ABSTRACTS

SENSORIMOTOR NEUROLOGY.


The conception of a postencephalitic variety of muscular atrophy strikes an unfamiliar note in English neurology. Wimmer’s series of 20 such cases however have in certain instances been confirmed pathologically. His cases fall into five groups:—the ‘bilateral distal,’ the ‘proximal,’ and the ‘hemiplegic’ varieties, a bulbar-myasthenic type and lastly a form resembling amyotrophic lateral sclerosis. Parkinsonism was present in four cases only. Postmortem examination was available in three of the series, the changes comprising an outfall of cells from the anterior horns of the cord with degeneration of the pyramidal tract. Hyaline changes were found in the blood-vessel walls, and the Virchow-Robin spaces were infiltrated with lymphocytes. A certain amount of scepticism is legitimate with regard to the diagnosis in many of Wimmer’s cases, the evidence of an encephalitic origin being extremely slender.

M. C.


The first patient, in addition to respiratory seizures of rapid breathing without apnoea, had generalized convulsive attacks occurring independently of, or following, the respiratory disturbance. The other two had paroxysms of apnoea in full inspiration with spasm of the respiratory muscles, followed by loss of consciousness and convulsive movements.

The authors believe that convulsions produced in experimental animals by greatly increasing intrathoracic pressure are comparable to the symptoms observed in the last two patients. Clinical observations upon one of these patients seemed to show that the intensity of respiratory seizures, like the convulsions occurring in other conditions, could be altered by changes in the chemical composition of the blood.

R. G. G.


The case of Schilder’s disease reported here is of interest especially on the clinical side. The illness started abruptly, the patient fell in the street, was unable to rise, and was then found to be extremely ataxic in both upper limbs. Periods of improvement were followed by periods of regression for 13 years, during which intention tremor, nystagmus and ataxia were the most prominent early symptoms, and were later followed by certain evidences of aphasia and right-sided Jacksonian fits. The patient finally died in coma. Various
diagnoses including neuritis and hysteria were made. The possibility of disseminated sclerosis was considered but rejected. Post-mortem the macroscopic appearances of the brain were typical of Schilder’s disease, the white matter especially of the frontal and occipital lobes on both sides being grey and sclerosed, with preservation of the cortex and of a thin subcortical layer of white fibres. In stained sections all parts of the brain were found to be affected. In the pons and medulla and cerebellar peduncles localised areas of degeneration were also found. In the cord in addition to degeneration of the pyramidal tracts, two other areas of degeneration were seen, one in the anterior tracts on one side of the cervical cord, the other in the dorsal columns in the thoracic region. In the degenerated areas the axis-cylinders were better preserved than the myelin sheaths. Apart from the presence of numerous corpora amylacea throughout the brain and spinal cord no other lesions of special interest were found.

J. G. G.


Two cases of ruptured aneurysm are reported, one of congenital origin and the other of the arteriosclerotic type. Each patient died as the result of intracranial haemorrhage, and in each the gradual and progressive character of the cerebral symptoms, combined with evidence of irritation of the nerves at the base of the brain, was sufficient at least to suggest a clinical diagnosis of ruptured intracranial aneurysm.

R.M.S.

[16] Gradenigo’s syndrome (Sindroma de Gradonigo com paralisis do facial dos anos apos o trauma, etc.)—M. ALBERNAZ. Revista Oto-Neuro-Oftalmol., 1929, iv, 117.

Gradenigo’s syndrome consists of (1) acute otitis media, (2) severe temporo-parietal pain, and (3) convergent strabismus, from paralysis of the external rectus. It is produced by lesions situated in the petrous temporal, lateral to the sella turcica and in the vicinity of the cavernous sinus. The case here reported was of traumatic origin, from a revolver bullet, which lodged in the region mentioned. It was complicated by a facial paralysis. Operation resulted in relief of the symptoms, but a cerebral abscess developed and caused death. Photographs and X-ray pictures show the exact site of the lesion.

S. A. K. W.


A survey of literature, more particularly of Adie’s and Wilson’s papers, is given; then follows a full report of a case showing sleeping attacks, falling,
and attacks of powerlessness associated with hallucinations of the patient’s
mother or brothers kneeling beside him, sometimes giving him calm and kindly
advice.

