FROM THE ARCHIVES


As a contribution to the section of Brain instituted by the founding editors in which articles published elsewhere are abstracted, Dr W.J. Dodds is charged with reporting ‘On the influence of the central nervous system on the irritability of the motor nerves’ and ‘A new theory of locomotor ataxia’ together with summaries of cases of ‘Progressive muscular atrophy’, ‘Multiple cerebro-spinal sclerosis’ and ‘Progressive amyotrophic bulbar-paralysis and its relations to symmetrical sclerosis of the lateral columns’, the latter reported by (Ernest von) Leyden [(1832–1910): Archiv für Psychiatrie (1878; vii. p. 641 et seq.)]. First, Dodds rehearse the earlier description of Bärwinkel (nk) who has qualified the position taken by (Guillaume-Benjamin-Amand) Duchenne [de Boulogne (1806–75)] that the condition is a peripheral affection of muscle, by demonstrating normal faradic irritability of the weak muscles, thereby implying that the lesion is central and in the medulla. (Armand) Trousseau (1801–67) has studied the pathology and finds fatty degeneration in the roots of the hypoglossal, vagus and facial nerves. (Jean-Martin) Charcot (1825–93), Duchenne and (Alex) Joffroy (1844–1908) have added to atrophy of the grey matter of the anterior horns and disappearance of their multipolar ganglion cells, pathological involvement of the motor strands of the spinal cord. Leyden describes the pathology of five cases: unequally distributed muscular atrophy most marked in the tongue, lips, hand and forearm; degeneration of the motor nerves throughout their course with preservation of the ‘nerve-sheath’; fatty degeneration and sclerotic atrophy resembling Wallerian degeneration; degeneration of the large ganglion cells of the anterior horns particularly in the cervical enlargment and medullary nuclei, especially the hypoglossal; degeneration of the pyramidal strands and anterolateral columns throughout their course below the pons with reduction in the number of nerve fibres and diameter of those that have survived; and no involvement of the posterior columns, posterior nerve roots or dorsal horns. ‘The distribution of the principal lesion is remarkable: it is symmetrical and, though widespread, limited to the motor cells and fibres of the spinal cord and medulla oblongata… it is an example of a lesion affecting a particular physiological system of fibres’. The disorder originates in the nerve cells, at different sites, and spreads along the fibres connected with them. Changes in the neuroglia are secondary. Leyden and Charcot differ in their formulation on the sequence of events: for Charcot, atrophy precedes paralysis, whereas Leyden considers that the paralysis is secondary to atrophy; for Charcot, the paralysis is spastic and rigid with contractures resulting from sclerosis of the lateral columns, but for Leyden, it is ‘paralysis with relaxation (atonic paralysis)’.

Dr (Charles Edward) Beevor (1854–1907) has been reminded by Dr (Sir William) Gowers (1845–1915) of a case diagnosed as amyotrophic lateral sclerosis that Beevor encountered as a resident at the National Hospital, Queen Square, when he worked for Dr (Jabez Spence) Ramskill (1824–97) ‘in which an increased tendon-reflex of the muscles of the lower jaw was obtained’. At the age of 46 years, a woman notices numbness of the face and left arm, which rapidly becomes paralyzed, followed by difficulty with speech and swallowing and weakness of the other upper limb: examination shows inability to swallow with nasal regurgitation due to palatal paralysis; weakness of the face and jaw muscles with inability to protrude the tongue; speech reduced to a moaning sound; and patchy weakness and wasting of the muscles of both hands and forearms but with ‘rigidity’ and a brisk response to ‘filipping the lower end of the radius on each side’. ‘The symptom which I think the most interesting… is… clonus of the lower jaw…[elicited] by placing the finger on the teeth of the lower jaw and then depressing it, when immediately the muscles which closed the jaw passed into a state of clonic contraction and the lower jaw vibrated… in the same manner as the heel… when ankle-clonus is produced’. Clonus can also be produced by stretching the masseters and percussing the zygomaticus major resulting in retraction of the corner of the mouth. ‘I do not know whether a similar case has been observed, but I do not remember to have heard of any one having been reported’. Then, Dr Beevor ruminates on whether chattering of the teeth with cold or rigor is the same as clonus of the jaw and associated with increased tendon reflexes throughout, and he plans to test this casual observation in the next case of rigor he sees. He has noticed that his own knee-jerk is increased after a cold bath, although he has not managed to elicit ankle clonus, ‘though I have heard on good authority of its occurrence in a healthy individual under such conditions’.

