
In a family of sixteen members, seven in three generations showed the complete picture of Graves’ disease. Four of them, seen by the author, are described. As a rule similar heredity is transmitted through the females, but these patients were cousins through their fathers.

W. J. Adie.

SENSORIMOTOR NEUROLOGY.


The writers explain the absence, in certain striate lesions, of the symptoms of one or other variety of involuntary movements, by severe concomitant involvement of the corticospinal paths, a view which has already been put forward by others and has much to commend it. Negatively, they do not know of any case of the syndrome of the corpus striatum being present in spite of pyramidal disease, but in the reviewer’s opinion this statement should not be taken too absolutely. The Vogts distinguish four types of pathological change in the corpus striatum.

1. État marbré (status marmoratus)—a ‘marbled’ condition of outfall of nerve-cells in irregular fashion and their replacement by nests of the finest myelinated fibrils (not glial overgrowth). It may be regarded as a dysplasia, a developmental defect, which the Vogts are inclined to associate with asphyxia neonatorum. They have examined six cases of état marbré, which all began in earliest infancy and were associated in each instance with the striate syndrome of choreo-athetoid movements, or tremors, associated movements, variable tonic muscular spasms, involuntary laughing and crying, etc., without genuine muscular paralysis. Some improvement in the cases was noted up to the fifth year of life. They were bilaterally spastic, but not in any sense paralyzed, and are comparable to some cases of Little’s disease. The authors, in short, limit the latter conception strictly to spasticity without paralysis, and couple with it the condition of ‘athetose double’, which they consider a “more severe form of Little’s rigidity ” : the two they separate rigorously from infantile cerebral hemiplegia or double hemiplegia.

2. État fibreux (status fibrosus)—a shrunken condition of the corpus striatum, as a result of which those myelinated fibres that remain seem unusually closely set, so that the ganglion gives a spurious appearance of being richer in fibres than ordinarily. With this the writers associate a slow progressive bilateral chorea without psychical impairment: they agree with other observers who have demonstrated the connection between disease of the corpus striatum and the involuntary movements of Huntington’s chorea.
3. **Total necrosis** of the corpus striatum. In this class are placed progressive lenticular degeneration (Wilson’s disease), and probably other subacute processes; they are connected also with changes in the liver.

4. **Acute lesions**: i.e., haemorrhages, acute softenings, inflammatory processes, etc., not of a familio-hereditary or degenerative character.

Other disease-processes may include in their incidence the corpus striatum, e.g., so-called pseudosclerosis, tuberose sclerosis, general paralysis, paralysis agitans, etc.

**Wilson.**

[22] **Torsion-dystonia (torsion spasm, dystonia musculorum deformans)**


Mendel’s communication is a complete little monograph on the disease known by the various names of dystonia musculorum deformans or dysbasia lordotica progressiva (Oppenheim), torsion-neurosis (Ziehen), progressive torsion-spasm in children (Flatau and Sterling), tortipelvis (Fraenkel), the Ziehen-Oppenheim variety of dystonia lenticularis (Thomalla). Mendel criticizes each of these denominations adversely, and proposes ‘torsion-dystonia’, which is satisfactory enough in a way. The important thing is to decide on one term, if practicable, and to stick to it.

Torsion-spasm or torsion-dystonia is an affection of comparative rarity, some 33 cases having been recorded since Schwalbe and Ziehen drew attention to it in 1908 and 1910. Much more common in the male than the female, it occurs almost exclusively in Polish Jews, though this is not a *conditio sine qua non*. Apparently some five cases outside the Jewish race have been described. An occasional familial element is traceable. The disease makes its appearance as a rule between 10 and 13, but it has been seen at 8, while one of Mendel’s patients was 45. It is characterized by the gradual onset of involuntary movements, described comprehensively if vaguely by Mendel as “a mixture of choreic, athetoid, tic-like, Parkinsonian-like movements”, at one time resembling one of these, at another time some other; they are clown-like, grotesque, bizarre, pointless, tonico-clonic, and especially apt to become tonic; coupled with this tonizing element is the characteristic tendency to twisting or torsion-like contraction of trunk and limbs. The face usually escapes. The movements more commonly affect the trunk, and proximal rather than distal segments of limbs, and disappear in sleep. There is a noteworthy degree of spine-twisting and lordosis in almost every case. In addition to the involuntary movements, the peculiar changing nature of the muscle tonus is a prominent feature; marked hypotonia is followed by hypertonia in the same group, resulting in a transient immobilization of the mobile spasm as in athetosis. Mendel says that passive movement reveals an obvious hypotonia, and that, with repetition of the movement, hypertonus, spasm, and tonico-clonic involuntary movements set in. The deep reflexes are reduced, occasionally exaggerated; pyramidal disease is conspicuous by
its absence, except in one or two cases considered doubtful by Mendel. Negatively, there are no muscular atrophy, speech defects, paralyses, sphincter impairment, or intellectual defect. The affection is to be distinguished from double athetosis, chorea, tic, juvenile paralysis agitans, myotonia, Wilson’s disease, and pseudosclerosis. It runs a chronic course, and cures are at present unknown. Its pathology is undetermined, for Mendel criticizes Thomalla’s diagnosis in his case (see this Journal, p. 87), considering it to belong definitely either to Wilson’s disease or to pseudosclerosis.

