REFRESHER COURSE FOR GENERAL PRACTITIONERS

DISSEMINATED SCLEROSIS

BY

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Disseminated sclerosis is one of the commonest neurological diseases in the United Kingdom and occurs rather more often in women than in men. The most frequent time of onset is between the ages of 15 and 30, but a considerable number of patients first have symptoms between 30 and 50, and occasionally the disease starts after the age of 50.

Diagnosis

This remains essentially a clinical problem, and little help can be obtained from pathological investigations except by the exclusion of other conditions in difficult cases. It is the evolution of the disease which gives the clue to the diagnosis, and typically there is evidence in the history of episodes of neurological disorder occurring at varying intervals of time and involving different parts of the nervous system, while on examination the physical signs indicate more than one lesion.

In taking the history very careful inquiry is necessary about the possibility of an earlier episode, as this may have been of short duration and forgotten by the patient, or some non-committal diagnosis such as neuritis, food-poisoning, or a nervous breakdown may have been given him. These early incidents are of various types: a common one, occurring in about 25% of cases, is a unilateral retrobulbar neuritis with dimness of vision in one eye coming on over the course of a few days, sometimes accompanied by pain on movement of the eyes, normal vision returning in three to four weeks or less.

Other early symptoms about which inquiry should be made are double vision lasting for a few days, weakness of one or more limbs often accompanied by numbness or tingling but not by pain, a severe attack of vertigo and vomiting without tinnitus or deafness, and difficulty in walking which lasts for a few days. When a history of one or more episodes of this type, sometimes occurring many years previously, can be obtained from a patient who has physical signs of involvement of the pyramidal, cerebellar, or sensory pathways, a diagnosis of disseminated sclerosis can usually be safely made. When, however, the patient is seen in the first episode with an uneventful previous history, and the physical signs can be accounted for by a single lesion in the nervous system, there may be considerable doubt about the diagnosis except in the case of unilateral retrobulbar neuritis, which, in the United Kingdom at any rate, is nearly always due to disseminated sclerosis. In cases of this type the course of the disease will usually clarify the diagnosis either by the appearance of lesions in other parts of the nervous system or by the patient making a symptomatic recovery. Patients who have persistent symptoms and physical signs should be investigated more thoroughly and kept under regular observation so that more potentially treatable conditions, such as myasthenia gravis, neurosyphilis, subacute combined

degeneration of the cord, and cerebral or spinal tumours are not overlooked.

Physical Signs

As would be expected from the disseminated and variable distribution of the plaques in the central nervous system, physical signs vary considerably from case to case, and also at different times in the same patient. In retrobulbar neuritis, which is common as an early symptom, but may also occur at later stages of the disease, the visual acuity is reduced but the optic disk is often normal, although in a minority of cases there is unilateral papilloedema. Testing of the visual fields by confrontation will, however, always show a central scotoma with a full peripheral field. In patients with temporary uselessness of an arm it is common to find impairment of position sense and of stereognosis, but cutaneous sensation is usually normal.

The pyramidal tracts tend to be involved early in the disease whether it advances by relapses and remissions or by gradual deterioration without remissions, and this results in a variable degree of spasticity and weakness with increased jerks and extensor plantar responses. The abdominal reflexes disappear at an early stage; this is a sign of some importance, as in certain other conditions affecting the pyramidal tracts—in particular, amyotrophic lateral sclerosis—the abdominal reflexes may be retained in spite of very brisk tendon jerks and extensor responses. Posterior column sensation, especially appreciation of vibration, also tends to be reduced in the early stages of the disease. Lower motor neurone lesions, with wasting and absent tendon jerks, are of great rarity, and if these are present a diagnosis other than disseminated sclerosis is probable. The really important finding is physical signs which cannot be accounted for by a single focal neurological lesion or by a systematized tract degeneration.

