Year 2003 Paper two: Questions supplied by Tricia

Question 97
A 63-year-old man is brough to the emergency department by his family with a one-hour history of continuing abnormal behaviour following an argument with his wife. His speech is fluent and his comprehension is normal. He has no recollection of the argument or the events leading up to his presentation. He continually requires reassurance regarding the current situation and has difficulty remembering the advice given. His long-term memory is intact. Neurological examination is otherwise normal. A cranial computed tomography (CT) scan is normal

The most likely diagnosis is:

A. conversion disorder  
B. transient global amnesia  
C. temporal lobe epileptic event  
D. thalamic stroke  
E. basilar artery thrombosis

Conversion disorder

Conversion disorder refers to symptoms or deficits of voluntary or sensory function suggesting a neurologic or general medical condition and associated with psychological factors. Typically there is a sudden onset of a dramatic but physiologically impossible condition like paralysis, aphonia, blindness, deafness, or pseudoseizures. The presentation fits the patient's view of the disorder rather than physiology. Unlike somatization disorder, patients with conversion disorder focus upon only one symptom. Careful neurologic examination documenting an intact system rules out an organic cause. Although the rate of misdiagnosis of conversion disorder (medically unexplained sensory, seizure, or paralysis) was 29 percent in the 1950s, published studies since the 1970s show only 4 percent of patients diagnosed with conversion are subsequently found to have a documented medical illness.

Conversion disorder tends to occur in young, naive, uneducated women and is not in the conscious control of the patient. It is often associated with an emotional conflict that is not easily resolved. Comorbid depression, psychosis, or neurologic disease should be considered. Personality disorder, dissociative disorder, and posttraumatic stress disorder may also be present. Patients often respond to suggestion or persuasion.

Transient global amnesia

Transient global amnesia (TGA) is a syndrome characterized by the acute onset of severe anterograde amnesia accompanied by retrograde amnesia, without other cognitive or focal neurologic impairment. The amnesia resolves within 24 hours. Most patients are middle aged or older adults. Episodes are usually not recurrent, but rare patients have infrequent attacks that recur over several years.

The etiology of TGA is uncertain. Most TGA episodes are probably related to vasoconstriction, but some may be caused by transient ischemia or complex partial seizures. TGA can be associated with small focal abnormalities on diffusion-weighted MRI, but the significance of these remains unclear.

Temporal lobe epileptic event

Complex partial seizures — The classification system for epileptic seizures includes several seizure types that are characterized by an abrupt loss of consciousness: complex partial seizures ("complex" means that consciousness and awareness of the surroundings are lost), absence seizures, and generalized tonic-clonic seizures (also known as convulsions; "tonic" refers to muscle stiffening and "clonic" refers to muscle jerking)

Complex partial seizures (previously called temporal lobe seizures or psychomotor seizures) are the most common type of seizure in epileptic adults. During the seizure patients appear to be awake but are not in contact with others in their environment and do not respond normally to instructions or questions. They often seem to stare into space and either remain motionless or engage in repetitive behaviors, called automatisms, such as facial grimacing, gesturing, chewing, lip smacking, snapping fingers, repeating words or phrases, walking, running, or undressing. Patients may become hostile or aggressive if physically restrained during complex partial seizures.
Complex partial seizures typically last less than three minutes and may be immediately preceded by a simple partial seizure. Afterward, the patient enters the postictal phase, often characterized by somnolence, confusion, and headache for up to several hours. The patient has no memory of what took place during the seizure other than, perhaps, the aura.

The behaviors that typify complex partial seizures are not specific for epileptic seizures and may be observed in association with NES.

**thalamic stroke**

Thalamic hemorrhage — A thalamic hemorrhage may extend in a transverse direction to the posterior limb of the internal capsule, downward to put pressure on the tectum of the midbrain, or may rupture into the third ventricle. Symptoms include hemiparesis, hemisensory loss, and occasionally transient homonymous hemianopsia. There may also be an upgaze palsy with miotic pupils that are unreactive, peering at the tip of the nose, skewed, or "wrong way eyes" toward the weak side (in contrast to hemispheric cortical injury in which the eyes are deviated away from the hemiparesis). Aphasia may occur if the bleed affects the dominant hemisphere or neglect in the nondominant hemisphere.

**basilar artery thrombosis**

BASILAR ARTERY — The basilar artery begins at the medulloponine junction and ends at the junction of the pons and midbrain. Occlusive lesions may occur anywhere along the basilar artery. In addition, thrombi engrafted upon occlusive lesions within the distal intracranial vertebral artery (ICVA), for example, near or at the ICVA-basilar artery junction, can extend into the proximal basilar artery.

Basilar artery occlusive disease most often presents as ischemia in the pons. The major burden of ischemia is in the middle of the pons, mostly in the paramedian base, and often also in the paramedian tegmentum.

The paramedian pontine base contains descending long motor tract and crossing cerebellar fibers. The paramedian tegmentum contains mostly oculomotor fibers. As a result, the predominant symptoms and signs in patients with basilar artery occlusive disease are motor and oculomotor. Sensory and vestibular nuclei and tracts located in the lateral tegmentum are relatively spared.

