*Ataxo-Spasmodic Tabes (Ataxic Paraplegia), occurring in a case of Primary Dementia.** By R. S. STEWART, M.D., Senior Assistant Medical Officer, Glamorgan County Asylum.

It is only within comparatively recent times, especially in England, that a distinct place in the nosological classification of diseases of the spinal cord has been granted to ataxic paraplegia, and that it has been separated, on the one hand, from spastic paraplegia, and on the other from ataxic tabes. Ross† classifies it as a compound form of lateral sclerosis, and Bramwell‡ mentions it as owing its origin to an occasional extension of the lesion from the postero-external columns in locomotor ataxia, while Erb (Ziemssen's "Cyclopædia") regards it as tabes complicated by lesion of the lateral columns, or lateral sclerosis complicated by lesion of the posterior columns, according to the preponderance of the symptoms of one or other disease. On the other hand, the most recent English work on diseases of the spinal cord, that of Gowers,§ devotes a separate section to the consideration of the affection, while on the Continent, especially in Germany and France, it has attracted considerable attention. In the "Archives de Neurologie" for March, May, and July of last year, a detailed description of the symptomatology, pathology, diagnosis, and treatment, with a tabulated *résumé* of 33 cases followed by autopsy, described by various French and German authors, is given by Grasset.

The following case, both from a clinical and pathological point of view, presents many of the features of this form of disease of the spinal cord.

Summary:—History of intemperance in the father. Commencement by speech-embarrassment and mental enfeeblement. Ataxic gait. Absence of knee-jerk. Retention of superficial reflexes. Partial anæsthesia. Absence of lightning pains. Gradually advancing motor failure. Rigidity of limbs. Fibrillary tremors. Emaciation. Muscular atrophy. Bedsores. Diarrhœa. Increasing loss of con-

* It may be questioned whether these terms, the introduction of which into our nosology is of dubious wisdom, are justified by the case here reported. It is not yet, we think, sufficiently recognised, how frequent is the combination of lateral and posterior column changes in General Paralysis. [EDS.]

† "Treatise on the Diseases of the Nervous System," 2nd Edit., Vol. ii., p. 80.

‡ "Diseases of the Spinal Cord," 2nd Edit., p. 224.

§ "Diseases of the Nervous System," Vol. i., p. 341.

sciousness. Temporary improvement. Affection of taste and smell. Auditory hallucinations. Returning loss of consciousness; coma; death 16 months from commencement. Degeneration and atrophy of nerve-cells of cerebral cortex and spinal cord. Primary lateral and posterior spinal sclerosis.

James B., aged 24, a smith's striker, native of Cardiff, was admitted on Nov. 25th, 1885. Up to within 12 months of his admission, according to information given by his wife, he had been an active, steady man, kindly-dispositioned, of temperate habits, and of uniformly good health. At that date he had been unable for some time to obtain employment, and it was observed that he was becoming dull and reserved, that his speech became slow and hesitating, his movements uncertain, and that his memory began to fail. During the 12 months prior to his admission these symptoms became gradually more and more marked. Very little information could be obtained as regards family antecedents, beyond the fact that his father was an habitually intemperate man. Patient himself had been married two years, and there was one child.

His condition on admission was as follows:—

He is a poorly-nourished man, pale and sallow, and of medium height; height 5ft. 4½in., weight 9st. 4lbs.; features emaciated, head well formed and amply developed anteriorly; hair dark; irides light blue. The pupils are much dilated, but equal and responsive to light. The tongue is pale, flabby, indented at the edges, and slightly coated, and voluntary attempts to protrude it take place in a highly tremulous and uncertain fashion. Speech is also very hesitating and drawling, amounting to little more than mere mumbling. There is nothing noteworthy as regards the heart, lungs, or abdominal viscera; urine, specific gravity 1011, acid reaction, straw colour, mucous sediment, no albumen.

The mental condition is one mainly of stupor; his expression is vacant and unintelligent; to many commonplace questions he is unable to give a rational reply, though he responds to such simple requests as asking him to put out his tongue, to walk a certain distance, &c.; memory both for remote and recent events is very much impaired, and his habits are defective.

