Neurology

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1 Introduction

This chapter on neurology will have a focus on the emergencies associated with damage to the brain tissue either by ischemia, hemorrhage, or inflammation. Transient ischemic attacks (TIAs) and cerebrovascular accidents, both embolic and hemorrhagic, will be covered in detail. Traumatic brain injury and its management from the primary care setting will be discussed. Infection of the central nervous system, meningitis and encephalitis, will be briefly reviewed. Other neurologic diseases with potentially emergent presentation such as multiple sclerosis and myasthenia gravis are covered. The approach to seizures in the primary care setting will be addressed. Miscellaneous topics such as Guillain–Barre syndrome, transverse myelitis, and Wernicke's encephalopathy conclude the chapter.

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2 Transient Ischemic Attack

Transient ischemic attack (TIA) is a transient occlusion of a cranial artery resulting in temporary neurologic deficits.

2.1 General Considerations

Particular attention must be paid to TIAs, as primary care clinicians will encounter this multiple times in practice. Approximately 500,000 TIAs occur annually and 20–25% of cerebrovascular accidents (CVA) are preceded by a TIA. Proper identification and management of TIA prevents significant morbidity and mortality. Previously, the clinical distinction between TIA and CVA was time related, with TIAs resolving after minutes to several hours, and CVAs taking hours to days to resolve, if at all. A TIA, according to new guidelines, is now diagnosed solely based on the history of symptoms in the absence of ischemic changes on imaging. A TIA is a medical emergency as the average risk of stroke is 5% within 7 days and a 10–15% risk or higher within 90 days. Management can be challenging, and some clinicians feel that all with TIA-like symptoms should be evaluated in the ED. The ABCD2 criteria provide an estimated risk of stroke within 48 hours (Table 1). On a seven-point scale, 0–3 points is associated with a risk of 1%, 4–5 points is associated with a stroke risk of 4.1%, and 6–7 points is associated with a risk of 8.1%. In the ABCD2 score, 1 point is given each to (A)ge equal to or greater than 60, (B)lood pressure equal to or greater than 140/90, a (C)linical feature of speech disturbance, a (D)uration of 10–60 min and (D)iabetes. Two points are given for a (C)linical feature of unilateral weakness or a (D)uration of more than 60 min. This ABCD2 criteria can help triage those patients with possible TIA and make a shared decision in the determination of ultimate disposition.

Table 1 The ABCD2 score is formulated to calculate the subsequent risk of a stroke after a TIA

ABCD2 score		Points
Age	≥60 years	1
Blood pressure elevation	Systolic >140 mmHg and/or diastolic ≥90 mmHg	1
Clinical features	Speech disturbance without weakness	1
	Unilateral weakness	2
Duration of symptoms	10–59 min ≥60 min	1 2
Diabetes	Type 2	1

Score	2-day risk (%)	7-day risk (%)	Risk of stroke
0–3	1.0	1.2	Low
4–5	4.1	5.9	Medium
6–7	8.1	11.7	High

Johnston, S.C., et al. *Lancet*. 2007

2.2 History

The history and exam are closely linked in the evaluation of TIA and CVA. In embolic CVAs and TIAs, intermittent symptoms indicating a TIA should be sought, such as motor weakness, sensory symptoms (i.e., numbness and tingling, especially on the unilateral face or circumoral region), and the inability to speak or understand. "Dizziness" is a special concern associated with posterior circulation TIA or CVA (ischemic central vertigo); however, it is most commonly reported with conditions other than central vertigo such as inner ear pathologies (peripheral vertigo see Chap. 8 "Otolaryngology"). Unlike peripheral vertigo, central vertigo is generally not position related and lasts longer than just several minutes. This rule is not hard and fast, as turning of the head can exacerbate central vertigo. However, in central vertigo, this effect is long-lasting, on the scale of hours, unlike the brief and intermittent episodes seen with peripheral vertigo. TIA and CVA involving the posterior circulation often evolve over 2–3 days in contrast to the sudden presentation in other CVA subtypes. Usually, a TIA or CVA of the posterior system is accompanied by other symptoms and signs besides just dizziness such as dysarthria and dysphagia and cranial nerve palsies. Very rarely, hearing can be affected in these TIAs and CVAs.

Another relatively common complaint is double vision. Painless binocular double vision can be caused by a hemorrhage or infarct in the brain stem or by potentially emergent cranial nerve palsies (see "Exam" section below under stroke). A partial loss of vision, described as a shade coming down over the eye, is called amaurosis fugax and is a form of TIA (see Chap. 12 on Ophthalmology). Visual changes can also be ischemic (see under CVA below). Seizures (see Sect. below) and migraines are sometimes confused with TIAs. In seizures, a common prodrome is the sense of epigastric rising or other auras. In migraines, patients may describe numbness or weakness beginning in one hand, gradually spreading up the arm and then spreading to the face, trunk or elsewhere, and this history is virtually diagnostic. However, migraine is a diagnosis of exclusion, and a workup for TIA is often necessary in patients without a past history of migraine.

2.3 Exam

By definition, a TIA will have no symptoms at the time of examination in the clinic. Carotid bruits should be assessed to account for possible carotid disease.

2.4 Diagnostics

An EKG should be obtained on patients with TIA though the yield is low with a normal cardiovascular examination. MRI/MRA of the brain and neck is the imaging of choice for suspected TIA. A carotid ultrasound is an option, if a TIA in the carotid distribution is suspected. Cardiac monitoring should be promptly arranged.

2.5 Therapeutics

All patients with a history of possible TIA should receive aspirin 325 mg. Clopidogrel 75 mg may be added while awaiting studies, however, this may delay surgery in carotid disease. The clopidogrel should be stopped after 3–4 weeks and the aspirin continued.