Alteration in the level of consciousness is an important sign in patients with basilar artery occlusion. They may present with coma when the bilateral medial pontine tegmentum is ischemic.

Motor symptoms and signs — Most patients with symptomatic basilar artery occlusive disease and pontine ischemia have some transient or persistent degree of paresis and corticospinal tract abnormalities. The initial motor weakness is often lateralized and has been referred to as the "herald hemiparesis" of basilar artery occlusion.

Hemiparetic patients with basilar artery occlusion almost always show some motor or reflex abnormalities on the nonhemiparetic side. As examples, slight weakness, hyperreflexia, an extensor plantar reflex, or abnormal spontaneous movements such as shivering, twitching, shaking, or jerking may be present on the relatively spared side. Asymmetry but bilaterality is the rule.

Adventitious movements of the arms and/or legs are occasionally seen and can be prominent. These movements are variable and sometimes intermittent. Small movements may resemble fasciculations. Larger movements may resemble shivering, shuddering, or jerking; another variant is that of tremulous shaking. Voluntary or passive limb movements or painful stimuli may precipitate a flurry of abnormal movements. At times there are large repetitive jerking and twitching movements, especially in limbs contralateral to a hemiparesis. These movements are often misdiagnosed as seizures.

Incoordination of limb movements is another common motor finding. Ataxia is invariably combined with some degree of weakness. Incoordination is usually more severe in the legs. Toe-to-object and heel-to-shin testing usually shows clumsiness and diminished coordination due to cerebellar dysfunction. The ataxia is invariably bilateral but may be asymmetric and more severe on the weaker side. Intention tremor is not common.

Bulbar involvement — Weakness of bulbar muscles is very common and is an important cause of morbidity with pontine infarction due to basilar occlusive disease. Bulbar symptoms include facial weakness, dysphonia, dysarthria, dysphagia, and limited jaw movements. The face, pharynx, larynx, and tongue are most often involved. The pattern may be that of crossed
motor loss, such as weakness involving one side of the face and the contralateral body, but more often the bulbar muscle weakness is bilateral.

Some patients totally lose the ability to speak, open their mouth, protrude their tongue, swallow, or move their face at will or on command. Secretions pool in the pharynx, and aspiration is an important and serious complication. When all voluntary movements other than the eyes are lost but consciousness is retained, the deficit is referred to as the "locked-in syndrome."

Patients with infarction of the pontine base frequently have exaggerated crying and laughing spells and are hypersensitive to emotional stimulus, a condition known as pseudobulbar affect or emotional lability.

Despite the loss of volitional muscle movement, reflexes of the jaw, face, and pharynx may be exaggerated. In addition, clonic jaw movements or clamping down on a tongue blade may occur as a response to attempts to pry the mouth open and to insert a tongue blade.

Some patients with pontine ischemia develop palatal myoclonus (a rhythmic involuntary jerking movement of the soft palate and pharyngopalatine arch) that can involve the diaphragm and larynx. This movement disorder usually begins sometime after the brainstem infarct. The movements of the palate vary in rate between 40 to 200 beats per minute. The movements are readily seen by watching the palate and pharynx when the mouth is open. The movements involve the eustachian tube and make a click that the patient and clinician can hear.

Oculomotor symptoms and signs — Oculomotor symptoms and signs are common with symptomatic basilar artery occlusive disease and pontine ischemia, and few patients with this condition have normal eye movements. Abnormalities include: Complete bilateral horizontal gaze palsy Unilateral horizontal conjugate gaze palsy Unilateral or bilateral internuclear ophthalmoplegia (INO) One-and-a-half syndrome (a conjugate gaze palsy combined with an INO).

Skew deviation of the eyes and ocular bobbing may also be present. Horizontal, gaze-paretic nystagmus is common and, when asymmetric, usually is more prominent when gaze is directed to the side of a unilateral pontine tegmental lesion. Dissociated nystagmus, that is nystagmus that is more severe in one eye and not rhythmically concordant in the two eyes, and vertical nystagmus are found in patients with an INO. Ptosis of the upper eyelids is also very frequent.

The pupils may remain normal or become small. In some patients, the pupils are bilaterally very small ("pinpoint"). Use of a magnifying glass can show that, despite their very small size, the pupillary response to light is preserved, although the amplitude of the response is slight.

Sensory symptoms and signs — Somatosensory abnormalities are generally not prominent in patients with basilar artery occlusions. Paresthesias on one side of the body and limbs reflects involvement of the contralateral medial lemniscus in the paramedian dorsal portion of the basis pontis. Bilateral paramedian lesions that include the medial lemnisci on both sides can cause bilateral paresthesias. Proprioceptive loss is usually minimal or absent despite the paresthesias.

Some patients with basilar artery occlusive disease have unusual burning pain in the face usually located in the center of the face near the midline. Tinnitus and hearing loss relate to involvement of the central auditory tracts and nuclei (auditory nuclei, lateral lemnisci, trapezoid bodies, inferior colliculi) or to ischemia of the eighth nerves or the cochlea.