Handbook for Differential Diagnosis of Neurologic Signs and Symptoms

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Preface

This handbook is designed to serve as a problem-oriented guide to the physician who is caring for patients with neurologic diseases. The traditional neurology texts are usually organized by disease states and their underlying etiology and pathology (eg, infectious diseases, neoplastic diseases, vascular diseases). Since the clinician frequently does not have a priori knowledge of the etiology of his patients' complaints, these books are of limited help in guiding him to the correct diagnosis. This problem-oriented text has been designed to assist the clinician through the deductive process needed to make an accurate diagnosis.

Unlike the traditional text, this problem-oriented guide does not review the natural history of the disease, the pathology, the prognosis, and many other aspects of specific diseases. After the clinician has made a diagnosis, it is important for him to know as much as possible about a specific disease. We would suggest that this handbook be used in conjunction with a standard neurology text. Finally, although we discuss treatment, prior to treating a patient with a pharmacologic agent the physician should acquaint himself with doses, indications and contraindications, side effects, and other aspects of the proposed treatment.

Contents

1
Episodic Disorders

HEADACHES

Headache is one of the most common neurologic complaints. Headache is defined as pain distributed in the upper region of the head (from orbits to suboccipital region).

In the head there are a limited number of pain-sensitive structures: (1) scalp and neck muscles; (2) blood vessels (eg, dural arteries, large arteries at the base of the brain, scalp vessels, dural sinuses); (3) nerves (including V, IX, X, and upper cervical nerves). These structures produce pain when they are stretched, compressed, dilated, or inflamed. In general, pain from intracranial structures above the superior surface of the tentorium is referred to the anterior portion of the head, and pain from intracranial structures below the tentorium is referred to the occipital and suboccipital regions.

There are five major types of headaches: (1) vascular headaches, (2) muscle contraction headaches, (3) traction headaches, (4) headache of cranial inflammation, and (5) extracranial headaches. In the evaluation of a patient with headache, it is important to diagnose which type of headache the patient is having; the differential diagnosis is outlined in Table 1. After diagnosing the major group, then consideration of the differential diagnosis within each group can be undertaken (Table 2).

A complete history and physical and neurologic evaluation should be performed for every patient with headache. Frequently it will be found that

further laboratory tests are not needed to make the diagnosis of migraine or tension headache. However, if one diagnoses tension headache and the pain is anterior, then one may want to obtain sinus films to help rule out sinusitis. Intraocular pressure measurements should be obtained to rule out glaucoma. In addition to sinusitis, muscle contraction headaches are commonly confused with traction headaches. If one wishes to rule out traction headaches, a computerized axial tomography (CAT) scan is the best screening test. In the absence of the apparatus necessary for a CAT scan, a brain scan and EEG should help exclude a mass lesion.

Recurrent migraine that always occurs on the same side may occasionally be caused by an arteriovenous malformation. A dynamic brain scan is a good screening test. If one suspects nonmigrainous vascular headache, then blood gasses and serum glucose should be obtained. Every patient should have blood pressure determinations. When vascular headaches are confused with other types of headaches, they are most frequently confused with headaches of cranial inflammation. If one suspects that cranial inflammation is producing a headache, then the patient should have a lumbar puncture (LP). Any patient with sudden onset of severe headache with or without loss of consciousness (without focal neurologic signs) should have an LP to rule out a subarachnoid hemorrhage. Patients over 50 years of age should have a determination of erythrocyte sedimentation rate to rule out temporal arteritis.

When possible, one should treat headaches of cranial inflammation (see Chap. 10), nonmigrainous vascular headaches, traction headaches (see Chap. 8), and extracranial headaches by treating the underlying disease. Treatment of tension headache and migraine headache is presented in Table 3. Prior to treatment the physician should familiarize himself with indications, contraindications, adverse effects, and dosages of the medications to be used.

SYNCOPE

Syncope is defined as a temporary loss of consciousness. Usually there is no permanent neuropathology associated with this loss of consciousness. Although syncope is frequently associated with self-limited and benign conditions, it may be a symptom of a serious underlying disease. Sudden loss of consciousness, in addition to being socially troublesome, may lead to serious injury. Therefore patients with syncope should be carefully evaluated. In the differential diagnosis of syncope, one first must be certain that one is dealing with syncope (no neuropathology), as opposed to the brief loss of consciousness that can be associated with seizures. Seizures often denote underlying neuropathology, and they must be worked up in a different

manner (see following section). The differential diagnosis between seizures and syncope is covered in Table 4.

After it is decided that one is dealing with syncope and not seizures, then the differential diagnosis of syncope (Table 5) must be considered. The history is of paramount importance. Unless a patient is having transient cranial nerve signs, it is difficult to make the diagnosis of basilar artery insufficiency. Syncope may be the only symptom of migraine, but frequently migraine is associated with other signs (see previous section). If syncope is being caused by a subarachnoid hemorrhage, there may be other signs such as headache (see previous section) or stiff neck. Lumbar puncture may be diagnostic. The history may also be helpful if the patient states that he passes out when he stands up quickly, has skipping of his heartbeat, or passes out after he turns his head, urinates, or coughs. A physical examination with special attention to the cardiovascular system will help to ascertain if syncope is being caused by a cardiovascular disease.