Key Points

- If a clear instance of TIA is encountered within 48 hours, it is reasonable to pursue expedited workup in the ED. This is especially true in those with a high ABCD2 score (see Table 1).
- Shared decision making may be used. If outpatient workup is pursued, aspirin and clopidogrel may be used.

3 Cerebrovascular Accident (Embolic and Hemorrhagic)

An embolic stroke or cerebrovascular accident (CVA) is caused by blockage of a cranial vessel that results in tissue damage and neurologic deficits.

3.1 General Considerations

CVAs are common; 200,000 cases are reported each year with 75% of cases being embolic. CVA in younger adult populations (20–40) is rare, but can occur due to the following unusual conditions:

- Fibromuscular dysplasia
- Cervico-vertebral dissection
- Hypercoagulable states, especially pregnancy (i.e. central venous thrombosis) and antiphospholipid syndrome
- Systemic embolization from a patent foramen ovale (paradoxical embolism)
- Posterior reversible encephalopathy syndrome (PRES) (a stroke-like syndrome with a characteristic radiographic appearance that is associated with eclampsia, post-partum, and migraines)
- Migraines (especially when oral contraceptives are used)
- Cocaine use (ischemic and hemorrhagic CVA).

From age 40 onward, the risk factors for stroke become those for other atherosclerotic processes such as hypertension, hyperlipidemia, diabetes, and smoking, and age. From 60 years old and onward, cardiac causes begin to play a role (approximately 15% of all CVA). Atrial fibrillation accounts for most of these strokes, but ventricular embolization associated with recent myocardial infarction and cardiomyopathy is also known to occur. Rare inflammatory and infectious etiologies, as

seen in vasculitis and endocarditis, can be found in all ages but skew toward older populations. Although guidelines are in flux, treatment with tissue plasminogen activator (TPA) must be started within 4.5 h; however, thrombectomy can be performed up to 24 hours later.

3.2 History

TIA and CVA are closely related with only the distinction of time and tissue damage defining them (see above). While in TIA the history is paramount, the exam drives the evaluation of CVA. However, the history deserves special consideration in some stroke syndromes. The symptoms of hoarseness, difficulty swallowing, nausea with or without vomiting plus dizziness with abnormal gait are typical of a cerebellar infarct called Wallenberg's (or Lateral Medullary) syndrome. A history of trauma is usually reported for cervico-vertebral dissections. Often a car accident is the precipitating factor, but sometimes even minor, trivial stress on the neck can be the cause. Pain is often reported in the ipsilateral neck, face, or head in the case of a carotid dissection, or the back of the neck or head with a basilar artery dissection. The patient may report a sound suggestive of a bruit in the ipsilateral neck. Hoarseness can also occur due to dysfunction of the ninth cranial nerve within the carotid sheath. In cases of central vein thrombosis, a headache, which can be severe, is often reported.

3.3 Exam

A discussion of the full neurological exam is beyond the scope of this book but will be covered briefly in this section. The presence of carotid bruits should be ascertained. Horner's syndrome (miosis—decreased pupil size, ptosis, and decreased sweating) is often seen with carotid dissections. Close examination of the pupils (sometimes best accomplished in a dark room) and extraocular movements should be performed (cranial nerves 2–6). Facial findings (seventh cranial nerve) such as furling of eyebrows, puffing of the cheeks, and showing of teeth should be examined. Sudden foot drop and shoulder weakness can be a syndrome of anterior cerebral artery occlusion. Facial droop is a common sign of stroke and should always be investigated. Occlusion of the middle cerebral artery can elicit various syndromes, and focal weakness can be found in the upper or lower extremity on one side. If a patient cannot stand upright with feet close together and eyes open, this is a possible posterior CVA, especially in the absence of a clinical picture of severe peripheral vertigo. The HINTS exam (head impulse test, nystagmus, and test of skew) can be used to differentiate peripheral from central vertigo. Sudden altered mental status and inability to look up with bilateral ptosis is caused by an embolus to the basilar artery. Several visual syndromes are also associated with

