**Question 76**

A 75-year-old woman with longstanding rheumatoid arthritis complains of neck pain. X-rays of her cervical spine demonstrate the changes shown below.

Which one of the following is most likely to accompany these changes?

A. sensory loss in the C2 dermatome
B. Horner’s syndrome
C. wasting of the small muscles of the hands
D. Lower motor neurone changes in the upper limbs.
E. Upper motor neurone changes in the upper limbs.

**Cervical subluxation in rheumatoid arthritis**

CERVICAL INVOLVEMENT — Cervical joint destruction in patients with RA may lead to vertebral malalignment (eg, subluxation). Risk factors for development of cervical subluxation include older age at onset of RA, more active synovitis, and rapidly progressive erosive peripheral joint disease. Both atlantoaxial and subaxial (below C1-C2) joints may be involved.

**Atlantoaxial disease** —

- Among the joints of the cervical spine, the atlantoaxial joint is prone to subluxation in multiple directions. The atlas (C1) can move anteriorly, posteriorly, vertically, laterally, or rotationally relative to the axis (odontoid and body of C2):
  - Abnormal anterior movement on the axis is the most common type of subluxation. It often results from laxity of the transverse ligament induced by proliferative synovial tissue in an adjacent synovial pouch, but may also occur as a result of erosion or fracture of the odontoid process.
  - Posterior movement on the axis can occur only if the odontoid peg has been fractured from the axis or destroyed.
  - Atraumatic posterior subluxation is rarely associated with cervical myelopathy.
  - Vertical movement in relation to the axis is least common; it results from destruction of the lateral atlantoaxial joints or of bone around the foramen magnum.
  - Vertical atlantoaxial subluxation may occur in those with initial anterior-posterior subluxation. Vertical subluxations are believed to have a worse prognosis than the other varieties.
Year 2003 Paper two: Questions supplied by Tricia

The involvement and severity of cervical spine disease in RA parallels the progression of peripheral joint erosions. As a result, cervical subluxation is more likely in those with erosions of the hand, feet, hips and/or knees.

Symptoms — Involvement of cervical joints may result in
- significant pain
- range of motion may be normal in the absence of muscle spasm.
- The earliest and most common symptom of cervical subluxation is pain radiating superiorly towards the occiput.
- Additional symptoms of subluxation include:
  - Spastic quadriparesis which is slowly progressive
  - Sensory findings are also common, including painless sensory loss in the hands or feet. In patients with C1-C2 subluxation, transient episodes of medullary dysfunction (such as respiratory irregularity) associated with vertical penetration of the odontoid process of C2 and probable vertebral artery compression. Movement of the hands in this setting may result in paresthesias in the shoulder or arms.
- Neurologic findings in patients with vertical atlantoaxial subluxation may also include decreased sensation in the distribution of the fifth cranial nerve, sensory loss in the C2 area, nystagmus, and pyramidal lesions. (one of a group not isolated finding)
- The symptoms of spinal cord compression that demand immediate attention and intervention include:
  - A sensation of the head falling forward upon flexion of the cervical spine
  - Changes in levels of consciousness
  - "Drop" attacks
  - Loss of sphincter control
  - Respiratory dysfunction
  - Dysphagia,
  - vertigo,
  - convulsions,
  - hemiplegia,
  - dysarthria, or nystagmus
  - Peripheral paresthesias without evidence of peripheral nerve disease or compression

Physical findings — Physical findings relating to the spine which are suggestive of atlantoaxial subluxation include: Loss of occipito-cervical lordosis Resistance to passive spine motion Abnormal protrusion of the axial arch felt by the examining finger on the posterior pharyngeal wall

In addition, neurologic findings appropriate to the symptoms described above may be seen, including:
- Increased deep tendon reflexes
- Extensor plantar responses
- Muscle weakness, spasticity, or muscle atrophy Gait disorders

All upper motor neuron changes

RADIOGRAPHIC FINDINGS — Among patients with atlantoaxial subluxation, plain radiographic views of the cervical spine (lateral, with the neck in flexion) may reveal more than 3 mm of separation between the odontoid peg and the C1 arch. Separation between C1 and C2 (anterior subluxation) of 9 mm or more, or a posterior atlanto-dental distance of less than 14 mm is associated with an increased incidence of cord compression. In addition, if the space available for the spinal cord is less than 13 mm anywhere in the cervical region, there is an increased risk for neurologic impairment. In symptomatic patients, the films in flexion should be taken only after radiographs (including an open-mouth view) have excluded an odontoid fracture or severe atlantoaxial subluxation.
Upper versus lower motor neuron lesions — Several examination findings help to distinguish central from peripheral lesions in the motor system.

Deep tendon reflexes:
- reflexes are typically hyperactive with a central lesion and hypoactive with a peripheral one.
- The Babinski sign is a reliable indicator of a central lesion.
- Atrophy and fasciculations are common with lower motor neuron disease and unusual with upper motor neuron disease.