There are full references to the literature.

**Wilson.**


The patient was a boy, age 14, with some of the characteristic symptoms of torsion-spasm (‘dysbasia lordotica progressiva’, ‘dystonia musculorum deformans’), viz., irregular, involuntary, mobile spasms of the trunk and limbs, producing almost clown-like distortions of the body, especially of pelvis and spine. As has been noted in most, though not all, of the recorded cases, he was a Polish Jew.

This case of Thomalla’s is interesting because of the pathological findings, viz., a small cirrhosed liver, and a bilaterally shrunken and softened putamen, with secondary glia-formation, especially of glia nuclei; the parenchyma of the putamen had almost entirely disappeared, but the globus pallidus was practically normal, and the caudate nucleus seemed unchanged. Thomalla gives excellent clinical photographs, and a good discussion on the problems presented by his case, especially in its pathological similarity to progressive lenticular degeneration. His conclusion is that under the general term ‘dystonia lenticularis’ might be included double athetosis, pseudosclerosis—(Westphal–Strümpell), Wilson’s disease, and torsion-spasm.

It should be stated that some doubt has been expressed as to the classification of this case in the group of torsion-spasm (see this Journal, p. 86).

**Wilson.**


The patient, a man, age 43, had suffered for several weeks from athetoid movements of the face and the left arm and leg, with variable attacks of spasm in the trunk and limbs, often curiously rhythmical; his attitude of flexion and his general rigidity resembled those of paralysis agitans. Further, he showed on occasion a sort of torsion-spasm of the trunk, neck, and proximal segments of the limbs. The Wassermann test was negative in blood and spinal fluid, though infection was admitted; the abdominal
reflexes were active, and Babinski's plantar reflex was absent. Some six weeks later the patient died.

Pathological examination revealed bilateral softening of vascular origin in the lenticular nucleus, with evidence throughout the brain of lymphocytic and plasma-cell perivascular infiltration, and changes in the walls of the blood-vessels. The liver showed commencing cirrhosis.

The case is one illustrating extrapyramidal motor disease, and lends support to the view associating certain forms of involuntary movements and spasmodec rigidity with disease of the corpus striatum. Westphal describes two other cases, in one of which he correlates a clinical state of paralysis agitans sine agitatione with an asymmetrical bilateral softening in the putamen found at the necropsy, of syphilitic origin; and he adopts the general attitude that paralysis agitans, progressive lenticular degeneration, pseudosclerosis, perhaps also Oppenheim's dystonia musculorum deformans or torsion-spasm, are definitely to be assigned to the lenticular nucleus and corpus striatum.

WILSON.


A woman, age 40, who had her left breast removed for carcinoma, began, four days after the operation, to suffer from choreiform movements of the whole of the right side, and from bilateral facial chorea; occasional choreic movements were observed at first in the left arm and leg also. Eleven days later the left limbs became paresed, and at a later date bulbar symptoms made their appearance. The chorea of the right side was associated with much hypotonia of the muscles. Other nervous symptoms need not here be particularized, with the exception that the patient, in Bárány's pointing test, always made an error towards the left with the right arm.

Pathologically, three small secondary carcinomatous metastases were found. One was in the left regio subthalamiae, about the size of a pea, involving in part the ansa lenticularis and Forel's field. Bremme gives several reasons against associating this lesion with the patient's choreiform movements, which are ascribed with greater justification, in the author's opinion, to a larger metastasis almost completely destroying the right superior cerebellar peduncle in the mid-brain, under the corpora quadrigemina, just where its fibres commence to cross the fibres of the left one. A small tumour was also found in the extreme lateral edge of the right cerebellar hemisphere (lobulus semilunaris superior), to which the pointing error is attributed. The patient showed no tremor, it appears, and it may be noted that the red nuclei were normal. Possibly the chorea produced by the peduncular tumour was subsequently accentuated by the lesion in the left regio subthalamiae.

WILSON.


Thirteen cases are enumerated in which the diagnosis was certain, while four where there was some doubt as to diagnosis are mentioned. There
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were no histories of preceding influenza, and there was no example of more than one case in the same house. No evidence of a contagious element in the disease was forthcoming. The case are divided into two types, roughly:

1. Slow onset, in 10 of the 13. This type had ophthalmoplegia, followed about a week later by a lethargic stage. The writer suggests an infection of the region of the pons as a starting-point for this type.

