation of the stimulus. Dorsiflexion of the foot was poorly carried out; plantar flexion against resistance was followed by a
lasting contraction of the calf muscles. The calves were exceptionally large, measuring 14½ inches. The reflexes were normal. There was no disturbance of sensation. Curious attacks occurred while the patient was under observation. These were preceded by a peculiar feeling in the lower part of the neck, which passed upwards. The arms were then shot out from the sides and the legs extended, the condition lasting a few seconds. Consciousness was not impaired in these attacks. The pulse-rate was usually slow, falling on more than one occasion to 40 per minute. There was no heart block. Pieces of muscles were removed for histological examination. Many of the muscular fibres were found to be greatly increased in diameter. In the affected forearm many of the fibres were almost circular in outline. There was, too, a pronounced increase in the number of nuclei within the muscle fibres. To sum up, this patient presented in addition to the myotonic condition of many of the muscles an atrophic palsy, the distribution of the muscular wasting and notably the pronounced atrophy of the sternomastoids bringing it into line with some of the cases reported by Batten and Gibb (Brain, vol. xxxii. p. 187) under the name of "Myotonia Atrophica," as originally suggested by Rossolimo. Batten and Gibb take up the position that the feature of the atrophy in some of these cases is similar to that met with in the myopathies. They suggest "the possibility of there existing a form of myopathy differentiated clinically from the classical types as much by the distribution of the wasting as by the presence of myotonia." The abstractor some months ago demonstrated at a meeting of the Medico-Chirurgical Society a case of myotonia atrophica in which the infraspinati muscles were abnormally well developed, and indicated that in his opinion this observation afforded support to the view advanced by Batten and Gibb. So far as he is aware this case reported by Wardrop Griffith, a case described by Godwin Greenfield (Rev. of Neurol. and Psychiat., 1911, p. 171) in which the infraspinati were said to have been "unduly prominent," and the personal observation above referred to are the only instances in the literature in which myotonia, muscular atrophy, and hypertrophy have been associated.

SURGERY.

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DEVELOPMENTAL ADHESIONS AFFECTING THE LOWER END OF THE ILEUM AND THE COLON.

In a paper with the above title (Aberdeen, The University Press) Gray and Anderson illustrate and discuss the origin, symptoms, and treat-