Movement Disorders

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I. The Basal Ganglia

The basal ganglia are composed of several structures including the caudate and putamen (collectively called the striatum), globus pallidus externa, globus pallidus interna, subthalamic nuclei and the substantia nigra.

The main input area for the basal ganglia is the striatum. Projections from the motor cortex and substantia nigra terminate in the striatum.

The striatum then projects to the globus pallidus interna (GPI) through either the direct or indirect pathway.

The direct pathway projects direct to the GPI.

The indirect pathway projects indirectly to the GPI through the globus pallidus externa and subthalamic nucleus.

The GPI summates the input from the direct and indirect pathway and projects to the thalamus. The GPI is the key structure in regulating the output through the basal ganglia.

The thalamus projects to the cortex to complete the circuit.

II. Movement Disorders

Movement disorders are a group of diseases which manifest themselves as abnormalities of movement. The first step in diagnosis is identifying the abnormal movement (IE, a tremor, dystonia, chorea, etc.). Identifying the abnormal movement will generate a differential diagnosis. One can then narrow the differential by searches for clues in the history and physical exam as well as ordering diagnostic test to help confirm your hypothesis.

III. Normal Movement

In order to identify an abnormal movement, one first must become familiar with normal movement. Although we have all been watching each other move our entire lives, usually we have not made careful observations. In the next few days, take a few moments to watch how your friends and family move (hopefully these people represent the Anormal population@). Notice how their faces do not move except when they are talking or showing emotion. At rest, people do not usually move their

arms or legs. Therefore, movements that occur at rest are not common and may represent an abnormal movement such as the rest tremor of Parkinson=s Disease or chorea in Huntington=s Disease. The rest of the lecture will center on describing several different abnormal movements. Once familiar with the characteristics of these movements, they are easily identified.

IV. Tremor

Tremor is a rhythmic oscillating movement across a joint usually representing the contraction of agonist and antagonistic muscles. The key to diagnosing tremor is the regular rhythm of the movement.

It is helpful to classify tremor according to the position in which it occurs. Therefore a tremor may be classified as rest, postural, or action.

Rest Tremor

Tremor primarily occurs when the body part is at rest. This may be the arms, legs or the jaw. The classic rest tremor made be relieved by action of the body part. This type of tremor is typically seen with Parkinson=s Disease.

Postural Tremor

Postural tremor is maximal when the patient holds the extremity (usually the arm and hand) suspended against gravity. This type of tremor is characteristic of Essential Tremor. The tremor may also be present with action but is usually symmetrical throughout the movement as opposed to intention or cerebellar tremor to be discussed later.

Action Tremor

Action tremor is maximal during the movement of a limb. The tremor may be symmetrically present throughout the movement or it may increase as one approaches the target. The latter is typical presentation of an Intention or Cerebellar Tremor. Patients with Intention Tremor may have other signs of cerebellar dysfunction such as dysmetria or dysdidokinesis.

V. Chorea

Chorea is a non-rhythmical movement of moderate amplitude and speed that flits from one body part to another in a unpredictable random sequence.

Chorea may occur in a variety of conditions including Huntington=s Disease and Tardive Dyskinesia.

Huntington=s Disease.(HD)

Eloquently describe by George Huntington, the disease consists of chorea, neuropsychiatric disturbances, progressive dementia and a positive family history. **HD is associated with an expanded trinucleotide repeat sequence (CAG) on the short arm on Chromosome 4.**

The chorea may affect the head, face, neck and extremities. The chorea may superimpose on normal movement giving them a lurching quality. The gait has a characteristic dance like quality.

VI. Dystonia

Dytonia is defined as a sustained abnormal muscle contraction producing pain and/or a positional deformity. There are many etiologies for dystonia and although in some the cause has been identified, the majority of cases are idiopathic.