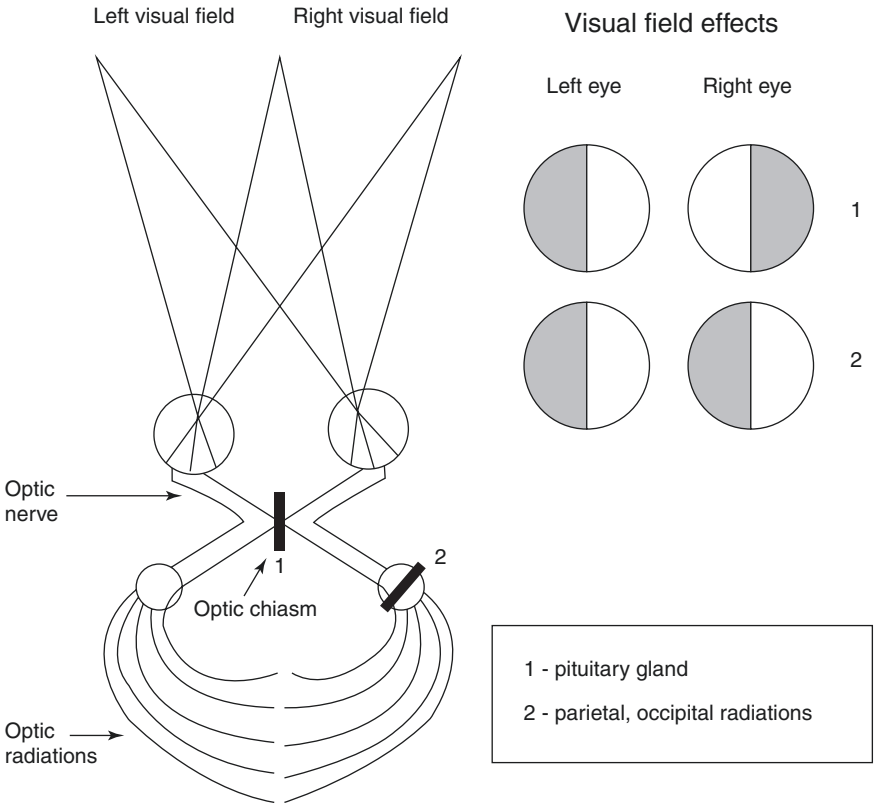

Fig. 1 1. Bitemporal hemianopsia. Bitemporal hemianopsia is usually caused by a lesion of the pituitary at the optic chiasm 2. Homonymous hemianopsia. Homonymous hemianopsia is caused by an occlusion or disruption in the parieto occipital radiations

CVA. Bitemporal hemianopsia (see Fig. 1) is associated with pituitary hemorrhage and apoplexy (see Chap. 10 on Endocrinology). Homonymous hemianopsia (see Fig. 1) can be associated with vascular disease in the parietal or occipital lobes; sometimes this finding is acute and noticed by the patient, depending on where the lesion is located.

Binocular diplopia, confirmed by absence of double vision with each eye covered sequentially, should prompt consideration of referral to the ED if acute. The exception is an incomplete third nerve palsy (sparing the pupil), almost always associated with diabetic patients. This benign diabetic cranial mononeuropathy is associated with pain one-half of the time.

Key Points

- In patients without obvious peripheral vertigo and who have trouble standing with feet together and eyes open, vertebral basilar stroke or other emergency is a strong consideration.

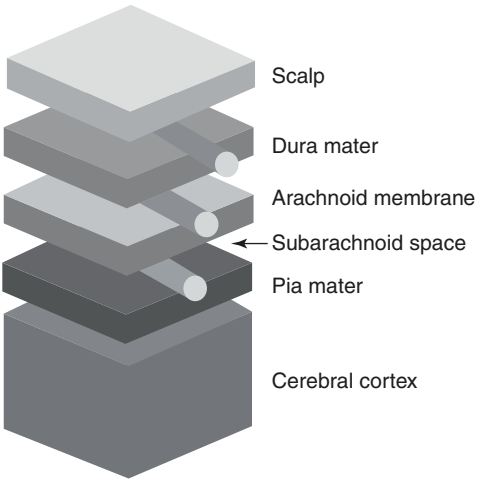

Fig. 2 Layers of the meninges

- Painless binocular double vision is usually an emergency unless explained by an incomplete third nerve palsy (see above), myasthenia gravis, or multiple sclerosis (see below).

3.3.1 Hemorrhagic Cerebrovascular Accident

A hemorrhagic cerebrovascular accident (CVA) causes neurological deficits, usually focal, by bleeding within the central nervous system (CNS). Hemorrhagic CVAs may be intracerebral (ICH), subarachnoid (SAH), subdural (SDH), or epidural (see Fig. 2).

3.4 General Considerations

Hemorrhagic CVA accounts for approximately 25% of the 200,000 CVAs that occur each year. Use of anticoagulants and, to a lesser degree, antiplatelet agents, is a risk factor for hemorrhagic CVA. Usually, but not always, the patient is older than the age of 35. The majority are ICH, followed by SAH and then SDH. A major identifiable risk factor in ICH is hypertension in addition to alcoholism. SDH and epidural hematoma is typically associated with trauma. Cancer patients can have metastatic CNS tumors (usually intracerebral) which are hemorrhagic. The neoplasms most commonly involved are melanoma, renal cancer, and lung cancer. Arteriovenous malformations (AVM) are commonly seen with SAH but also ICH and SDH. However, the most common cause of SAH is aneurysms. SAH is rare accounting for only 1% of ED visits for headache. Cocaine is often implicated in

hemorrhagic stroke and patients under the age of 40 and first-time users can be affected. Trauma (traumatic brain injury or TBI) is another common cause of ICH and SDH and will be addressed in the next section (see below).

3.5 History

Family history of hemorrhagic CVA can be found, most often with SAH due to aneurysm. Patients with adult polycystic kidney disease and hemophilia are at risk for hemorrhagic CVA. Inquiries should be made concerning alcohol and cocaine use. SAH often causes a sudden loss of consciousness that may be preceded by or followed by a severe headache, thus up to 6% of syncope is caused by stroke. Thunderclap headache is a severe headache that reaches its maximal intensity in less than 10 s and should be referred to the ED immediately. "Worst headache of my life" is a historical question that can elicit a history leading to a diagnosis of SAH. Associated symptoms of nausea (with or without vomiting) and photophobia are commonly associated with hemorrhagic CVA and should prompt immediate imaging. Headaches occurring during heavy exercise and sexual intercourse are common complaints in subarachnoid hemorrhage (SAH), but SAH symptoms can also occur at rest. A headache associated with altered mental status could be a hemorrhage and should be sent to the ED. Headaches associated with sudden changes in visual acuity or bilateral double vision are concerning for an emergency. If there is pain in the eye, this is generally not a neurologic condition, as the structures in the eye from the retina moving posterior are not pain sensitive. Headaches associated with paresthesias especially unilateral (does not apply to migraineur) are concerning and a stat CT should be obtained.