The pattern of muscle involvement.
- A central lesion usually results in weakness that is more pronounced in the flexors of the lower extremities than in the extensors, but in the upper extremities the extensors are weaker than the flexors. This is often called pyramidal weakness, but it does not occur with pure lesions of the pyramidal tracts. Instead, it is the net result of disrupting all the descending motor tracts and is probably most appropriately called an upper motor neuron (UMN) pattern of weakness.
- The UMN pattern of weakness also causes supination of the upper extremity to be weaker than pronation; this accounts for the finding of a pronator drift, in which the arm pronates and drifts downward when the patient is asked to hold it extended with palms up (supinated). This is a fairly sensitive indicator of subtle UMN weakness. It is also useful as a test for internal consistency because patients with nonorganic weakness will often allow their arm to drift downward but fail to pronate it.

Muscle tone
- A central lesion is characterized by spasticity, whereas tone is normal or reduced with a peripheral lesion.

Horner’s syndrome

NEUROANATOMY — A Horner's syndrome can result from a lesion anywhere along a three-neuron sympathetic (adrenergic) pathway that originates in the hypothalamus:
- The first-order neuron descends caudally from the hypothalamus to the first synapse, which is located in the cervical spinal cord (levels C8-T2, also called ciliospinal center of Budge).
- The second-order neuron travels from the sympathetic trunk, through the brachial plexus, over the lung apex. It then ascends to the superior cervical ganglion, located near the angle of the mandible and the bifurcation of the common carotid artery.
- The third-order neuron then ascends within the adventitia of the internal carotid artery, through the cavernous sinus, where it is in close relation to the sixth cranial nerve.
- The oculosympathetic pathway then joins the ophthalmic (V1) division of the fifth cranial nerve (trigeminal nerve). In the orbit and the eye, the oculosympathetic fibers innervate the iris dilator muscle as well as Müller's muscle, a small smooth muscle in the eyelids responsible for a minor portion of the upper lid elevation and lower lid retraction.

CLINICAL FEATURES — The classic signs of a Horner's syndrome are ptosis, miosis, and anhidrosis. The degree of anisocoria is more marked in the dark than in the light.

Horner’s syndromes are frequently caused by trauma and/or a surgical procedure involving the chest or neck.

ETIOLOGY — The etiology of Horner’s syndrome in adults relates to the lesion location
First-order syndrome — Lesions of the sympathetic tracts in the brainstem or cervicothoracic spinal cord can produce a first-order Horner's syndrome.
- The most common cause is a lateral medullary infarction, which produces a Horner's syndrome as part of the Wallenberg syndrome. Typically the patient presents with vertigo and ataxia, which overshadow the Horner's syndrome. Other neurologic symptoms and signs include abnormal eye movements, ipsilateral limb ataxia, and a dissociated sensory loss (loss of pain and temperature sensation on the ipsilateral face and contralateral trunk). Hoarseness and dysphagia are also often present.
Strokes, tumors, and demyelinating lesions affecting the sympathetic tracts in the hypothalamus, midbrain, pons, medulla, or cervicothoracic spinal cord are other potential causes of a central Horner's syndrome. Syringomyelia and cervical cord trauma can also produce a Horner's syndrome when the intermediolateral columns are affected.

Second-order syndrome — Second-order or preganglionic Horner's syndromes can occur with trauma or surgery involving the spinal cord, thoracic outlet, or lung apex.

- Lumbar epidural anesthesia can also produce a Horner's syndrome due to pharmacologic disruption of the preganglionic neuron as it exits the spinal cord. This is most often described in association with obstetrical procedures.

Third-order syndrome — Third-order Horner's syndromes often indicate lesions of the internal carotid artery such as an arterial dissection, thrombosis, or cavernous sinus aneurysm. Carotid endarterectomy and carotid artery stenting can also produce a Horner's syndrome

- An acute Horner's syndrome with neck or facial pain should be presumed to be caused by carotid dissection until proven otherwise
- Other causes of postganglionic Horner's syndrome include neck masses, otitis media, and pathology involving the cavernous sinus. When the cavernous sinus is involved, other oculomotor deficits, particularly a sixth nerve palsy, commonly occur
- A Horner's syndrome is a common feature of cluster headache, occurring with unilateral eye or temple pain and lacrimation, generally lasting no more than an hour or two

**Wasting of the small muscles of the hands**

In the hand, the median nerve supplies the lateral two lumbricals, opponens pollicis, abductor pollicis brevis, and flexor pollicis brevis; the remainder are served by the ulnar nerve.

Wasting of the interossei (prominent guttering of the back of the hand), of the web space between thumb and index finger, and softening and flattening of the hypothenar eminence with sparing of abductor pollicis brevis indicates an ulnar nerve lesion.

Isolated wasting of abductor pollicis brevis indicate median nerve lesion in carpal tunnel syndrome.

Global wasting of hand indicate median and ulnar nerve lesion; probably, with damage to T1 root.

More extensive arm wasting may indicate any of the following: syringomyelia of MND; bilateral, symmetrical wasting indicate peripheral neuropathy.