His gait, though by no means characteristic, approximates to that of locomotor ataxia rather than that of spastic paralysis. The knee-jerk is completely absent on both sides; the plantar and other superficial reflexes are normally active. Sensation as regards painful impressions is very much blunted, and the same applies to the localization of touch, and the discrimination by touch of different objects or parts of objects—such as the head from the point of a pin. There is considerable diminution of voluntary motor power, and some ataxia, both of the lower and upper limbs, manifested in the walk and such actions as touching

the tip of the nose with the fore-finger. There is likewise swaying of the body when the eyes are closed and the feet approximated. In both upper and lower extremities there is considerable rigidity, and resistance to passive movement. As he lies in bed the legs are strongly flexed and adducted, and the arms flexed and closely applied to the chest wall. Fibrillary tremors very generally distributed, and affecting the superficially placed muscles, are noted.

Three weeks after admission, owing to increasing helplessness and stupidity, he had to be confined to bed. Consciousness became more and more involved, until he became almost comatose. He lay on his back all day unless moved, the saliva dribbling from his open mouth. He paid no attention to remarks addressed to him, nor could he be roused by vigorous slapping of the face. Evacuations were passed in bed, and there was considerable paralysis of deglutition.

On January 12th it is noted as follows :—There has been a considerable change for the worse. He is still very confused and stupid; he has become very emaciated, the muscular masses are much atrophied, and bedsores, dry, superficial, and leathery in character, have formed over the sacrum and left trochanter (a water bed has been in use all along). The knee-jerk is still absent; the plantar reflex active. He suffers from an intractable form of diarrhœa, not yielding to large doses of bismuth, but controlled to some extent by a combination of tincture of opium and aromatic sulphuric acid.

February 2nd. --A considerable improvement is indicated by the note made at this date. He has become bright and observant, noting what is going on around him. On testing the special senses, it is found that hearing is normally acute, but that taste and smell are both affected, more so the latter. For example, quassia tastes "sour," acid "sweet," sugar "sweet," and salt "salty;" oil of cloves smells "like gin," turpentine "like rum," and assafœtida "like cocoa-nut." Auditory hallucinations have lately developed; he hears his father and mother outside, and he often holds conversations with them. Although he has begun to gain both flesh and strength, a loss of 31 lbs. has taken place since his admission (three months). The eschars exhibit healing action and are improving rapidly. The appetite improves, and he takes large quantities of food without any apparent difficulty as regards swallowing, while the diarrhœa has quite disappeared. This improvement has taken place during the administration of the opium and sulphuric acid, and to these are added cod-liver oil and Parrish's syrup.

In the early part of March he was able to be up part of each day, but by the middle of the month he was again confined to bed. There he remained, and the further progress of the case was steadily and progressively downward. Emaciation and muscular

wasting became extreme; his face became haggard and ghastly, and for two days prior to his death, which occurred on March 31st, 1886, a little over four months after his admission, he gave little sign of life beyond slow regular breathing and a feeble pulse.

The autopsy was performed 40 hours after death, and the following notes were taken.

The spinal cord weighs 17 drams, and its measurements are as follows :—*

	Transverse.	Sagittal.
Cervical	13	10
Dorsal	10	8
Lumbar	10	9 mm.

The cerebro-spinal fluid is in considerable excess. The cord itself is firm throughout, but more especially so in the lumbar enlargement. The dura is normal. The soft membranes are congested, particularly over the posterior aspect of the lumbar enlargement, where, in addition, they present a grayish appearance. On section, the central gray matter appears slightly congested. In the lumbar region a grayish patch is apparent in each postero-external column, while the postero-internal division is also grayer than normal, and somewhat pink. No other change is apparent to the naked eye.

The skull-cap weighs 10½ozs., and is thin generally. The dura mater is normal. The encephalon weighs 53½ozs.; the right hemisphere, 22½; the left, 22; the cerebellum, pons, and medulla, 7. The soft membranes are gelatinous, opaque, and tough; but they are nowhere adherent, being separated from the underlying convolutions by a considerable quantity of subpial fluid. The brain tissue is œdematous and soft; the cortex is congested in a somewhat patchy manner, but not apparently atrophied. The ventricular fluid is slightly increased, but the walls are perfectly smooth. The central medullary substance is of a pure white colour.

The heart weighs 7ozs.; its cavities are contracted; its tissue pale and firm, and its orifices normal. There is rather extensive atheroma of the ascending aorta. The left lung weighs 10½ozs., the right 24ozs.; in the latter there is basal congestion; otherwise both are normal. Spleen 2ozs.; left kidney 3½ozs., right 3ozs.