Laboratory studies such as chest x-ray, EKG, cardiac monitoring, blood glucose determination, and drug screens may also be helpful in determining the etiology of syncope.

Therapy should be aimed at the underlying disease.

SEIZURE DISORDERS

Seizures may be symptoms of (1) genetic predisposition to neuronal excitability, (2) metabolic abnormalities, or (3) focal pathology of the central nervous system. The classification of seizures can be found in Table 6. Frequently, seizures can be confused with syncope (see Table 4), and in children, seizures can be confused with breath-holding spells (see Table 7).

In each age group there are different diseases that may produce seizures. The principal causes of seizures in different groups can be found in Table 8. Frequently the clinician may need laboratory studies to help him in the differential diagnosis. An EEG is one of the most important tests in helping to differentiate the different types of seizures (see Table 9). Table 10 is a flowchart of how a patient with a seizure disorder should be evaluated.

In regard to treatment, there are six major principles: (1) Start with one drug. (2) Increase the dosage until the patient is either seizure-free or shows evidence of toxicity. (3) Add a second drug if necessary. (4) Change the dosage only after a trial lasting a minimum of 1 week. (5) Do not terminate medication unless the patient is seizure-free for at least 2 years. (6) It is also helpful if blood levels can be obtained. The drugs used in the treatment of seizures can be found in Table 11; their pharmacologic properties are listed in Table 12. The treatment of status epilepticus can be found in Table 13.

TABLE 1

Differential Diagnosis of Headaches

MANIFESTATIONS	VASCULAR HEADACHE	MUSCLE CONTRACTION HEADACHE	TRACTION HEADACHE	CRANIAL INFLAMMATION HEADACHE	EXTRACRANIAL HEADACHE
Laterality	Usually unilateral onset	Usually bilateral	Unilateral or bilateral	Bilateral	Unilateral or bilateral
Severity	Severe	Mild to severe	Usually mild	Severe	Mild to severe
Throbbing	Present at onset	Usually absent at onset, but may be present during peak	Usually absent	Present	Usually absent
Change with head position	Severe	Mild	Moderate	Severe	Mild
Time course	Acute	Subacute to chronic	Subacute to chronic	Acute	Acute to subacute
Gastrointestinal disturbance	Severe	Absent or mild	Moderate	Mild to severe	Absent
Visual disturbance	Present	Absent	May be present	May be present	Absent
Tenderness	Mild over extracranial vessels	Severe in suboccipital and temporalis muscles	Absent	Absent except in temporal arteries	Present in sinusitis
Focal neurologic signs	May be present	Absent	May be present	May be present	Absent
Stiff neck	Absent	Mild	Mild to severe	Severe	Absent

TABLE 2

Differential Diagnosis of Vascular Headaches

MIGRAINE HEADACHES

Classic migraine: contralateral neurologic symptoms (eg, hemanopsia, scotoma, amblyopia, paresthesia, numbness, weakness, speech disturbances) are followed by a unilateral throbbing headache that lasts several hours and is associated with nausea and vomiting. Frequently there is a strong family history

Common migraine: unilateral or bilateral throbbing headache without neurologic manifestations

Ophthalmologic migraine: ophthalmoplegia may occur after headache, which is on same side as eye findings

Cluster headache (histamine cephalagia, Horton's headache): severe unilateral pain that frequently arouses patient from sleep and is associated with unilateral lacrimation, conjunctival injection, ptosis, miosis, and nasal stuffiness; these headaches come in a series or in clusters, and pain is mainly in the eye and temporal regions

NONMIGRAINOUS VASCULAR HEADACHES

Changes in blood composition
- Hypercapnia
- Hypoxia
- Hypoglycemia
- Carbon monoxide

Drug-induced
- Nitrates
- Caffeine withdrawal
- Hangover

Other
- Post convulsion
- Post seizure
- Fever
- Hypertension

TABLE 3

Treatment of Migraine and Tension Headaches

TREATMENT	MIGRAINE HEADACHE	MUSCLE CONTRACTION HEADACHE
Vasoactive agents	Ergot preparations (eg, ergotamine tartrate)	Not useful
Analgesics	(1) Acetaminophen or (2) acetylsalicylic acid (ASA) or (3) Fiorinal or other analgesics	Same as migraine
Prophylaxis	(1) D/C oral contraceptives; (2) propranolol or (3) cyproheptadine HCl (Periactin) or (4) methysergide maleate (Sansert) or (5) low-tyramine diet	Muscle relaxants, eg, diazepam (Valium), or mood elevators, eg, amitriptyline HCl (Elavil)
Behavioral-Mechanical	Biofeedback, psychotherapy	Biofeedback (EMG), relaxation exercises; psychotherapy
Other	For nausea and vomiting, perchlorperazine (Compazine) or trimethobenzamide (Tigan)	—