R. G. G.

[18] A cavernous angioma of the mesencephalon (Di un caso di angioma
cavernoso del mesencefalo).—C. DEMEL. Il Cervello, 1929, viii, 1.
In the case of a man of 51 who died of lobar pneumonia, a cavernous angioma
of the superior corpora quadrigemina was an accidental finding at autopsy.
A description of the histological character of the tumour is followed by a
discussion of a number of analogous cases from the literature. In view of the
site of the lesion it is regrettable that no clinical examination of the nervous
system appears to have been made.

S. A. K. W.

[19] On the pathogenesis of ophthalmoplegic migraine.—HOLGER EHRLERS.
The association of migraine with recurrent paralysis of the oculomotor nerve
was first described by Gubler in 1860; the name ‘migraine ophthalmologique’
was suggested 30 years later by Charcot. In an excellent review of this syn-
drome, Ehlers emphasizes the long duration of the history and the ultimate
permanence of the condition in many instances. Opinions are divided as to
the lesion in respect of the third nerve; the strict unilaterality of the signs
and the absence of involvement of other cranial nerves suggest the peripheral
portion of the oculomotorius as the probable location. The close anatomical
association of the third nerve with the posterior cerebral artery within Tarini’s
fossa (the fossa interpeduncularis) leads Ehlers to the following conception
of ophthalmoplegic migraine. Local vasomotor disturbance is the probable
pathogenic agent in migraine: the posterior cerebral artery is the vessel
supplying that part of the cerebrum to which the teichopsis and the hemianopia
are referable. It is probable therefore that an intermittent vascular change
in the posterior cerebral artery is responsible. Dilatation of this artery will
press upon the adjacent third nerve and so interfere with its function (cf.
cases of oculomotor palsy due to pressure of aneurysms of the posterior cere-
bral artery). The author admits that his hypothesis is not applicable to those
rare cases of ophthalmoplegic migraine in which some ocular nerve other than
the third is implicated.

M. C.

nerv. e ment., 1929, xxxiv, 1.
This is a superb communication of some 162 pages, based on a personal case
of a typical kind, and beautifully illustrated. Since the appearance of Hall’s
monograph there has been nothing to compare with it for its wide conspectus
of the general problems arising out of the subject and for its critical discussion
of the phenomena of the malady. The reader will find here a complete investigation of semiology, pathology, pathogenesis; of the relation of the sympathetic system to the condition; of 'postencephalitic Wilsonism' (sic); of the kinship between it and Westphal's pseudosclerosis, as also torsion-spasm. All this is admirably done. The writer points out, as have others, that Alzheimer's atypical glia cells are not pathognomonic of any particular disease. He concludes that pseudosclerosis and Wilson's disease are two types of the same affection, though distinguishable. There is a bibliography which contains some 250 references.

S. A. K. W.


This is a careful study of a case of Wilson's disease, with characteristic clinical and pathological findings, in a boy of 16. The liver was cirrhosed and the spleen enlarged. There is no record of any corneal 'ring' pigmentation. In summary, it embraces the following conclusions:

1. There was an isolated, bilateral, symmetrical softening of the putamen, all other parts of the brain exhibiting no chronic histological change whatever.
2. This putaminal softening consisted in degeneration of all ectodermal tissues (parenchyma and glia), with widespread fatty change.
3. The so-called atypical glia-cells of Alzheimer are in reality degenerative forms of protoplasmic glia, which are seen in other conditions than that with which this paper is concerned.
4. The 'status spongiosus' of the putamen develops when the neuroglia is also involved, when its faculty of developing fibres fails.
5. Wilson's disease and pseudosclerosis are similar conditions varying in localisation and degree of involvement of parenchyma.
6. These, and some forms of torsion-spasm, belong to the group of toxic ectodermotropic affections.
7. They do not belong to a heredofamilial class; they are the expression of constitutional but not of hereditary factors.