3.6 Exam

The neuro exam outlined above for embolic CVA also applies to hemorrhagic CVA. Some exam specifics are notable for hemorrhages and aneurysm. Cushing's sign (hypertension with bradycardia) may be present, but it is a late and unreliable finding of increased intracranial pressure. A fixed dilated pupil can be a sign of cerebral herniation and, if both pupils are affected, the brainstem may be involved. Neck stiffness (nuchal rigidity) or neck tenderness (less specific) can be seen with hemorrhagic CVA. Photophobia to pen light exam may be prominent. Complete third nerve palsy (medial eye movement disruption with dilation of the pupil) is a posterior communicating aneurysm until proven otherwise. An isolated sixth nerve palsy can be seen with increased intracranial pressure. Bitemporal hemianopsia can be seen in disruptions of the pituitary (see Fig. 1). As with embolic stroke, homonymous hemianopsia can be seen with disruption of the parietotemporal lobe or occipital lobe (see Fig. 1).

3.7 Diagnostics

A CT of the head without contrast is the diagnostic test of choice. In the case of SAH, the CT is 80% sensitive. If suspicion is high, the patient should be sent to the ED for a lumbar puncture to increase the sensitivity of the evaluation.

Key Points

- A thunderclap headache or "worst headache of my life" are descriptions of a potential emergent neurologic condition.
- Nausea, photophobia, or neck pain may be present.
- Any headache associated with loss of consciousness, altered mental status, or visual symptoms (unless migraineur) should be considered for an ED referral.

4 Traumatic Brain Injury

Traumatic brain injury (TBI) is a closed head injury resulting in damage to the cerebral cortex and incorporates the term concussion but is not synonymous with it.

4.1 General Considerations

Approximately, 1.5 million people in the U.S. suffer a TBI annually. A TBI can cause hemorrhage within any layers of the cranium (see Fig. 2). In the primary care setting, most of the presentations will be after the fact, and the clinician must discern between common sequelae following head injury that are expected, versus symptoms and signs that need or may need immediate attention. Loss of consciousness and amnesia should always be assessed but are not always present with a TBI that necessitates emergent care. Despite this caveat, the Canadian head CT rule has been developed to give guidance on which patients need an urgent or emergent head CT (see Table 2).

The Canadian Head CT rule applies to those who have lost consciousness and have amnesia that lasts less than 30 min after the event. The Canadian Head CT rule does not apply to any patients with a seizure, on blood thinners, or age less than 16 years old. The Canadian Head CT rules state that a patient with a brief loss of consciousness or amnesia (less than 30 min), and with (1) only 1 or no episodes of vomiting, (2) age less than 65 years old, (3) no signs of skull fracture (see below), and (4) a normal Glasgow coma score at 2 h, does not need an emergent head CT. A normal Glasgow score is characterized by a patient with their eyes open spontaneously, alert and oriented (normal verbal responses), and able to follow simple commands. Two other risk factors were scored as medium risk: (1) retrograde amnesia to the event of greater than 30 min and (2) a dangerous mechanism such as a fall greater than 3 feet or greater than five stairs. Medium risk factors warranted consideration of CT depending on clinical situation.

Table 2 Canadian Head CT rule for traumatic brain injury (TBI)

High risk (for neurosurgical intervention)
GCS score <15 at 2 h after injury
Suspected open or depressed skull fracture
Any sign of basilar skull fracture[a]
Vomiting ≥2 episodes
Age ≥65 years
Medium risk (for brain injury on CT)
1. Amnesia before impact ≥30 min
2. Dangerous mechanism[b] (*pedestrian, occupant ejected, fall from elevation*)

[a]Signs of basilar skull fracture
- Hemotympanum, "raccoon" eyes, CSF otorrhea/rhinorrhea, Battle's sign

[b]Dangerous mechanism
- Pedestrian struck by vehicle
- Occupant ejected from motor vehicle
- Fall from elevation ≥3 feet or 5 stairs

Rule not applicable if:
- Non-trauma cases
- GCS <13
- Age <16 years
- Coumadin or bleeding disorder
- Obvious open skull fracture

CSF cerebrospinal fluid, *GCS* Glasgow Coma Scale
Stiell, I.G., et al. *Lancet* 2001

4.2 History

The patient will report a headache. Nausea and vomiting are always cause for concern that a hemorrhage has occurred. Of note, the post-concussive syndrome is common following a head trauma and does not require imaging though the patient may be very distressed by these symptoms and their long-lasting nature. Somatic symptoms of the post-concussive syndrome are headache, sleep disturbance, vertigo, nausea, fatigue, and sensitivity to noise or light. Cognitive symptoms affecting memory and attention as well as affective symptoms such as anxiety, depression, and irritability may also occur. As disturbing to patients as these symptoms are, especially when lasting up to 3 months, (found in 80% of concussion patients) or longer (up to a year in 15% of patients), these symptoms do not require any additional emergent imaging.

4.3 Exam

Cushing's sign (hypertension with bradycardia) may be present, but is a late and unreliable finding indicating increased intracranial pressure. A fixed dilated pupil can be a sign of cerebral herniation; and, if both pupils are affected, the brainstem may be involved. Raccoon eyes, Battle's sign (retroauricular ecchymosis), and hemotympanum are signs of basilar skull fracture. Cerebrospinal fluid (CSF) leak, resulting in otorrhea or rhinorrhea, may also signal a basilar skull fracture. The rest

of the neurologic examination is similar to that seen with CVA, as hemorrhage is the major concern with the TBI patient.

