LECTURE NOTES

Neurology

LIONEL GINSBERG

9th edition

WILEY-
BLACKWELL

Lecture Notes: Neurology

To my children
Amelia, Toby and Connie
and in memory of their mother
Andrea Marguerite Cobon

Lecture Notes

Neurology

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Preface to the ninth edition

The seventh edition of *Lecture Notes on Neurology* involved a complete revision of the text, an opportunity afforded by the change of author. With the eighth and ninth editions, there have been further refinements as follows:

- Selected case histories, drawn from life, are given at the end of each chapter in Part 2 of the book, so that important clinical points made in the main text can be illustrated and expanded.
- A series of sample examination questions and answers is given at the end of the volume. Desmond Kidd, Consultant Neurologist, and Thomas Solbach, Consultant Neuroradiologist, kindly provided figures to accompany some of these questions.
- Some of the figures in the main text have been replaced and a few new ones added.
- The text has been fully updated to reflect the continuing rapid advances in neurological management.

Tony Wilson and Charlie Davie, Consultant Neurologists, kindly read and commented on the case histories and examination questions, respectively. Susan Huson, Consultant Clinical Geneticist, commented on Table 18.3. Editorial staff at Wiley-Blackwell, notably Laura Murphy and Karen Moore, were patient and tolerant as always. Final thanks must again go to Sue, my wife, for her encouragement and support.

Lionel Ginsberg

Preface to the seventh edition

More than a decade has passed since the sixth edition of Dr Ivan Draper's *Lecture Notes on Neurology*. This seventh edition has been prepared with the dual aims of reflecting advances in neurology in the intervening period and changes in the undergraduate medical curriculum.

There have been dramatic developments in neurological practice in recent years, paralleling achievements in basic neuroscience research. These include new imaging techniques, which have greatly refined diagnostic accuracy and spared patients the discomfort of previous investigative approaches. Novel therapies are beginning to appear for conditions once considered untreatable. Molecular genetic research has cast new light on disease pathogenesis and should ultimately pay dividends in the treatment, as well as diagnosis, of inherited disorders.

Despite these advances, neurology remains *par excellence* a clinical discipline. Contrary to popular opinion outside the specialty, it is not a sterile and obscure diagnostic exercise. Neurological disorders are common, permeating the whole of general medicine and surgery. Their diagnosis is based on accurate history-taking and physical examination, coupled with the application of logical rules derived from knowledge of the underlying anatomy, physiology and pathology.

This volume is intended to emphasize these general principles. In line with the concept of an undergraduate 'core curriculum', it also concentrates on common diseases. The book falls naturally into two parts. The first section, 'The Neurological Approach', is concerned with history-taking and examination, where possible linked to relevant anatomy and physiology. A final chapter in this part outlines the expanding range of neurological investigations. The second section, 'Neurological Disorders', is a systematic account of the common conditions, rarities being relegated to the tables, or to a brief thumbnail sketch. There are also chapters on neurological emergencies, neurorehabilitation and the interface between neurology and other specialties. The separation of general from systematic is incomplete. For convenience, some disorders are discussed in the first part of the book and some principles appear for the first time in the second. The author also apologizes for occasions where his enthusiasm has allowed discussion of a few topics that might be considered beyond the range of a 'core curriculum'. Their inclusion may, it is hoped, be justified on grounds of continuity and interest, particularly where they reflect growing areas of neurological research and practice.

This new edition of *Lecture Notes on Neurology* should function as a portable companion during a student's neurology clinical attachment, and also senior medical clerkships. It may serve as a revision aid, and contains a grounding for early postgraduate work in general medicine.

Lionel Ginsberg

Acknowledgements

This work would not have been completed without the secretarial assistance of Barbara Parker, who uncomplainingly coped with numerous drafts and corrections. Many colleagues read selected chapters. I am grateful to Bob Bradford, Consultant Neurosurgeon, Heather Angus-Leppan, Jeremy Gibbs, John Hodges, Gareth Llewelyn, Gordon Plant, Tony Schapira, Neil Scolding, Tom Warner and Tony Wilson, Consultant Neurologists, for their comments. The responsibility for any remaining errors is, of course, my own. Several student reviewers at Blackwell Publishing also made helpful suggestions, which have been incorporated wherever possible. The editorial staff at Blackwell Publishing, particularly Mike Stein and Andrew Robinson, have been embodiments of patience and tolerance.