2. Type with rapid onset, characterized by headache, delirium, and little evidence of ocular palsies. For this type an infection of the frontal lobes via the ethmoid region is put forward for consideration.

Individual symptoms: euphoria, followed by depression in convalescent stage. Apathy rather than lethargy. Diplopia, present in 11 cases, with vague palsies of other cranial nerves, the external eye muscles showing a paresis rather than a clear paralysis such as occurs with definite lesions of the nerves or their nuclei. Temperature: the pyrexia varied from 101° to 104.6°. Rigidity not real as in meningitis. Skin eruptions: discrete pinhead petechiae. Cerebrospinal fluid: increased pressure, sterile on culture; cell-count varied from 6 to 103 mononuclears. Leucocytosis in blood: 3 out of 12 examined, 15,000 to 20,000. Mortality: 4 patients died out of 13. No post-mortem details available.

J. LE FLEMING BURROW.

[27] Fifteen cases of involuntary movements following influenza and encephalitis lethargica (Quinze cas de mouvements involontaires apparus à la suite d'épisodes grippaux et d'encéphalite léthargique).
—Pierre Marie and G. Lévy. La Médecine, 1920, i, 270.

Since the autumn of 1918 a number of cases of involuntary movements following a febrile illness have come under notice. The following forms were observed: fine oscillatory movements of the limbs, most marked distally; slow regular movements of wide range, sometimes synchronous in the upper and lower limbs, most marked proximally; movements resembling Sydenham’s and Huntington’s chorea; and in two cases extraordinary rhythmical contortions of the trunk which interrupted progression and ended with a lateral eversion of the head and an antagonistic gesture. The movements in any form may be confined to one side. Several cases presented the picture of the rigid form of Parkinson’s disease. The movements began two or three months after the febrile illness; in most cases they were still present a year after the onset. They tend to disappear, but very slowly, and not always completely.

W. J. ADIE.


Early in 1917, and again in 1918, a number of cases of a peculiar form of encephalomyelitis occurred in certain country districts of Australia. The disease was acute, and often abrupt in onset, with a mortality of 70 per cent. The symptoms included pyrexia, coma, convulsions, and rigidity; in most cases there was no evidence of paralysis. The affection was conveyed by intracerebral inoculation to monkeys, from monkeys to sheep,
and from these back to monkeys or to other sheep. It was also conveyed to a calf and a horse. Histologically there was found distention of the perivenuous sheaths by lymphoeyte-like cells throughout the brain and cord, most often and most intensely in the corpus striatum, pons, and medulla.

The authors consider that in the group of diseases to which the Australian disease belongs, this cellular response around the veins is the result of a chemical irritant, and that the symptoms are due, not to the mechanical presence of the virus, nor immediately to its toxins, but to the effects of the cellular response itself. In an epidemic of one of the diseases of this group, e.g., of acute poliomyelitis, the paucity of cases is due chiefly to the fact that many individuals react to the virus to such a slight degree that no interference with function results; in a few the reaction, i.e., the cellular response, is great, and interference with function is prominent.

W. J. Adie.


Eight cases are described fully, with special opinions upon the various systems, and with full details of the clinical pathology of the blood, cerebrospinal fluid, etc. Four gave a history of recent ‘influenza’. Six had double vision or some paresis of cranial nerves, and one a masklike face such as would occur in Parkinson’s disease. One had root pains in the body, while half the cases were drowsy or apathetic. The Wassermann reaction with blood and spinal fluid was negative, and the average leucocytosis was about 10,000 per c.mm. in the blood. There was some excess of globulin in the spinal fluid in two cases. No new features of the disease are brought forward, and the clinical details are loosely put together, so that no clear picture of the state of the patients is presented.

J. Le Fleming Burrow.


This is an interesting paper in which a general historical account is given of the pathological conditions which may give rise to a hemilesion of the cord. These were originally looked upon as syphilitic in origin, until Gowers and Horsley demonstrated that they might be due to tumours, a condition amenable to surgical interference. The author very properly points out that the diagnosis of spinal tumour is not so easy as was once thought, and refers to the pathological conditions which may simulate (or complicate) tumour. Of these, he mentions localized simple meningitis, meningitis serosa chronica circumscripta, and arachnoperineuritis chronica serofibrinosa. These conditions cannot be said to have a very
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definite pathogenesis or clinical picture; they are associated with pressure symptoms by reason of the liquid tumours to which they give rise. Spinal tumour may be simulated occasionally by disseminated sclerosis, especially in the region of the conus medullaris, and rarely by ordinary myelitis. On the other hand, it not infrequently happens that spinal tumour is confidently diagnosed and yet no pathological condition of any kind is found at operation, or even, as in a case described by Nonne, on histological examination of the cord.