Key Points

- If the encounter is soon after the occurrence of injury and criteria are not met for a CT, ensure that the patient is able to be closely observed for 24 hours in the outpatient setting.
- Loss of consciousness or anterograde amnesia are not necessary for severe brain trauma and the clinical rules are meant to provide guidance and do not outweigh judgment.

5 Bell's Palsy

Bell's palsy is unilateral damage to the facial nerve resulting in a syndrome that is easily confused with CVA.

5.1 General Considerations

Idiopathic facial nerve palsy (or Bell's palsy) is not an emergency but presents very similar to a CVA. Approximately, 80,000 cases of Bell's palsy occur every year. The etiology of Bell's palsy is thought to be viral and patients are usually treated with antiviral medications and steroids though the evidence for this is not strong. In endemic areas, Lyme disease has also been implicated. If function of the facial nerve has not begun to recover after 4 months, the patient should be imaged and reevaluated.

5.2 History

The onset of Bell's palsy is acute usually over a day or two and causes decreased tearing, loss of taste or hearing and sagging of the face on the affected side. More than one cranial neuropathy can occur which can be confusing to the clinician and will generally require prompt imaging.

5.3 Exam

The hallmark of the exam is dysfunction of the peripheral facial nerve involving eye closing, cheek puffing, and mouth closing on one side PLUS interruption of forehead wrinkling. (A unilateral central lesion usually spares the function of the

forehead muscles due to the dual innervation of the forehead muscles). However, if the forehead is spared, this does not always imply a central lesion.

Key Point

- A lesion that affects the function of the peripheral facial nerve (including the action of the forehead muscles) is peripheral and does not constitute an emergency.

6 Meningitis and Encephalitis

CNS infections cause inflammation of the tissues of the CNS and are often caused by various microorganisms. These infections can be fatal.

6.1 General Considerations

CNS infections are not the only cause of meningitis as medications, vasculitis, and miscellaneous etiologies exist. However, the primary care practitioner should be aware that these are diagnoses of exclusion and vigilance for infectious causes must be maintained. Viral meningitis is common with enteroviral meningitis alone causing 75,000 infections every year. The incidence of bacterial meningitis is less with 10–15,000 cases per year, resulting in 2000 deaths. Encephalitis is equally as rare with only 20,000 cases reported each year. Risk factors for bacterial meningitis include previous neurosurgical procedures (i.e., shunts) or skull fracture resulting in CSF leak. Bacterial meningitis can also spread contiguously from infections such as sinusitis, otitis media, and mastoiditis. Systemic risks are asplenia, sickle cell disease, post-transplant status, cancer, and HIV. Alcoholism and cirrhosis are also risk factors. Meningitis can spread to the surrounding brain tissue and cause meningoencephalitis. This is more common in the aseptic causes of meningitis caused by viruses, fungi, and other bacterial classes such as rickettsia and spirochetes, such as neurosyphilis. Neurosyphilis is seen with tertiary syphilis and requires intravenous antibiotics. Encephalitis is a diffuse infection of the brain parenchyma causing vague, generalized symptoms with insidious onset. Seizures may occur. Viral encephalitis is more common in the summer months in endemic areas for arthropod borne (arbo) viruses. Organisms such as West Nile virus, Eastern and Western Equine viruses, and St. Louis encephalitis virus are common. The treatment for encephalitis is supportive except for cases of herpes virus which can be treated with intravenous (IV) acyclovir. Immediate treatment with IV acyclovir is necessary in all encephalitis patients to prevent morbidity and mortality as assays for herpes virus take time to complete.

6.2 History

The classic signs of CNS infections such as headache, fever, neck stiffness, photophobia, nausea, and vomiting may be absent or incompletely presented, especially in the elderly who present with listlessness and other vague complaints. Altered mental status due to cerebral edema and increased intracranial pressure is often present and is more common in diffuse infections such as encephalitis. Cerebral edema in encephalitis can lead to the characteristic vague symptoms such as drowsiness, irritability, and delirium, which is often accompanied by fever. Subtle cognitive and psychiatric symptoms may also be present. In brain abscesses, onset can be subacute with the presence of subtle symptoms for up to 8 weeks. Herpes Simplex 2 meningitis associated with genital infections is generally a benign entity and is often recurrent. However, in severe cases, genital-associated HSV meningitis may need to be treated with IV instead of oral antivirals. Sexual history may prove useful as tertiary syphilis may present without a prior history of primary or secondary syphilis.

6.3 Exam

Fever may be present. Focal neurologic signs may be present and a full neuro exam should be performed. Nuchal rigidity may be present (30%) in bacterial meningitis and is best elicited with the patient lying supine and the head passively flexed. Meningeal signs such as Kernig and Brudzinski's sign may be present but are often a late finding and rare (5%). Kernig's sign is pain in the back or contraction of the hamstrings with flexing the hip and extension at the knee. Brudzinski's sign is flexion of the hips and knees with passive flexion of the head. Petechial or ecchymotic rashes on the extremities are common for meningococcal CNS infections and meningoencephalitis from Rocky Mountain Spotted Fever (see Chap. 2 on "Infectious Diseases" under "Febrile Traveler").

Key Points

- Classic symptoms and signs of meningitis are often absent.
- Bacterial meningitis usually has a rapid onset (within 48 hours).
- Viral meningitis usually has a more insidious onset, but when the two cannot be differentiated, the patient should be referred to the ED for a lumbar puncture.
- Patients suspected of encephalitis must be referred immediately to the hospital for treatment with IV acyclovir.

7 Spinal Cord Compression

Spinal cord compression usually comes from the epidural space of the spinal CNS, most commonly from malignant or infectious causes.