S. A. K. W.


Two cases of teratomatous cysts of the spinal cord are described, which were successfully removed at operation. Both contained ciliated cells. The first patient, a child of two years, who came under observation because of irritability and 'tender' abdomen, gave a history of always having dragged its right foot. Repeated lumbar punctures at the level of the eleventh dorsal vertebra gave thick egg-white fluid filled with ciliated cells. At operation a large
flattened cyst was found extending from the tenth dorsal to the fourth lumbar vertebra. Histologically it proved to be a relatively simple teratoma lined with ciliated columnar epithelium. The child recovered.

The second patient, a young woman of 27, had had five attacks of left hemiplegia with pain in the left cervical region and Brown-Séquard dissociation of sensation in trunk and extremities (pain and thermanæsthesia on the right, loss of position sense and astereognosis on the left) at varying intervals since the age of two. Her present attack developed into an almost complete quadriplegia with an upper level of sensory disturbance at the fourth cervical segment, and embarrassment of respiration. A cyst filled with mucus and ciliated cells, attached to the left side of the cord at that site, was removed. Histologically there were also found in it long flat bands of smooth muscle, groups of serous and mucous glands, fat-cells, masses of lymphoid tissue, many blood-vessels, some cartilage, many small bundles of myelinated nerve fibres, and some large round cells resembling nerve-cells. After operation an excellent return of power occurred, with less complete return of sensation.

Both cysts were congenital and probably represent ependymal diverticula of the central canal of the spinal cord. References to the scanty literature on this subject are given.

J.V.


The case here described in detail is certainly unusual and perhaps capable of more than one interpretation. It is that of a man who at the age of 52 sustained a rather serious injury of the left knee, with slow development thereafter of osseous change and a consecutive neuritis. The latter continued to develop, and after four years signs of amyotrophic lateral sclerosis made their appearance, resulting in death some 14 months later. The amyotrophies commenced in the injured limb (left quadriceps) and in the right they affected the corresponding groups (in analogy with the traumatic neuritis); only the right arm was in the end unimpaired. The affection of the left limbs finally involved the face, giving rise to a kind of left hemiparesis.

In his discussion the author is evidently inclined to place the amyotrophy as reflex in origin—that is, the result of abnormal sensory excitations. In the brain, however, the pathological appearances included numerous acellular plaques in mainly the third and fifth cortical layers; they were also abundant in the neostriatum and in the thalamus. The appearances associated with amyotrophic lateral sclerosis were evident in cord and also in cerebrum. Argentophil granulations were obvious in the plaques; and in the nuclei of various cranial nerves (nucleus of inferior oblique, facial, nucleus ambiguus, hypoglossal) a deposition of what seemed to be melanin had occurred in some
quantity. He dismisses all theories assigning an 'inflammatory' origin to the affection, and stresses its transneuronal nature. He considers it feasible that the degeneration is the result of an "extreme functional overwork" associated with the abnormal excitations mentioned above. In the event, a kind of premature senium set in, revealed by plaques analogous to, if not identical with, those seen in arterio-pathic conditions.

J. V.


In the opinion of the author tremor can be distinguished from dysmetria as follows:

1. Action tremor appears under the influence of any internal or external stimulus, inclusive of movements. Cerebellar dysmetria appears exclusively during movement; it is a locomotor phenomenon, whereas the other may or may not be locomotor.

2. Action tremor is absolutely rhythmical, but dysmetria is non-rhythmical and irregular.

3. In cerebellar dysmetria the limb oscillates as a whole; in action tremor limited muscular groups may be in activity.

4. 'After-discharge' is seen in some action tremors, but there is no such phenomenon in dysmetria.

In some 28 cerebellar cases the author claims to have distinguished pure dysmetria in 23, and a combination of dysmetria and action-tremor in 5. In three decerebellarized dogs this latter combination was found; in them the cerebellum was completely extirpated, which "proves that cerebellar phenomena are produced outside the cerebellum, perhaps in the Rolandic convolutions."