* The average weight of the spinal cord in 73 male insane persons dying under the age of 30 is given by Boyd ("Table of Average Weights of the Body and Brain") as 1·1 oz. The weight of the cord varies, according to Quain ("Anatomy," 8th Edit.), from 16 to 28 drams. The measurements of the normal cord are:—

	Transverse.	Sagittal.
Cervical	13 or 14	10
Dorsal	10	8
Lumbar	12	9 mm.

— Erb in Ziemssen's "Cyclopædia," Vol. xiii., p. 11.

Liver, 55ozs., is slightly fatty. Intestines normal. Enlargement and caseation of mesenteric glands.

Microscopic examination.—Brain. In sections taken from the upper end of the central convolutions, and stained with carmine, the large pyramidal nerve-cells of the third layer of the cortex present evidences of a considerable degree of atrophy and degeneration. They are smaller than normal, and they have indefinite outlines and withered-looking processes; they have a generalized yellow-granular appearance, and in many instances the nucleus is completely obscured. In sections stained with osmic acid ($\frac{1}{6}$ per cent. sol.) the degenerated nerve-cells take on a dark stain, varying from a deep brown to almost complete black. The vascular walls are nowhere thickened, but the perivascular sheaths in many of the smaller arteries is occupied by hæmatoidin particles, lying free or enclosed in granular cells.

Crura cerebri.—The nerve-cells of the locus niger are filled with brown and often quite black pigmentary particles, so that the nucleus is only exceptionally to be detected. Hæmatoidin particles occur in the perivascular sheaths, but as regards the medullary substance no material alteration is to be noted, and in particular no sclerotic process either in the region of the pyramidal tract or elsewhere.

Cervical cord.—The microscopic appearances indicate a degree of generalized sclerosis, with specialized areas of degeneration of greater intensity. The supporting connective tissue over the whole section is coarser than normal; the neuroglia-cells are large and prominent; the vascular walls are considerably thickened, and hæmatoidin particles occur occasionally in the walls and perivascular spaces. In carmine-stained sections, the areas of more advanced sclerosis are indicated by a deeper staining. These areas (Fig. 1) affect the lateral and posterior columns. That in the lateral columns assumes a triangular form; it has badly-defined outlines; it is separated externally from the periphery by a narrow zone of more healthy tissue; posteriorly it touches the posterior cornu, and anteriorly it reaches as far forward as the level of the central canal, shading off gradually into the more normal tissue of the anterior root-zone. In this area the nerve-fibres are diminished in number, but many of those remaining are of normally large size. This description applies to both lateral columns, the degeneration being strictly symmetrical. In the posterior columns the degeneration is less intense; it affects the whole extent of the internal divisions, but only a small portion of the external divisions, forming a wedge-shaped area which reaches quite to the periphery, but is separated from the posterior cornua and central parts of the gray substance by a zone of more healthy tissue.

The smaller nerve-cells of the anterior cornua of the gray substance seem fewer in number than normal. The large multipolar

corpuscles are extensively degenerated and slightly atrophied; their outlines are wanting in definition; their processes are shrunken-looking, and their interior is occupied in varying proportion by brownish granules, collected sometimes in one or other of the polar recesses, or distributed more generally through the cell-substance, more or less completely obscuring the nucleus. In carmine-tinted sections these degenerated parts of the nerve-cells do not take on the staining, but appear as brownish-yellow areas, and in sections stained with osmic acid they assume a tint varying from deep brown to black. The vessels are numerous and dilated, and the central canal is obliterated and replaced by a mass of round cells.

Lumbar cord.—In this region there are also evidences of a generalized slight sclerosis, and localized areas of more advanced degeneration. The area (Fig. 2) of lateral sclerosis is here much diminished, and it is confined to the posterior extremity of the column, reaching quite up to the periphery, but separated from the central gray substance by a zone of comparatively healthy tissue. The posterior sclerosis does not affect the deeper parts of the columns, nor, except at the extreme outer part of the external divisions, the parts lying towards the periphery. It extends transversely over the whole extent of the columns in their middle three-fifths, and it varies somewhat in intensity. Here also the nerve-cells of the central gray substance are extensively degenerated, and it is noticeable that the smaller bipolar cells of the posterior cornua share in some degree in the degeneration. The central canal is normal.