There are three groups of physical signs which at times have been overemphasized in the diagnosis of disseminated sclerosis. The first is Charcot's triad of nystagmus, intention tremor, and scanning speech. These are signs of cerebellar dysfunction which may occur with various pathological processes. They may of course occur in disseminated sclerosis, but only in a relatively small proportion of cases and then often in the late stages of the disease. The second is bitemporal pallor of the optic disks: the colour of the normal disks varies considerably from person to person, the temporal side usually being paler than the nasal, and pathological pallor is often very difficult to assess. Unless there is a history of retrobulbar neuritis or impairment of visual acuity with visual field changes, considerable care should be taken in the assessment of temporal pallor. The third of the groups is euphoria, which is said to be common in disseminated sclerosis: this is a quality difficult to estimate unless one knows the patient well, and certainly many sufferers from the disease are not euphoric. Though euphoria may occasionally be present, a much commoner finding, especially in the later stages, is emotional lability, and at times pathological laughter and crying.

The patients in whom diagnosis is most difficult are those, frequently over the age of 40, in whom there is slow but steady progress of symptoms, such as a spastic weakness of the legs with minimal sensory involvement, or a slowly progressive hemiparesis without evidence in the history or

physical examination of disseminated lesions, and in these patients further investigation is necessary. In the patient with slowly progressive weakness of the legs the blood Wassermann reaction and a lumbar puncture are necessary, and if the Queckenstedt response gives any indication of spinal compression, or if the protein content of the cerebrospinal fluid is raised, a myelogram should be carried out. With a progressive hemiparesis x-ray examination of the skull, the blood Wassermann reaction, and a lumbar puncture will be needed, and possibly an air-encephalogram to exclude a cerebral tumour.

The Cerebrospinal Fluid

The common finding in disseminated sclerosis is a normal fluid. In about 30% of cases the lymphocyte count is raised to about 10 per c.mm., and occasionally up to 40-50 per c.mm. The protein content is raised to 50-100 mg. per 100 ml. in a similar proportion of cases, and is very occasionally over 100 mg. per 100 ml. The most striking abnormality is a first-zone, or "paretic," Lange gold curve, which occurs in about 25% of cases; when this is found with an otherwise normal cerebrospinal fluid and the Wassermann reaction is negative in both the blood and the cerebrospinal fluid, and there is no history of antisyphilitic treatment, it is important confirmatory evidence of the diagnosis.

Prognosis

This is a matter of considerable difficulty in any individual case in the early stages, as there are three different ways in which the disease behaves. (1) There may be a series of relapses and remissions with, in general, the relapses becoming more severe and lasting longer on each occasion; during remissions the patient may have no disability, but residual signs such as absent abdominal reflexes and extensor plantar responses may persist. The length of remission varies from a few weeks up to 20 or more years, and it is probable that there are occasional patients who have one or two attacks and then remain symptomless for the rest of their lives. (2) In some patients there is a slow but steady progress of the disease, sometimes punctuated by periods of slight improvement, but a complete remission never occurs. (3) There is a small group of patients in whom the disability progresses to a certain stage of disability, which may be slight or severe, and then remains stationary for many years.

In general, a remitting course is usual when the disease starts before the age of 35-40, and a slowly progressive course without remissions when it starts above this age, but there are exceptions to this. Attacks of acute onset are more likely to remit than those which start gradually, while involvement of cranial nerves, in particular retrobulbar neuritis, or of sensory pathways more often recovers completely than does involvement of the pyramidal or cerebellar pathways. Once an attack has persisted for six months a notable remission is improbable.

In exceptional acute cases with a plaque in the brain-stem, death may result from respiratory failure, but the common causes of death in the chronic cases are respiratory or urinary infections or toxæmia from bed-sores, while a number of patients die of some coincidental disease. It has been estimated that the average duration of life is about 20 years from the onset of the disease.

Management and Treatment

In early cases it is generally advisable not to tell the patient the name of the disease, as "popular" medical books are apt to be consulted and a uniformly gloomy prognosis assumed, which may well be wrong in the individual case. The patient who recovers from an attack of retrobulbar neuritis or a temporary paralysis of a limb who is told that she has disseminated sclerosis, and who then waits for the next attack of the disease and eventual complete incapacity,

when in fact she may have many years of good health or minor disability, presents a dismal spectacle. It is usually, however, advisable to explain the situation more thoroughly to a responsible relation and to give warning of the probability of relapses.