Another condition simulating tumour was described by Henneberg under the name myelitis funicularis; this may be non-progressive or even regressive. The five cases of which details are given in the paper, as well as similar ones described by Oppenheim, appear to come under this heading. They are characterized by the following group of symptoms: slow onset, usually with thermal paraesthesiae in one leg, or diffuse, not radicular, pains, weakness in the opposite leg, impotence, and mild urinary incontinence. The physical signs are those of a pyramidal lesion in one leg, diminution of painful and thermal sensations in the other, negative X-ray, normal cerebrospinal fluid, and negative Wassermann. The age of onset varied between 29 and 50, and all the patients were Jews. The course is typical; the condition reaches its maximum in a few months, and then gradually improves, the physical signs remaining practically unaltered. Remissions are common. Four of the cases had been under observation for more than ten years.

Goldflam labels the condition myelitis funicularis unilateralis, and strongly advises against operation. In a patient of Oppenheim's, who died three weeks after an exploratory laminectomy, a lesion was found limited to the white matter of the cord on one side, consisting of several foci which had coalesced, involving the lateral and posterior columns, and spreading to the cerebellar tracts.

The differential diagnosis from tumour, which in the early stages may be very difficult, is discussed at length.

J. L. Birley.

[31] Spinal tumours: statistics in a series of 330 collected cases.—


Forty varieties in the structure of spinal tumours have been recorded. As regards their location, it is possible to divide them into four groups: (1) Vertebral (primary and secondary); (2) Extramedullary (intradural and extradural); (3) Intramedullary; and (4) Caudal.

Early Symptoms.—

1. Vertebral group: Due to pressure on nerve roots and the cord. Pain is frequent, is persistent in character, and is often girdle in type. Local tenderness and rigidity of the spine may be present.

2. Extramedullary group: Here the nature of the pain depends on the site of the tumour. If on the posterior surface of the cord, pain in the back results. If on the nerve roots, the pain follows a typical root distribution. If the growth is near the anterior aspect of the cord, muscle palsies and cramps result.
3. Intramedullary group: Numbness and tingling in one or more extremities, followed by motor disturbances.

Tumours of lumbar region produce early sphincter troubles. Those in the upper cervical region cause respiratory disturbance.

Starr gives the sequence of the symptoms, as they arise in spinal-cord tumours, in the following order: (a) Peculiar pains with a limited distribution; (b) Increase of reflexes below the lesion; (c) Paraplegia; (d) Sensory loss; (e) Loss of all subjacent reflexes.

TREATMENT.—Gumma being excluded from the diagnosis, removal by operation is indicated. For the severe pain in inoperable malignant disease of the spine, the posterior spinal roots may be divided, or—and this gives more lasting benefit—the anterolateral columns of the cord may be cut.

Extradural tumours are removable, with a mortality of 38 per cent; intradural present more difficulty, but the mortality is no more than 35 per cent. Caudal tumours, on account of the involvement of the roots of the cauda equina, also present great difficulty. The mortality is 46 per cent. Intramedullary tumours are very satisfactorily dealt with by the two-stage operation. In the first stage the tumour is exposed only, and then at a later date removal takes place. (Horsley and Elsberg both had a nil mortality in dealing with these tumours.)

PATHOLOGICAL TYPES IN 330 SPINAL TUMOURS, WITH THEIR RELATIVE FREQUENCY.—

1. Vertebral group: Primary growths (mostly sarcomata), 18·6 per cent. Secondary growths (especially carcinomatous mammae), 4 per cent.

2. Extradural group: (a) Extradural (mostly fibromata and sarcomata), 16·6 per cent. (b) Intradural from pia arachnoid (mostly fibromata, endotheliomata, psammomata, and sarcomata), 29·4 per cent.

3. Intramedullary group (mostly gliomata, gliosarcomata), 11 per cent.

4. Caudal group (mostly endotheliomata, fibromata, and sarcomata), 9·1 per cent.

Location unstated in 16·3 per cent.

Tables are given illustrating further analyses of the cases. In the recoverable cases, the author lays stress on the necessity for early and careful operation, followed by efficient post-operative treatment.

W. JOHNSON.

TREATMENT.


Four cases are described as occurring in 550 peripheral nerve injuries. Of these four, three were operated upon and injected with 60 per cent alcohol, being cured after a previous simple neurolysis had failed to abolish the symptoms. The remaining case was under observation at the time of writing. Only one was observed as long as four months, and in this there was no return of the causalgia. No gross changes in the nerves were observed except superficial adhesions. The cases all occurred in the median, except one which affected the internal popliteal division of the sciatic nerve.

J. LE FLEMING BURROW.