Most of the radiological figures were kindly provided by Alan Valentine, Consultant Neuroradiologist. Figure 10.1 was from the Department of Clinical Neurophysiology at the Royal Free Hospital. Pathological plates were prepared by Jim McLaughlin, Consultant Neuropathologist. The Department of Medical Illustration at the Royal Free Hospital helped provide most of the clinical photographs. Several other figures were prepared by Kieran Price. Finally, I thank my wife, Sue, who encouraged and supported me, and drafted most of the line drawings.

Lionel Ginsberg

Part 1

The Neurological Approach

Chapter 1

Neurological history-taking

The diagnosis and management of diseases of the nervous system have been revolutionized in recent years by new techniques of investigation and new treatments. But neurology continues to rely as much as any other branch of medicine on the fundamental clinical skills of history-taking and physical examination.

Neurological diagnosis

The neurological diagnosis is generally separable into two parts:

- Anatomical: What is the site of the lesion in the nervous system?
- Pathological: What disease process has occurred at that site?

This division is helpful as it can reduce possible confusion caused by the many available sites for neurological disorder (Table 1.1).

The history is of paramount importance in determining both the anatomical and pathological diagnoses. Indeed, many neurological patients have no abnormal signs, or simply have physical features that confirm clinical suspicions based on the history.

Sometimes, however, particularly with complex problems, the history can only yield a 'shortlist' of potential sites of the lesion(s) and final localization must await the formal examination. This is because disease at one site in the nervous system may produce symptoms mimicking a lesion at another.

Table 1.1 Potential sites of neurological disease.

Cerebral cortex
Cerebral white matter
Basal ganglia
Cerebellum
Brainstem
Cranial nerves
Spinal cord
Spinal roots
Peripheral nerves
Neuromuscular junction
Muscle

History of presenting complaint

How can the history best be taken to provide the maximum diagnostic information? An important rule is first to allow the patient sufficient uninterrupted time to speak. Most patients can give a reasonable account of their symptoms within 2 or 3 minutes and time spent listening at this stage is not wasted.

The nature of the main complaint and its duration will usually have been established in this early part of the interview, along with three further essential pieces of information about the patient:

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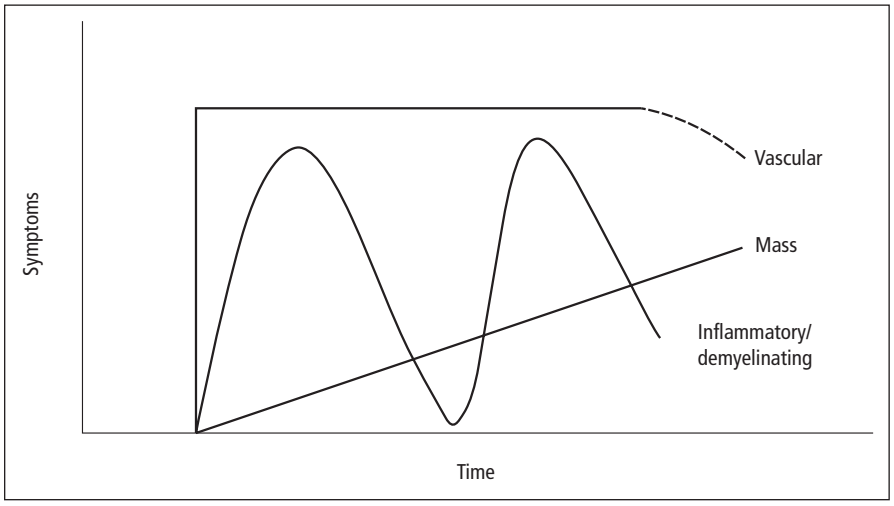

Figure 1.1 Temporal patterns associated with specific neuropathological causes. Using the example of a cerebral hemisphere lesion presenting with contralateral weakness, a rapid onset (seconds, minutes or at most hours) and static subsequent course, ultimately possibly with some improvement, suggest a vascular event (**stroke**), i.e. haemorrhage or infarction. A slowly progressive course (days, weeks or months) is more indicative of a **mass** lesion, i.e. a tumour. A relapsing and remitting pattern (with symptoms typically developing and resolving over days or weeks, then perhaps recurring with a similar time course) generally implies a **chronic inflammatory** or **demyelinating** process, of which multiple sclerosis is the prime example in the central nervous system.