A. Craniofacial Dystonia

This category of dystonia contains many different syndromes including laryngeospasm (tonic contraction of the vocal cords), blepharospasm (forced eye lid closure), Meige Syndrome (blepharospasm associated with lower facial dyskinesias), and hemifacial spasm (tonic semirythmic contraction of one side of the face),

B. Occupational Dystoinas

This category of dystonia refers to syndromes in which the dystonia develops in associated with some task. The most common is Writer=s Cramp in which the fingers, hand, and/or forearm develops dystonia associated with writing. This type of dystonia may also be seen in association with cooking or playing a musical instrument. One theory is that the repetitive movements associated with the above activities some how induce mild trauma and serve as a trigger for the development of the dystonia.

VII. Cervical Dystonia (Torticollis)

This category of dytonia refers to dytonia involving the cervical neck muscles. Although usually called Torticollis this name is probably inappropriate. Torticollis refers to the twisting or rotation of a patient=s neck cause by dytonic neck muscle contraction. Patients with cervical dystonia may also have laterocollis (lateral flexion), anterocollis (forwards flexion), retrocollis (posterior flexion), etc

A particular unique feature in dystonia especially in cervical dystonia is the geste antagonist or sensory trick. By using the geste (the patients usually touches their face or head in a certain way), patients are able to temporarily alter the dystonic neck muscle contraction and enjoy reduced positional deformity.

Dopa Responsive Dystonia (Segawa=s Disease)

This disorder is an autosomal dominant dystonia that usually presents in childhood. Although rare it is an important syndrome to recognize since it is easily treated with excellent results. The key to the diagnosis is the diurnal fluctuation of the abnormal movement. The affected child is usually better in the morning hours although there may be some baseline dystonia. Symptoms become progressively worsen during the day, until evening when the symptoms are maximal.

It is now known that this disorder is caused by a deficiency in tetrhydrobioterin production. Levodopa in small doses produces a dramatic and long lasting improvement in these patients.

Secondary Dystonia

Dystonia may be secondary to multiple etiologies including vascular, tumor), demylinating disease, and truama. All of these etiologies usually result in structural lesions in the basal ganglion especially in the putamen. The dystonia may be generalized and severe.

VII. Myoclonus

Myoclonus is a lightening like muscle jerk that may involve any body part. It has multiple etiologies most of which are idiopathic. Myoclonus may be disabling by superimposing on the patient=s normal movements thus making them incordinated and inaffective.

VIII. Tics

Tics described abnormal repetitive sterotypic movements (motor) or sounds (vocal) that usually begin in childhood and adolescence. Examples of tics include repetitive eye blink, facial movement, head shakes, or vocalizations such as sniffing, coughing or clearing the throat.

Tourette=s Syndrome (TS)
This syndrome begins before the age of 18 and the patient exhibits multiple motor and at least one vocal tics. The tics are presistent, cause significant impairment including social dysfunction, and are not secodary to some other condition such as stimulants or as disease such as Huntington=s Disease.

Tics are accompanied by an involuntary urge to perform the tic. Patients can surpress the movements but doing so cause anxiety which is relieved by performing the tic.

TS may be associated with Attention Deficit Hyerpactivity Disorder (ADHD) and Oppessive Compulsive Disorder (OCD). Although these disorders occur in conjunction with TS, they require separate evaluation and treatment.

IX. Hemiballismus

Hemiballism is a choreic type movement of median to large amplitude that is confined to one area of the body. Hemiballism often results from a lesion in the contralateral subthalamic nucleus.

The movement can be very disabling. Some patients will have resolution of their symptoms over time whereas others may not.

X. The Cerebellum

The cerebellum can be basically divided into two parts: the midline structures (vermis and flocculonodular lobe) and the cerebellar hemispheres (including the dentate nucleus).

Cerebellar lesions produce signs and symptoms IPSILATERAL to the lesion. This is because the cerebellum is a shifty double crossing animal. Projections from the cerebellum cross the contralateral side in the midbrain and then proceed to the motor cortex via the thalamus. Projections from the motor cortex then project to the spinal cord crossing at the pyramidal decussation. Therefore a lesion in the Right cerebellar hemisphere would influence the Left motor cortex which in turns projects to the Right side of the body. Clear as mud, isn't.

Midline cerebellar lesions produce ataxia or an unsteady broad based gait.

Hemispheric cerebellar lesions produce dysmetria, dysdiadochokinesia, and an intention tremor.