In several of its features, *e.g.*, the tremor of the lingual muscles and the speech-embarrassment, this case resembles one of general paralysis, but never, during the whole course of the affection, either before admission, so far as could be gathered from the history, or during his residence in the asylum, did he manifest any symptoms of that mental exaltation which is so common a characteristic of the early stage of confirmed general paralysis. On the other hand, the prevailing mental condition was one of more or less progressively advancing enfeeblement, pointing rather to primary dementia.

The physical signs indicate a widespread affection of the whole cerebro-spinal nervous centre, but from the point of view of the affection of the spinal cord, the case presents the features mainly of ataxic paraplegia. The gradual failure of motor power, the rigidity of the limbs, and resistance to passive movement, indicate an affection of the lateral columns, while the absence of the knee-jerk, the ataxia, the diminished sensibility, and the deficient equilibra-

tion on closure of the eyes, constitute the symptoms of posterior sclerosis. It must be remarked, however, that the case involves more than the question merely of sclerosis of the posterior and lateral columns. Symptoms indicating extension of the morbid affection to the gray substance are not awanting. The gradual wasting of the muscles, and the fibrillary tremors, indicate a tropho-irritative affection of the nerve-cells of the anterior cornua, and the dermic necroses an irritative affection of the posterior parts of the central gray substance.

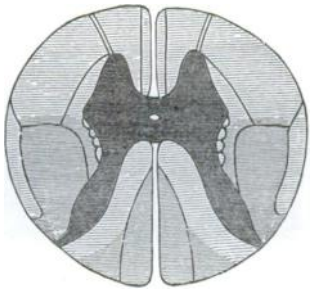
There is on some points a discrepancy of opinion between the two most recent writers on this subject—Grasset and Gowers. From a perusal of the section devoted to the description of this affection, I should say that probably the latter had not, at the date of publication of his work on "Diseases of the Nervous System," seen Grasset's article in the "Archives de Neurologie." While Grasset's article comprises three cases observed by himself, and a tabulated summary of 33 other cases, in all of which autopsies had been performed, Gowers says "a few pathological observations have been published." According to Gowers, the knee-jerk is in the majority of cases quick and extensive; in Grasset's 33 tabulated cases explicit reference is made to the condition of the patellar tendon-reflex in 19 instances, and of these abolition is noted in 12, exaggeration in 7. In the case here described, in a case of melancholia which I have elsewhere* described, and in a case of general paralysis whose cord I have recently examined, in all of which there was found, *post-mortem*, sclerosis, both of the lateral and posterior columns, the knee-jerk was abolished, so that I am rather inclined to adopt the view of Grasset, viz., that "abolition is much more frequent than exaggeration."

The facts of this case, so far, at least, as regards the lower extremities, are not altogether in accordance with the view expressed by Westphal and Zacher, viz., that in a combined lesion of the pyramidal and posterior columns the spastic phenomena are not developed in the superior or inferior members when the lesion of the posterior columns affects the posterior radicular zones in the corresponding sections of the cord.

Grasset, who proposes for this form of disease the name *Combined Tabes*, classifies it as one of the *Mixed Myelitis*, in-

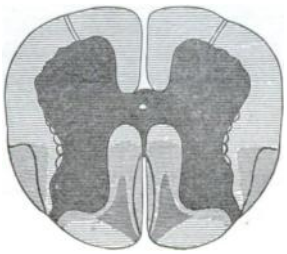
* "Glasgow Medical Journal," October, 1886, p. 250.

cluding under that term those myelites which are at once *diffused* and *systematic*—the posterior sclerosis being systematic, the lateral diffuse. The lateral sclerosis I look upon as *primary*, as contradistinguished from secondary descending degeneration, the anatomical features approximating more to those of the former than the latter.



CERVICAL (J. Bradley).

FIG. 1.—Spinal cord; cervical region; from a case of ataxic paraplegia; degeneration of lateral and posterior columns.



LUMBAR (J. Bradley).

SPINAL CORD.

FIG. 2.—Spinal cord; lumbar region; from a case of ataxic paraplegia; degeneration of lateral and posterior columns.

NOTE.—The writer may here mention that Grasset considers that the description of the cases collected in his tables affords a reply to the question raised by Dr. Bramwell in the foot-note of page 224 of the second edition of his work on "Diseases of the Spinal Cord" as to the condition of the knee-jerk in cases of locomotor ataxia which have become complicated with lateral sclerosis.