- Age
  - Certain neurological disorders are associated with specific age groups.
- Occupation
  - A patient may have experienced occupational exposure to a toxin or other potential causative agent of disease.
  - Some neurological symptoms may limit the patient's ability to perform certain occupations.
- Handedness
  - To obtain information about cerebral hemisphere dominance.
  - To establish the extent to which a patient is disabled if the presenting complaint concerns the upper limbs.

Having heard the patient's description of the symptoms, it is usually necessary to probe the history of the presenting complaint in specific areas.

Timing of symptoms

Determining the temporal features of a patient's symptoms is essential to reach a pathological diagnosis:

- onset,
- progression,
- duration,
- recovery,
- frequency.

For example, a patient may present with weakness of one side of the body, suggesting a lesion in the contralateral cerebral hemisphere. Detailed further questioning on the timing of the symptoms may clarify the pathological nature of this lesion (Fig. 1.1).

'Discriminant' questions

If the initial history only partially solves the anatomical diagnosis, the 'shortlist' of potential sites may be reduced by asking the patient direct questions (Table 1.2).

For example, a patient presenting with numbness in both hands and both feet is likely to have a diffuse disorder of all the peripheral sensory nerves of the extremities (**sensory polyneuropathy**). But a similar '**glove-and-stocking**' sensory loss may occasionally be produced by

Table 1.2 Neurological systematic review questions: Has the patient suffered any of the following?

Pain
Headache
Facial, neck, back or limb pain
Disturbance of consciousness
Blackouts, faints, fits*
Altered sleep pattern
Cognitive and affective dysfunction
Memory, language
Depression, irritability
Cranial nerve symptoms
Loss of vision, blurred or double vision*
Hearing, sense of taste and smell
Vertigo, dizziness, giddiness*
'Bulbar' problems (swallowing, articulation of speech)
Limb symptoms
Difficulty in lifting, gripping, fine finger movements; clumsiness
Gait disorder, leg weakness or stiffness, balance problems
Loss of sensation, altered sensation, numbness*
Involuntary movements, incoordination
Sphincter disturbance
Bladder, bowel, sexual dysfunction

*If the patient uses terms like blackouts, fainting, dizziness, giddiness, double vision or numbness, it is worthwhile establishing their exact meaning, as the standard medical usage of the term may not correspond to the patient's intended meaning.

a **cervical spinal cord lesion**, mimicking a polyneuropathy.

In this instance, selecting questions from Table 1.2 likely to *discriminate* between these two anatomical diagnoses, a history of neck pain or injury will strongly favour the diagnosis of cervical cord lesion, as will the presence of sphincter dysfunction. Bladder disturbance is an early feature of spinal cord disease but only occurs in patients with a sensory polyneuropathy if there is a coexistent **autonomic** neuropathy.

Upper limit of symptoms

A useful further refinement in neurological history-taking is to check the '**upper anatomical limit**' of the symptoms. Thus, in a patient presenting with weakness of one leg the anatomical diagnostic range is wide. But specifically asking whether there are equivalent symptoms in the ipsilateral arm immediately narrows this range, the patient then being far more likely to have a **hemiparesis** caused by a lesion on the opposite side of the brain than anything else.

Negative and positive symptoms

A valid distinction may be made between 'negative' and 'positive' neurological symptoms.

Negative symptoms, or loss of particular functions, signify destructive lesions of the nervous system. Thus, a vascular event in one cerebral hemisphere will generally lead to loss of function as indicated, for example, by paralysis of the opposite side of the body.

Conversely, **positive symptoms** are those that suggest an irritative lesion, i.e. an area of abnormal excessive electrical activity in the nervous system. An irritative lesion in one cerebral hemisphere may produce repetitive involuntary (**clonic**) movements of groups of muscles on the opposite side of the body (**partial epilepsy**) rather than paralysis.

Remainder of the history

In neurology, as in other branches of medicine, valuable information, particularly about the pathological diagnosis, can be obtained by asking directly about:

- previous medical history,
- family history,
- social history,
- therapeutic history.

Considering again the patient who presents with glove-and-stocking sensory loss caused by a sensory polyneuropathy:

- **Previous medical history**: A history of diabetes mellitus would be especially relevant, this being a common cause of a sensory polyneuropathy.
- **Family history**: Some causes of a polyneuropathy are inherited.