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Beevor knows nothing more of the fate of the patient with jaw clonus after she returns home.

As editor of *Brain*, Dr (Armand) de Watteville (1846–1925) feels entitled to add a note on the phenomenon, of which neurologists generally seem ignorant, whereby a reflex can be elicited by tapping the jaw, analogous to the knee-jerk, and which he would like to call the ‘jaw-jerk’ rather than the more clumsy ‘mandibular (or masseteric) tendon-reaction (or reflex)’. His preferred method is to slip a tongue depressor or paper knife between the canine or pre-molar teeth and knock this with a ruler, slim volume or tendon hammer, whereupon there is a clenching of the teeth felt by a finger inserted into the mouth on the opposite side. When exaggerated, nothing more than a tap with the finger is needed to elicit a brisk response. ‘I feel bound to state that this was quite familiar to me before it came to my knowledge that the phenomenon had been observed in a case in America…’

Dr (Joseph Arderne) Ormerod (1848–1925) argues that among the most distinctive aspects of amyotrophic lateral sclerosis is the clinical combination of atrophic paralysis with spasticity in the limbs and pathology which offers an excellent example of a “systematic” or “tract” disease. Charcot can be credited with making the distinction from other forms of paralysis described by (Jean) Cruveilhier (1791–1874) and by Duchenne and (François Amilcar) Aran (1817–61)—in which progressive atrophy with fibrillary twitching in the upper limbs, not limited to the ‘district of particular nerves’, and without sensory phenomena also occurs—by the more rapid progression and poor prognosis, paralysis with contractures of the limbs and bulbar involvement. A typical case, affecting a man aged 32 years, is described: presentation with weakness, wasting and fibrillary tremors of the tongue, lower face and throat spread to the neck, chest and limbs, with death usually within 3 years due to food entering the air passages and causing pneumonia. The pathology combines degeneration of motor nuclei and the anterior horn cells together with the pyramidal tracts from the cervical enlargement through the medulla up to the pons, but ‘as we shall see… these are exactly the lesions which are found in amyotrophic lateral sclerosis’.

However, are there any genuine differences between the disorders described by Leyden and Charcot? For Dr Ormerod, some patients with Charcot’s ‘amyotrophic lateral sclerosis’ may present with the typical bulbar symptoms of ‘Leyden’s disease’; paralysis and atrophy progress pari passu in Leyden’s cases, whereas, as Dr Dodds has noted, Charcot describes weakness followed by muscle wasting. Charcot insists on the presence of spasticity, whereas for Leyden, ‘spastic symptoms, such as have attracted so much attention lately, are absent in our type of the disease’, and if contractures do occur, these are not owing to ‘irritative phenomena’ but, rather, to ‘habitual posture of the paralysed limbs’. Charcot has challenged Leyden on this point and suggested that ‘those most delicate tests… were not properly investigated by him’. Although Dr Ormerod concedes that the state of the tendon reflexes is a genuine point of difference between Leyden’s disease and Charcot’s amyotrophic lateral sclerosis, his explanation is that, starting in the lateral columns, the pathology may sometimes spread preferentially to the anterior cornua and grey matter of the medulla and spinal cord allowing atrophy to dominate the clinical picture or, alternatively, remain confined to the motor tracts during the initial phase of the clinical course. Questioning why the atrophic form usually manifests first in the bulbar musculature and the spastic type in the limbs, Dr Ormerod’s survey of published cases allows the conclusion that: ‘there are intermediate cases which tend to bridge over the gap between Charcot’s and Leyden’s type… [showing]… that clinically they are, what their morbid anatomy indicates, varieties of one and the same disease’.