Treatment in bed is needed only in severe acute episodes of the disease and in severely disabled chronic cases who cannot be looked after otherwise. Other patients are better not confined to bed, particularly those with ataxia of the legs, who often become more ataxic when kept in bed. Fatigue aggravates symptoms and should be avoided so far as is possible, but, in general, patients should continue at work when this is at all practicable, as this is important from the point of view of morale. Febrile illnesses cause temporary aggravation of symptoms in many patients, and sufferers from the disease should take a cold in the head or influenza more seriously than normal people and not try "to fight it off" by continuing at work, but should have a few days in bed. There has been a general impression that relapses tend to occur in pregnancy or in the puerperium, but recent follow-up studies have not confirmed this as a general rule, and disseminated sclerosis is not in itself a good indication for termination of pregnancy. It must, however, be remembered that the care of an infant may be a serious extra burden for a woman partially disabled by the disease.

Medicinal Treatment

The effects of treatment are notoriously difficult to assess in a disease which has spontaneous remissions, but it is generally agreed that no drug treatment known at present has any effect in arresting the course of the disease. Many drugs have, of course, been employed: arsenic has for long been used empirically, given by the mouth as liq. arsenicalis 3 minims (0.18 ml.) in solution three times a day for six weeks at a time, or by weekly intravenous injections of 0.45 g. of neoarsphenamine, six to eight injections being given in a course; but toxic reactions may occur with neoarsphenamine and chronic arsenic poisoning with liq. arsenicalis. Other substances which have been used include copper, gold, dicoumarol, and tetraethylammonium bromide, but all these are potentially dangerous and have no definitely beneficial effect. In a condition in which there is no reliable evidence that any medicinal treatment can alter the course of the disease it is unwise to use a potentially dangerous drug, and it is preferable to employ as a placebo some substance that will not harm the patient; it is in this category that liver injections and quinine have been used, the former in 2-ml. injections once a week in courses lasting eight weeks, the latter as iodobismuthate of quinine, 1 ml., given by deep intramuscular injection weekly for a similar period. Fever therapy, induced either by intravenous injections of increasing doses of T.A.B. vaccine or by a Kettering hypertherm, has been employed in treatment; but results are far from encouraging, and some patients, especially those with bulbar or sphincter disturbance, are worse after this method of treatment.

Symptomatic Treatment

Physiotherapy, especially re-educational walking exercises for ataxic patients, is of real help—in particular in re-establishing confidence and in helping them to make the best possible use of their disabled limbs.

Urgency of micturition is common in patients with bilateral pyramidal involvement, and this can often be helped by ephedrine, $\frac{1}{2}$ gr. (32 mg.) twice a day, by a pill containing dry extract of belladonna $\frac{1}{2}$ gr. (32 mg.), and extract of hyoscyamus $1\frac{1}{2}$ gr. (0.1 g.), or by a tablet containing ergot, belladonna, and phenobarbitone. Less commonly, retention of urine or retention with overflow occurs, and in these patients carbachol, 2 mg. by the mouth six-hourly, may be helpful.

Spasticity is a common cause of difficulty in walking in patients in whom reasonably good muscular power remains,

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but unfortunately it is difficult to influence this. Injections of curare in peanut oil have been tried, but these are very uncertain in action and are not to be recommended. Mephenesin acts as a depressant of spinal cord reflexes, and theoretically should be helpful in reducing spasticity, but in practice it is usually impossible to give a dose large enough to achieve this without producing toxic symptoms. In a minority of spastic cases, however, it is of some value, and should be given orally as the elixir, starting with one drachm (3.5 ml.) six-hourly, and increasing the dose gradually to three drachms (10.5 ml.) six-hourly if giddiness and double vision are not produced as evidence of overdosage. With a spastic foot-drop, a toe-raising spring is often helpful in preventing the patient constantly catching his toe on the ground when walking. In the later stages of the disease, when flexor spasms frequently bother the patient at night, a cradle should be put over the legs to prevent contact with the bedclothes, as this contact may initiate the reflex flexor spasms, and a barbiturate sedative given.

