CASE 8: HEAD TILT AND UNSTEADY GAIT (Slide CC8-1)

1. Coordinated movement of the limbs depends on the synergistic activity of several different sensory and motor systems. One can easily be fooled into thinking that a patient has "ataxia" due to a cerebellar lesion while, in fact, the patient may instead have a disturbance in the pyramidal system, proprioception, basal ganglia or even a peripheral nerve injury causing awkward or unsteady movements. Therefore, it is essential to test motor strength, tone, and joint position sense before doing tests intended to elicit cerebellar abnormalities.

Our patient had normal strength, tone and reflexes (joint position sense was not tested), however, she had ataxia and dysmetria of reaching movements with the right hand. This suggests a lesion involving either the right cerebellar hemisphere, the right spinocerebellar pathways, the left corticopontine pathways, or the left pontine nuclei. Of these possibilities, the right cerebellar hemisphere is the most likely location, since there were no associated symptoms or signs expected for a lesion in the spinal cord, cerebellar peduncles, pons, or cerebrum.

Head tilt can be seen in cerebellar lesions for three reasons: 1. Disturbance of pathways influencing the tectospinal, reticulospinal, vestibulospinal and anterior corticospinal tracts will affect proximal muscles controlling posture. 2. Lesions of the cerebellum extending into the superior medullary velum often compress the trochlear nerve. This will cause the ipsilateral eye to move slightly upward and the head will then tilt to compensate. 3. Cerebellar lesions can produce unilateral meningeal irritation in the posterior fossa and consequent head tilt.

As discussed in the clinical case for Lab 3, unsteady gait can be caused by damage to *many* different structures including: the cerebral cortex, basal ganglia, cerebellum, descending pathways to the spinal cord or brain stem, the spinal cord, brain stem, peripheral nerves, vestibular apparatus, and skeletomuscular system. Cerebellar lesions involving the midline vermis and flocculonodular lobes disrupt pathways involved in gait, posture and muscle tone, producing an unsteady, staggering gait.

2. The above findings are consistent with a lesion extending from the midline vermis into the right cerebellar hemisphere. The most common lesions in this region in children are tumors such as medulloblastoma, astrocytoma and ependymoma. Other less common possibilities include glioma, epidermoid cyst, haemangioblastoma, arterio-venous malformation, and choroid plexus papilloma.

CT Scan:

A CT scan was done in our patient because the pediatrician suspected an intracranial neoplasm (see slide). Sequential images are shown without IV contrast from more caudal (upper left image) to more rostral (lower right image) levels of the cerebellum. A round hypodense lesion can be seen involving the vermis and right cerebellar hemisphere, with impingement on the pons and midbrain. In the upper right image significant surrounding edema (darker areas) can be seen. Interestingly, the fourth ventricle appears to be obliterated by the lesion yet there is only slight dilatation of the lateral and third ventricles (i.e. very mild hydrocephalus). Recall that our patient did not have signs of elevated intracranial pressure such as nausea, vomiting and papilledema. This is yet another example of the variable ways that disease processes can present in actual clinical practice. Additional scans were done with IV contrast (not shown) which demonstrated enhancement of the round mass in the cerebellum.

Clinical Course:

The patient was admitted to the hospital for resection of the tumor. A frozen section of the tumor performed in the operating room revealed a low grade astrocytoma. This type of tumor can have a five year survival of up to 90% and a cure rate of 70 to 80% with complete surgical resection. The tumor was, therefore, completely resected in the operating room with no subsequent radiotherapy or chemotherapy. Permanent fixed pathologic sections confirmed the diagnosis. Follow-up CT scans at two weeks, six months and two years revealed no evidence of recurrence. She remains with a slightly unsteady gait, but is otherwise without deficits.