TABLE 4

Differential Diagnosis of Seizures and Syncope

	SEIZURES	SYNCOPE
Motor activity	Tonic rigidity, clonic activity, mouth movement, automatic behavior	Usually limp without movements
Injuries	Tongue-biting and injuries secondary to clonic activity	Injuries secondary to fall
Incontinence	Present with major motor seizure	Usually absent
Cry	Usually at beginning of major motor seizure	Absent
Changes in respiration	Usually associated with tonic-clonic activity	May increase or decrease with syncope caused by changes in blood composition
Pulse	Usually increased	May decrease or may be irregular
Postictal lethargy	May be present	Absent
Postictal neurologic sign	May be present	Absent
EEG	May be abnormal	Should be normal (when patient is not unconscious)

TABLE 5

Principal Causes of Syncope

CENTRAL NERVOUS SYSTEM
- Hysteria

LOCAL ISCHEMIA OF CENTRAL NERVOUS SYSTEM
- Basilar artery insufficiency
- Migraine
- Subarachnoid hemorrhage

GENERALIZED FALL IN BLOOD PRESSURE
- Vasovagal
- Carotid sinus
- Aortic valvular disease
- Myocardial infarction
- Pulmonary embolus
- Dissecting aneurysm
- Orthostatic hypotension
- Arrhythmia
- Atrial myxoma
- Cough syncope
- Urination syncope

CHANGES IN BLOOD COMPOSITION
- Hypercapnia
- Hypoglycemia
- Hypoxia
- Hypocapnia
- Drugs

TABLE 6

Classification of Seizures

PARTIAL SEIZURES

The focal clinical features are from activation of a specific group of neurons with a correlative focal EEG abnormality. These phenomena may remain focal or may become generalized, and there is usually an underlying pathologic substrate. The onset may be at any age, and there may be a focal abnormality on routine examination or a postictal focal abnormality

With elementary symptoms: usually no generalization and therefore no loss of consciousness
- Motor (Jacksonian, adversive, aphasic, etc)
- Sensory (somatic. visual, auditory, olfactory, vertiginous, etc)
- Autonomic (rare)
- Compound (combined elementary and/or complex symptoms)

With complex symptoms: corresponds to temporal lobe (psychomotor) epilepsy that usually leads to impaired consciousness; may have elementary onset
- Only impaired consciousness
- Cognitive (déjà vu, forced thinking, etc)
- Affective
- Psychosensory (hallucinations, macropsia, etc)
- Psychomotor automatisms
- Combinations

Partial seizures secondarily generalized: usually tonic-clonic seizure developing from a partial seizure

GENERALIZED SEIZURES

These are without local onset. There is usually initial loss of consciousness with generalized motor findings and an EEG correlate of bilateral synchronous discharge

Primary: absence of etiology and presumed genetic in origin; examination usually normal; onset usually in childhood or adolescence, with persistence to adulthood
- Absence (petit mal)
  - Simple: only impaired consciousness
  - Complex: impaired consciousness with one or more of the following:
    - Mild clonic movements (myoclonic absences)
    - Increased postural tone (retropulsive absence)
    - Decreased postural tone (atonic absence)
    - Automatisms (automatic absence)
    - Autonomic (eg, enuretic)
    - Mixed
- Grand mal
- Myoclonus (massive bilateral)

Secondary: Caused by diffuse cerebral disease with abnormal exam.
- Atypical petit mal
- Tonic-clonic (grand mal)
- Atonic
- Massive bilateral myoclonus

UNILATERAL

Presents with clinical features restricted to one side of the body with EEG discharge over the contralateral hemisphere. These may be tonic, clonic, or tonic-clonic, with or without impaired consciousness, may shift sides, but do not become symmetrical.

TABLE 7

Distinguishing Features of Some Paroxysmal Events in Children

FEATURE	PETIT MAL	PSYCHOMOTOR	BREATH-HOLDING	FEBRILE CONVULSIONS
Age at onset	4–10 years	Older child, adolescent	3 months to 4 years	Less than 6 years; most occur between 6 months and 3 years
Frequency	Frequent, several per day	Variable, many per day followed by days or weeks free from seizures	Infrequent, but increase to about age 4, when they cease	Infrequent, occurs soon after onset of a fever
Precipitating factor	Hyperventilation, hypoglycemia	Sleep, drugs (eg, Brevital, Thorazine)	Always present (eg, minor injury or emotional upset)	Extracranial infection with fever
Crying	Never	May occur as part of aura	Always precedes	Not precipitating factor
Cyanosis	Never	Never	Always precedes loss of consciousness rather than occurring with loss of consciousness or during major motor convulsive activity	No
Aura	No	Yes	No	No
Tone changes or postictal motor movements	Normal tone, but with eyelid, eyebrow, and hand twitching; rarely, atonic, with falling to floor	Eyelid blinking or fluttering; complex automatisms (eg, lip-smacking, chewing, fumbling); motor onset	Opisthotonos	Grand mal
Postictal lethargy	No	Yes	Sometimes	Yes
Duration	Less than 30 sec	From seconds to several minutes	About 1 minute for consciousness and/or clonic movements	Very brief, to minutes
EEG	Specific	Abnormal, usually focal temporal; may need nasopharyngeal or sphenoidal electrodes to visualize discharge	Normal between events	Normal 1 week after seizure