Epileptic attacks and paroxysmal trigeminal neuralgia occasionally occur as symptoms of disseminated sclerosis. The epileptic attacks can be controlled by the medicinal treatment used for idiopathic epilepsy as described in an earlier article in this series (*British Medical Journal*, 1950, 2, 1047), and trigeminal neuralgia responds as well as does the idiopathic variety to alcohol injection of the Gasserian ganglion or operative section of the sensory root of the fifth nerve.

Next Refresher Course Article.—"Interpretation of Wounds," by Dr. F. E. Camps.

A GERIATRIC UNIT IN A GENERAL HOSPITAL

BY

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With the approval of the Sheffield Regional Hospital Board a geriatric unit was formed in 1949 at the City Hospital, Nottingham. As associations with other hospitals and with statutory and voluntary bodies engaged in the care of the elderly developed, the local set-up began to conform to the plan for an area geriatric service proposed by the Special Committee of the British Medical Association on the Care of the Elderly (1947). The "lines of communication" were established, but free movement along the lines was restricted by insuffi-

cient accommodation at all points. Following the outlines of the plan of the B.M.A. Special Committee a diagram of the geriatric scheme in Nottingham and district is here given.

Central Geriatric Unit at the City Hospital

The City Hospital is a general hospital for acute cases. It has 833 beds and a full complement of consultant staff, special departments, etc. There are two "acute geriatric" wards, one for men and one for women, each of 25 beds. To these wards are admitted medical cases of pensionable age (men over 65, women over 60), but there is no absolute age bar, and patients under pensionable age with long-term illness may be accepted. On the other hand, no attempt is made to deal in the geriatric wards with all elderly medical cases referred to the hospital. In the year reviewed 74% of the men but only 49% of the women of the total medical admissions to the hospital of pensionable age were admitted to the geriatric wards. The avoidance of any suggestion of a "closed shop" policy undoubtedly contributed to the general acceptance of the experiment.

The geriatric wards are equipped with parallel bars for walking exercises, several beds have Balkan beams with slings, and the simpler instruments of physiotherapy such as wax baths are available. The only other furnishings not usually found in general medical wards are specially designed armchairs. Each ward has a small day-room.

Medical, Nursing, and Ancillary Staffs.—A consultant physician is in charge and is assisted by a medical registrar (shared) and a house-physician. A psychiatrist visits weekly and the services of other consultants are available when required. Each ward has a sister or male charge nurse, a staff nurse, a variable number of student nurses, and four or five ward orderlies. The almoner prepares a brief case history for each admission, and this forms part of the patient's medical record. Before discharge each case is discussed with the almoner, who has interviewed patients and relatives. Physiotherapy in the aged requires encouragement rather than electricity, and most of the treatments given are in the form of walking and other remedial exercises. Under the guidance of a physiotherapist many of the early day-to-day treatments are given in the ward; later the patients attend the physiotherapy department. Other ancillary services include occupational therapy, speech therapy, and chiropody.

Out-patient and Follow-up Clinics.—The scope of the geriatric out-patient clinic is restricted because of difficulty in transporting elderly patients; also, many are referred to the general medical and surgical clinics for specialist's advice. Nevertheless, 30% of patients referred have been treated as out-patients. Patients are seen at intervals of three to six months after discharge from hospital. Elderly patients enjoy revisiting the hospital, and get a feeling of added security from the continued interest in their cases. Moreover, this service appears to be appreciated by their general practitioners. The cause of failure to keep a follow-up appointment is investigated by the almoner.

Admissions

All cases are initially admitted to an "acute" geriatric ward, with the exception of those requiring long-stay accommodation transferred from another ward or hospital. There are four channels of admission:

(a) *Emergency Admissions.*—The wards “receive” two days a week for medical emergencies of pensionable age. This relieves pressure on the general medical wards.

(b) *From Waiting-list.*—The waiting-list is kept by the records officer, and is compiled mainly from requests from general practi-

Diagram of the geriatric scheme in Nottingham and district. H=Hospital.
L.A.=Local authority.