7.1 General Considerations

About 20,000 cases of spinal cord compression happen every year due to cancer. Despite its rarity, the clinician must maintain vigilance when dealing with back pain, as early intervention in cases of cord compression can mitigate permanent disabling injury. Malignant metastasis to the spine is of great concern in patients with a history of active or remote cancer and is most common with breast, lung, or prostate cancer. Epidural abscess is a rare condition and up to half of these infections originate from hematogenous spread of bacteria from soft tissue, urinary or respiratory tract infections. Other risk factors for abscess are immunosuppression including diabetes, IV drug abuse and recent procedures such as surgery, lumbar puncture or epidural anesthesia. Hematoma is another rare cause and awareness of this entity should be high with patients on anticoagulation. Another cause of compression to the central or peripheral nervous system of the spine includes disc disease which is common and can also necessitate urgent intervention.

7.2 History

Back pain is present in 90% of epidural metastases and 70–90% of cases of epidural abscesses. Unlike the back pain with musculoskeletal causes, there is no positional component and the patient will not likely relate an inciting mechanism. Inability to find a comfortable position is often related. Subjective fevers are an ominous sign when occurring with back pain. The development of radicular syndromes is the first symptom of cord compromise and is followed by fecal or urinary incontinence or sexual dysfunction. Motor weakness and paralysis in the lower extremities occurs in the later stages. Saddle anesthesia indicative of cauda equina syndrome can be seen and is considered an emergency.

7.3 Exam

Searching for localized tenderness over the midline spine is of great importance. Motor weakness should be sought for with heel and toe walking and squatting on one leg. Hyperreflexia and Babinski sign indicates upper motor (CNS) neuron involvement. Cauda equina syndrome findings with decreased rectal tone (sensitivity of 60–80%) and decreased perineal sensation (75% sensitivity) should be assessed.

7.4 Diagnostics

In unclear cases of epidural abscess, CBC, ESR, and CRP may be ordered in addition to an MRI of the spine which should be obtained within 24–48 hours. Leukocytosis may be absent, however inflammatory markers should be elevated but

are a non-specific finding. Findings on plain films can lag by up to 2 months so MRI is the test of choice.

Key Points
- Back pain in the midline on the spinal processes is concerning for secondary back pain.
- Unexplained back pain in a patient with a history of cancer is concerning.
- Fever and back pain are ominous signs of epidural abscess.
- In unclear cases of epidural abscess inflammatory markers can be helpful as can MRI.
- Back pain associated with muscle weakness needs urgent attention.

8 Multiple Sclerosis

Multiple sclerosis (MS) is a disease of neurologic dysfunction caused by autoimmune destruction of the myelin sheath of nerves in the central nervous system (CNS).

8.1 General Considerations

MS is extremely common, affecting 350,000 people in the U.S. and is the most common cause of neurologic disability in the young. MS is twice as common in females as it is in males. MS is defined by multiple discrete synchronous lesions in the CNS, which cause a wide variety of symptoms and exam findings. By definition, there must be more than one CNS lesion and more than one exacerbation to make the diagnosis. Usually the presentation is subacute; however, sometimes the presentation is characterized by abrupt focal signs and resembles a CVA. Most MS cases are relapsing remitting (90%), but other cases are purely progressive.

8.2 History

MS can cause a wide variety of motor (35%), sensory (37%), visual (50%), and cerebellar (17%) symptoms. A pseudo-exacerbation is often caused by stress, heat, or infections and will resolve within 48 hours with rest or treatment of the bacterial infection. UTI is a very common cause of a pseudo-exacerbation. However, MS causes progressive bladder spasticity resulting in urgency and frequency resembling UTI which often makes the diagnosis difficult. Incontinence to both bowel and bladder can occur, but this is generally a late finding. Cognitive and emotional problems are common in established cases. Heat sensitivity, the appearance or worsening of symptoms on exposure to heat (often a hot shower), is present in most patients.

Vertigo may occur and is often associated with symptoms resembling lesions of the fifth or seventh cranial nerve. Optic neuritis (see Chap. 12 on "Ophthalmic Emergencies") is common and is associated with eye pain and vision loss. Double vision or blurry vision may be due to a sixth nerve palsy or internuclear ophthalmoplegia (INO) (see below). Bilateral INO in an awake patient is highly suggestive of MS. Trigeminal neuralgia is lancinating shock pain in the distribution of the trigeminal nerve and, when bilateral in a patient under 50, is highly suggestive of MS.

8.3 Exam

Lhermitte sign is classic though not entirely specific for MS and is described as electric shocks, vibration, or pain radiating down the back and often into the legs caused by flexion of the neck. It is rare at approximately 3%. Strength may be decreased in a focal or more generalized pattern. Pain and temperature sensation may be decreased. Optic neuritis with loss of central vision (see Chapter on "Ophthalmic Emergencies") is the presenting finding in 30% of MS patients. INO is characterized by failure of eye adduction and accompanying horizontal nystagmus of the abducting eye and, when bilateral, is strongly suggestive of MS. As MS is a disease of the central nervous system, hyperreflexia is present. Vertigo is accompanied by ataxia and nystagmus without latency, direction reversal, or fatigue. Hearing loss, facial hemispasm, or facial nerve palsy may occur but are rare.

8.4 Diagnostics

MRI of the brain and spine is used to identify an exacerbation.

Key Points

- Management of MS exacerbations is extremely challenging and should ideally be managed in close conjunction with the patient's neurologist.
- Monosymptomatic presentations can be managed with prompt MRI testing.
- More moderate or severe presentations are managed with IV steroids after hospitalization.
- Pseudo-exacerbations are often caused by UTI.