S. A. K. W.


In the great majority of cases of disseminated sclerosis the tremor belongs to what the author describes as 'type A,' which is identical with cerebellar dysmetria. In other cases, it may belong to 'type B,' identical with pyramidal tremor (tremor during pyramidal action); or to 'type B1,' identical with the tremor of cerebellar action; or to 'type C,' which is a mixture of A with one or other of the B types. There is no such phenomenon as a specific 'intentional tremor;' and in most cases the involuntary movement is not a tremor at all, but a dysmetria—an irregular execution of movements. The author holds that intention-tremor has nothing to do with 'intention' but corresponds merely to the movement itself. He also considers that augmentation of the
oscillation at the end of a movement (in disseminated sclerosis) is distinctly rare. The 'tremor at rest' of paralysis agitans cases is due to a rhythmic discharge in certain groups of nerve-cells and differs fundamentally from the 'tremor' of disseminated sclerosis (type A); but the B and B\(^1\) types do not differ essentially from the former.

S. A. K. W.

---

[26] **Extraneural and neural anomalies in Friedreich's ataxia.**—B. J. ALPERS and R. W. WAGGONER. *Arch. of Neurol. and Psychiat.*, 1929, xxi, 47.

Spina bifida occulta was found to be present in seven members of a family of ten, all of whom were susceptible to the development of Friedreich's ataxia. Only four of these were frank, outspoken types of hereditary ataxia, while two others were examples of the abortive type, or the "forme fruste" of Rombold and Riley. The presence of spina bifida occulta suggested a search of the literature for other developmental malformations and it was found that practically every system of the body may show physical evidence of degeneration. The anomalies include: disturbances of the eyes, such as astigmatism, cataracts, optic atrophy and retinitis pigmentosa; endocrine imbalance, manifesting itself as myxoedema, cretinism and infantilism; deformities of the hands and feet in the form of syndactylism and main-en-griffe; bony deformities of the skull, micrognathia and fragilitas ossium; anomalies of the hair, muscles and skin; malformations of the ear and degeneration of the cochlear division of the eighth nerve.

The occurrence of such purely developmental malformations can hardly be explained on any basis other than that of a tendency to direct inheritance, and there are good grounds for believing that Friedreich's ataxia is not only a familial but a hereditary disease which appears sporadically, involving either single or many members in the group.

R. M. S.

---


The patient, in addition to a psychoneurosis, suffered from a Brown-Séquard type of paralysis with pain in the area of the tenth, eleventh, and twelfth dorsal segments. Laminectomy was performed on expectation of finding a tumour, but an adhesive arachnoiditis was found instead; the thickening and adhesions were removed and the above-mentioned dorsal roots were cut. The patient made a good recovery and was able to return to work walking normally.

R. G. G.

---


The record of this case is of especial interest to the neurologist. A man, age 41 years, had a severe hemoptysis in 1916. A complete resection of the
NEUROLOGY

stomach was performed in 1918. He remained well till 1926 when he complained of paraesthesia. His blood count was typical of a primary anaemia. In 1928 he died of bronchopneumonia, and at postmortem findings consistent with the diagnosis of pernicious anaemia were found. The spinal cord showed degeneration in the central areas of the posterior columns. This is therefore a record of a case of subacute combined degeneration of the spinal cord in a patient who had his stomach removed eight years previously. E. A. C.


The author asserts from clinical observation that inversion of the abdominal reflexes may occur independently of paresis of the muscles on the side stimulated; that in spinal compression a genuine crossed abdominal reflex can be obtained; and that these inversions are usually missed because in the first place strong excitation is necessary, and, in the second, because a latent period may exist.

His theory is, apparently, that in spinal compression pyramidal function is interrupted, whence the loss of the ordinary abdominal reflexes. But if sensory conduction is not completely blocked, then the stimulus can make use of a non-pyramidal motor system.

J. V.

PROGNOSIS AND TREATMENT.


This article is worthy of attention as it raises questions regarding the advisability of placing epileptic children upon a prolonged and rigorous ketogenic diet. Unfortunately the experiment has not been carried out over any prolonged period of time. The author found that on a ketogenic diet the output of calcium and phosphorus exceeded the intake; she also found that there was a major shift of the excretion from the faces to the urine. During the period of the experiment the calcium and phosphorus levels in the blood were within normal limits. It, therefore, appears that on such a diet children must call upon their stores of calcium and phosphorus, a state of affairs which may result in damage to the central nervous system or bones.

E. A. C.


Patients with moderately severe epilepsy were placed on a high ketogenic diet until seizures were prevented; conditions were then modified in various