Weakness  (Dr. Merchut)

1. Lower motor neuron patterns of weakness

   Patients may have lower motor neuron (LMN) signs (more focal weakness and prominent muscle atrophy, decreased muscle stretch reflexes and tone, and fasciculations) from lesions occurring anywhere along the length of spinal cord or brain stem lower motor neurons. Involvement of a single peripheral nerve leads to LMN signs as well as sensory impairment confined to its anatomical territory, which may be accompanied by painful paresthesia or dysesthesia. In most cases of peripheral neuropathy or polyneuropathy, multiple nerve involvement manifests as distal limb LMN signs and sensory ("stocking and glove") loss, typically beginning in the lower limbs. A lesion of the brachial or lumbosacral plexus causes LMN signs and sensory deficit according to the anatomical territory of the trunk, division or cord of the plexus involved. A radiculopathy usually involves neck or back pain which may radiate into a limb or the trunk in a dermatomal distribution, along which tingling or numbness may also occur. LMN signs occur in muscles innervated by the involved spinal nerve root. Anterior horn cell lesions may cause weakness in a distal or proximal segment of a limb, eventually becoming more widespread and bilateral with prominent fasciculations in amyotrophic lateral sclerosis (ALS). ALS may also affect the brain stem lower motor neurons involved with speaking, chewing or swallowing. In general, pain may often accompany lesions of roots, plexus or nerves, but not lesions of anterior horn cells.

2. Upper motor neuron patterns of weakness

   Often patients have upper motor neuron (UMN) signs (more diffuse weakness with relatively less muscle atrophy, hyper-reflexia, spasticity, and Babinski signs) in the limbs on one side of the body described as hemiparesis or hemiplegia. The responsible lesion or lesions may occur anywhere along the extent of the corticospinal tract from the motor cortex down to the spinal cord. More exact localization of the lesion is aided by other signs or clinical clues. Hemiplegia from an ipsilateral cervical spinal cord lesion may be accompanied by neck and radicular pain if a cervical root is also involved at the level of the lesion, which may also create LMN signs at that root level. There may be ipsilateral sensory deficits for position sense and vibration (posterior or dorsal columns) and contralateral sensory deficits for pain (pinprick) and temperature (spinothalamic tract) up to a dermatomal level. Hemiplegia from a brain stem lesion may be accompanied by facial weakness, dysarthria, or dysphagia. A "crossed" brain stem syndrome may occur, with a cranial nerve lesion on one side contralateral to the hemiplegia, or there may be involvement of specific brain stem tracts such as the medial longitudinal fasciculus (MLF). Hemiplegia from a subcortical lesion in the internal capsule or corona radiata causes a relatively equal weakness in the contralateral lower face and upper and lower limbs, since the descending motor fibers are compacted closely in this area. Hemiplegia from a cortical lesion may have unequal weakness between the affected upper and lower limbs. Hemiplegia with the leg weaker than the face and arm is caused by a lesion in the more medial (parasagittal) portion of the contralateral motor
cortex. Ischemic infarction of this area is produced by an occlusion of the anterior cerebral artery (Fig. 1) or one of its branches. If the nearby sensory cortex is also involved, sensory deficits may also predominate in the lower limb. Hemiplegia with the face and arm weaker than the leg is caused by a lesion in the more lateral portion of the contralateral motor cortex. Ischemic infarction of this area is produced by an occlusion of the middle cerebral artery (Fig. 1) or one of its branches. Involvement of other adjacent cortical areas may produce accompanying symptoms of aphasia (in the dominant hemisphere), contralateral visual field deficits or sensory deficits predominating in the face and upper limb. It should be noted that motor cortex is not perfused by the posterior cerebral artery.

The time it takes for upper motor neuron weakness to develop is another clue to its etiology. The sudden onset of hemiplegia is more typical of a cerebrovascular lesion, such as an ischemic infarct or hemorrhage, or the result of trauma. The more gradual onset of upper motor neuron weakness suggests a tumor or a degenerative disease such as amyotrophic lateral sclerosis.

Fig. 1
Vascular territory of the cerebral arteries. (From Greenberg DA, Aminoff MJ, Simon RP: Clinical neurology, ed 2, Norwalk, Conn., 1993, Appleton & Lange.)
3. Bulbar versus pseudobulbar weakness

"Bulbar" refers to the "bulb" or lower brain stem, where lesions of the fifth, seventh, ninth, tenth or twelfth cranial nerves can cause weakness of chewing, speaking or swallowing, referred to as bulbar function(s). In the presence of bulbar weakness (bulbar palsy), LMN signs are manifest as atrophy and fasciculations of muscles of the face, jaws, palate or tongue. These LMN signs in non-limb muscles are more difficult to appreciate on physical examination, but may be best observed in the tongue. Pseudobulbar weakness (pseudobulbar palsy) also causes weakness of chewing, speaking or swallowing, but the causative lesions involve the upper motor neurons (corticobulbar tract) which control the respective cranial nerve nuclei in the brainstem. UMN signs include a hyperactive jaw jerk (masseter reflex) in the absence of atrophy or fasciculations of the weakened muscles.

4. Weakness without lower or upper motor neuron signs

Weak patients lacking LMN or UMN signs may have a disorder of muscle or the neuromuscular junction. In either case, sensation is preserved. In muscle disease, the weakness typically affects the proximal limbs. The muscle stretch reflexes are preserved initially, disappearing only after significant muscle atrophy occurs. In neuromuscular junction disorders like myasthenia gravis, there is variable weakness and fatigue of the limbs, often accompanied by ptosis, diplopia, dysarthria, dysphagia, or dyspnea.

5. Other systems affecting motor control

Patients complaining of "weakness" may actually have slowness or clumsiness instead, which may be due to problems with the extrapyramidal system (primarily the basal ganglia) or the cerebellar system. The basal ganglia influence postural control, muscle tone and more "automatic" types of movement. The cerebellum and its connections are important for balance, smoothness and coordination of movement.