9 Myasthenia Gravis

Myasthenia gravis is a failure of the electrical impulse to conduct at the neuromuscular junction caused by autoantibodies to the acetylcholine receptor on the postsynaptic membrane.

9.1 General Considerations

Approximately, 90,000 cases of MG occur annually. The disease may appear alone or in conjunction with other autoimmune disorders such as Grave's disease or lupus. The peak incidence is 20–30 years old in women and 50–60 years old in men. The outpatient clinician may be diagnosing the condition or managing an exacerbation. In both situations, a myasthenic crisis may result in a sudden worsening of muscular function including the respiratory muscles resulting in the need for a monitored setting. A common precipitating factor is an infection. High dose steroids should not be used initially as this can cause a paradoxical weakening in one third of patients.

9.2 History

Exacerbations can be caused by infections, treatment with certain drugs, or occur spontaneously. Sensory symptoms and pain are absent. The cardinal pattern of MG is fluctuating motor weakness. Repeated use of a muscle results in weakening which is at least partially relieved by rest of the affected muscle(s). Double vision that improves by resting the eyes with lids closed may occur. Difficulty swallowing and speaking implies involvement of the bulbar muscles and may portend respiratory collapse.

9.3 Exam

There are many exam findings that strongly suggest MG. The presence of weakness of the face muscles occurs in 80% of patients and along with levator palpebrae weakness (ptosis) with sparing of the pupils is virtually diagnostic. This differentiates MG from botulism where the pupils are affected. The effect on the facial muscles causes the characteristic "snarl" when the patient is asked to smile. Counting backward from 100 may cause the speech to become slurred and indistinct. A finding that is improved with rest. Sustained gaze in one direction may worsen double vision and can be corrected by resting the eyes. The skeletal muscles may be affected in virtually any pattern with proximal muscles usually more affected than distal muscles. Reflexes are usually intact unless the weakness is severe.

Key Points

- MG should be considered in the differential in any case of muscle weakness.
- The pupils are unaffected by MG.
- Exacerbations can be caused by infections and common drugs including statins, steroids, and antibiotics.

- MG can cause sudden severe weakness of the muscles and these patients should usually be hospitalized. This is especially true when muscles of the pharynx are compromised as this is commonly associated with respiratory compromise.

10 Seizures

Seizure is abnormal discharging of neurons that can occur in various locations in the brain resulting in partial or generalized sensory, motor, autonomic, or psychic symptoms.

10.1 General Considerations

The primary care physician does not need to know the details on how to manage an acute seizure. But occasionally, a patient will present to the provider after the event, with or without a clear diagnosis. For safety, the patient should be told that they cannot drive. Some states require that the clinician report the patient to the Department of Motor Vehicles (DMV). Driving is less of an issue with epileptic patients, and they should be referred back to their neurologists after ordering blood levels of their anticonvulsants, if necessary. A seizure that is idiopathic is referred to as a primary seizure. Otherwise, seizures are referred to as secondary and have a variety of causes such as active CVA, CNS infections (such as HIV), hypoglycemia (very common), and electrolyte abnormalities. Overdoses (alcohol, cocaine, and tricyclic antidepressants) and eclampsia are also precipitants of secondary seizures.

10.2 History

A seizure often has various auras, especially a sense of epigastric rising. Various automatisms such as lip smacking, swallowing, chewing, or fumbling may be described. Behavioral symptoms may be seen, and non-convulsive status epilepticus (NCS) may be responsible for non-resolving psychiatric symptoms following primary or secondary seizures. The patient may report unilateral motor symptoms following the seizure, termed Todd's paralysis, often the sign of a structural brain lesion. Incontinence and biting along the lateral surfaces of the tongue (as opposed to the tip of the tongue seen in syncope) are classically reported. Hypoglycemia is a common cause for seizures, and other symptoms of hypoglycemia should be ascertained (such as faintness, tremor, nausea, or diaphoresis). Unfortunately, these symptoms can be confused with vasovagal syncope. Additionally, syncope can cause convulsions, a condition termed convulsive syncope (see Chap. 1 on "Syncope in Cardiovascular Disease"). The convulsions in seizure usually are more dramatic

and longer lasting approximately 2 min or more. Syncope and seizure are difficult to diagnose on clinical grounds and often specialists in both the fields of neurology and cardiology will need to be consulted.

10.3 Exam

The primary care physician will encounter a normal physical exam by the time the patient presents to the office. However, NCS should be on the differential of behavior changes after the seizure event.

10.4 Diagnostics

A finger stick can be performed in the office that may signal hypoglycemia as the cause for the seizure.

Key Points

- If hypoglycemia can be identified as the cause for a secondary seizure, then a workup can be obtained as an outpatient. Otherwise, a referral to the ED for expedited workup of a first seizure (CT and labs) should be considered if outpatient studies cannot be obtained quickly.
- Non-convulsive status epilepticus may be responsible for behavioral changes after a seizure.

11 Miscellaneous

11.1 Guillain–Barre Syndrome

Guillain–Barre syndrome (GBS) is an acute polyneuropathy caused by inflammation of the myelin sheath of peripheral nerves by an autoimmune mechanism.

11.2 General Considerations

GBS is rare with approximately 3500 cases annually in the U.S. This autoimmune reaction is associated with various infections classically *Campylobacter jejuni* or vaccinations which is an extremely rare event. Other associations are the herpes class of viruses (EBV, CMV) and surgical procedures. Protein in the CSF becomes elevated by the first week of symptoms. A normal lumbar puncture does not rule out

the diagnosis because protein can be absent at the beginning of symptoms. The treatment is intravenous immunoglobulin (IVIG) or plasmapheresis.