The aetiology is completely unknown; familial cases are described only in childhood onset disease, the course predictably progressive and invariably fatal and the diagnosis rarely confused with other neurological disorders. But, mainly, Dr Ormerod wants to consider the morbid anatomy of the disease. The muscles are atrophic with reduction in the number and size of fibres showing granular or fatty disintegration, multiplication of the sarcoclemmal nuclei and interstitial fatty infiltration. The motor nerves are thin and grey and conspicuously different from sensory fibres, especially where both are present in the same trunk. In the spinal cord, the focus is on cells of the anterior horn: ‘there is reason to think that the disease begins in the cells’ with subsequent selective involvement of the direct and crossed pyramidal tracts. Although reminiscent of the degeneration that follows a transverse lesion of the spinal cord or bilateral cerebral damage, there are no such lesions in these cases. Some spread of the disease process may occur away from the pyramidal tracts, but this can be attributed to local irritation from the diseased anterior cornua and...
descending motor pathways. It is clear that the absolute sparing of cerebellar and sensory pathways distinguishes amyotrophic lateral sclerosis from other forms of myelitis or spinal cord disease. The pathological processes, confined to the motor pathways and nuclei, identify amyotrophic lateral sclerosis (and Leyden’s disease) as disorders of a defined system.

Usually, but not invariably, the disease stops at the level of the pons. However, (Otto) Kahler (1849–93) and (Arnold) Pick (1851–1924) have described a case in which the clinical course during life was typical of bulbar and spinal amyotrophic lateral sclerosis; at autopsy, in addition to the expected abnormalities in the pyramidal tracts and anterior cornua, there was marked atrophy of the central and precentral cerebral sulci. Although, in this instance, the association of regional cerebral atrophy and disease of the lower motor tracts is inferred, the case described by (Aleksei) Kojewnikoff (1836–1902)—a man aged 29 years with progressive spasticity of the legs and wasting of the arms without bulbar involvement who died of phthisis 3.5 years after onset—has shown, in addition to atrophy of the anterior horn cells, granular bodies in the sclerosed pyramidal tracts that can be used to trace the pathology through the pons into the cerebral peduncles and internal capsules radiating thereafter like a fan into the upper part of each frontal convolution but without apparent damage to the grey matter. Charcot and (Pierre) Marie (1853–1940) have also described cases with granular bodies and destruction of large pyramidal cells in the cerebral cortex: two women, each aged 60 years with the typical history of amyotrophic lateral sclerosis, including prominent bulbar involvement, and both dying within 1 year from onset, have pathological changes in the pyramidal tracts and anterior cornua, as expected; in addition, granular bodies that mark the diseased pathways are seen in pyramidal tracts within the internal capsule radiating into the frontal and parietal convolutions, where the giant pyramidal cells are much reduced in number and survivors morphologically abnormal.

Does Dr Ormerod anticipate the current link between TAR DNA-binding protein 43 (TDP-43)-associated fronto-temporal dementia and motor neuron disease in concluding that: ‘much interest attaches to the morbid anatomy of these cases, as showing, that in the cerebrum as in the cord the disease affects the elements of the motor tract; and secondly, that it may affect any level of the motor tract, the cells of the cortex cerebri, the white substance of brain or cord, the anterior corneal cells of the cord, the nerve trunks, or the muscles’? Together, these authors highlight the points made by Martin Turner and colleagues in their occasional paper on descriptions of amyotrophic lateral sclerosis in the 100 years before 1950 (see page 2883)—young age at onset and predominant bulbar involvement.

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