11.3 History

A preceding illness can be identified approximately one-third of the time, often gastrointestinal or viral. Often the first report is difficulty getting out of a car or standing from a seated position. Difficulty climbing stairs is sometimes the first report. Aching pain may be present in the thighs and back. This is followed by a gradual onset of weakness (days to weeks) usually in an ascending pattern from legs upward. Sometimes the weakness can have an explosive onset and happen within a day or two. The cranial nerves can be affected. In the Fisher variant, the weakness begins in the face and upper body first, sometimes with ataxia, and can be confused with myasthenia gravis or CVA. Sensory symptoms are lacking or mild and are not noticed because pain fibers are not generally involved. Autonomic dysfunction may be present, resulting in difficulty urinating or constipation. Facial flushing may be reported. Presyncope may be reported due to orthostatic blood pressures. The diaphragm may be affected causing a gradual onset of shortness of breath.

11.4 Exam

Blood pressure may be high or low depending on the position of the patient. Autonomic dysfunction is differentiated from orthostasis by being sustained more than 30 seconds. Due to autonomic dysfunction, the heart rate may be fluctuating with tachycardia or bradycardia. An ophthalmoplegia or other cranial neuropathies, especially the facial nerve, may be present (50% of the time) and usually will be bilateral. The weakness is usually symmetrical and will typically be ascending found in the lower extremities first. Hyporeflexia or areflexia will be elicited due to the peripheral nature of this nervous system disorder.

Key Points

- GBS is a rare peripheral neuropathy causing paralysis, usually in an ascending pattern, and autonomic dysfunction.
- Cranial nerves especially the facial nerve can be affected.
- GBS can cause respiratory compromise and the patient needs to be in a monitored setting.

12 Transverse Myelitis

Transverse myelitis is an inflammation of the spinal cord causing neurologic deficits downstream of the lesion.

12.1 General Considerations

The condition is rare with approximately 1500 cases reported annually in the U.S. Up to 40% of cases are associated with an antecedent infection. Many infections have been associated such as influenza, herpes viruses (EBV and CMV), the childhood viruses (measles, mumps, and rubella), and varicella. Multiple sclerosis may also be implicated in some cases.

12.2 History

The initial symptom is focal neck or back pain followed by various combinations of sensory symptoms, weakness, and sphincter dysfunction. Urinary retention or overflow incontinence can be reported. Isolated hemicord involvement can cause unilateral symptoms. The symptoms can evolve quickly within hours or develop within days. The symptoms can start distally and ascend like Guillain–Barre syndrome (GBS); however, involvement of the trunk is the distinction between the two entities.

12.3 Exam

A sharply demarcated cord level is seen on sensory exam. Weakness in the arms and/or legs is typical. Hyperreflexia is the rule but areflexia can be seen with spinal shock.

Key Points

- Transverse myelitis is an emergency characterized by a sensory cord level on the trunk.
- The treatment is IV steroids after hospitalization.

13 Wernicke's Encephalopathy

Wernicke's encephalopathy is a constellation of neurologic symptoms caused by deficiency of thiamine (vitamin B1).

13.1 General Considerations

This is mainly a disease of alcoholics though only a fraction of alcoholics develop these symptoms. Wernicke disease is also associated with malnutrition, renal dialysis, cancer or acquired immune deficiency syndrome (AIDS).

13.2 History

A history of disorders of malnutrition should be obtained. A history of alcoholism may be related as well. Double vision and difficulty ambulating may be reported. In many instances, the patient will be confused or indifferent and inattentive.

13.3 Exam

Ophthalmoplegia is characterized by lateral rectus palsy (usually bilateral) and horizontal nystagmus on lateral gaze. Ptosis and miosis may rarely occur. A wide based gait with inability to tandem walk is observed.

Key Points

- Wernicke's disease is a medical emergency as lack of IV thiamine can result in stupor and death.
- A patient who is suspected to have Wernicke's encephalopathy should be referred to the ED.

14 Botulism

Botulism is an extremely rare, life-threatening neuromuscular disease caused by the bacterium *Clostridium botulinum*.

14.1 General Considerations

Botulism is covered in the neurology section due to its dramatic neurologic sequelae. Botulism is toxin mediated and adults can acquire the condition via infected food or an infected wound. The disease is extremely rare with only reports of roughly 23 cases of food borne botulism per year. Food borne botulism is associated with a low pH, low salt, low sugar environment often seen with home canning. Wound botulism is often associated with injection drug users, especially black tar heroin tissue injection. The disease progresses throughout the entire body eventually affecting all the skeletal muscles including the diaphragm. The result is asphyxiation and death.

14.2 History

A history of home canning or injection drug use may be reported. In foodborne botulism, abdominal pain, nausea, and vomiting may be reported 12–72 h after food ingestion. Double vision is reported. Dysphagia, dysphonia, and slurred speech are common termed bulbar symptoms. Altered mental status is not reported as the toxin does not cross the blood–brain barrier.

14.3 Exam

Multiple cranial nerves are affected therefore isolated palsies are often not seen. Cranial nerve palsies are symmetrical causing bilateral ptosis and bilateral pupillary dilation. The patient may be slurring words. Weakness of muscles can be reported in any combination throughout the body.

Key Points

- Botulism is an extremely rare toxin mediated condition characterized by multiple symmetrical cranial nerve palsies with dilation of the pupils. The toxin can be generated by clostridium found in canned goods or infected wounds.
- The diagnosis is clinical, and the Centers for Disease Control and Prevention (CDC) has a 24-h hotline for consultations in suspected cases.

Further Reading

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