CASE REPORT

Locked-in syndrome caused by basilar artery ectasia

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Abstract

Despite various presentations of vertebrobasilar artery ectasia, the true incidence and its distribution and the frequency with which it produces symptoms are still uncertain. We report on a case of Locked-in syndrome caused by an ectatic elongated basilar artery, which predisposed to thrombosis and brain stem infarction.

Keywords: basilar artery ectasia, Locked-in syndrome

Case history

A 59-year-old man was admitted to our department with sudden onset of dizziness and left sided weakness whilst at work. There was no headache, nausea or vomiting and no past history of significant illness. He smoked 5 cigarettes/day and consumed a small amount of alcohol occasionally.

Neurological examination revealed a Glasgow Coma Scale (GCS) of 15/15 with slurred speech, normal fundi, and poor gag reflex. No bruit was heard over the carotids.

Five hours later he developed right-sided weakness and became emotionally labile with excessive yawning. His GCS dropped to 13/15, with bilateral facial weakness, horizontal gaze palsy and ocular bobbing; the power was 2/5 with exaggerated reflexes and up going Babinski bilaterally. There were no signs of meningeal irritation. While arrangements for an urgent computed tomography (CT) scan of the head were in progress his GCS dropped further to 8/15, with occasional decerebrate posturing and one short-lived apnoeic episode.

The CT scan showed an abnormally dilated tortuous basilar artery. Diagnosis of basilar artery ectasia was made. This was substantiated later on by CT angiography. He was transferred to the intensive therapy unit (ITU) and commenced on heparin. He remained stable, maintaining his breathing and GCS back to 15/15 but was totally paralysed except for eye movements i.e. Locked-in syndrome. Gastrostomy feeding was started and he was referred for neurorehabilitation.

Discussion

Ectasia

Ectasia of the cerebral arteries can be asymptomatic [1] and may be found during cerebral angiography, but can also lead to devastating neurological manifestations.

Ectasia of the basilar artery is a rare condition and it occurs when the diameter of the basilar artery is greater than normal along all or part of its course, and/or is abnormally tortuous.

The clinical manifestations produced by ectasia of the cerebral vessels are mainly attributed to compression and/or ischaemia.

Locked-in syndrome

Locked-in syndrome (synonyms: cerebromedullospinal disconnection, de-efferented state, pseudocoma) is a condition characterized by paralysis of all voluntary muscles, except for those performing eye movements. Individuals with Locked-in syndrome are conscious, but unable to speak. However they can learn to communicate through an eye blink code. It is a very rare condition...
affecting males and females equally and had been caused by trauma, tumours, and infections of the brain, prolonged anoxia, myelinolysis and polyneuritis.

The case described here demonstrates an unusual manifestation of basilar artery ectasia. The progressive course of this stroke seemed to be due to extension of the thrombus causing bilateral deficits. The aetiology of basilar ectasia has been subject to debate. Pathological studies have shown marked atherosclerosis in some cases [2], while other cases have had alteration in the media and a defect in the internal elastic membrane of the artery wall [3]. It has been suggested that in the latter group the disease may be present from birth and be exacerbated by the effect of hypertension later in the life [4]. Brainstem transient ischaemic attacks and infarction may complicate long-standing symptoms and signs, creating an even more complex clinical picture [4], as in this case.

Cerebral infarcts associated with ectatic basilar artery are due to different mechanisms, for example, arterial thrombosis, artery-to-artery embolism, mass effect with angulation and obstruction of the vertebral and basilar branches.

The association between ectatic basilar artery and stroke is more prevalent in males over the age of 50. Women tend to present at a later age [5]. Systemic hypertension is very common among such patients. It can cause a variety of neurological complications including trigeminal neuralgia [6], glossopharyngeal and vagal neuralgia [7], hemifacial spasm [8], oculomotor palsy [9], hydrocephalus [10–12] and deafness [13]. The presentation may be similar to cerebello-pontine-angle tumour [14]. The patient may present with progressive visual loss and bitemporal hemianopia due to compression of the optic chiasma [15], or homonymous hemianopia due to involvement of the optic tract [16].

Key points
- Basilar artery ectasia is not a rare condition.
- It can be an accidental finding, but can also lead to devastating neurological conditions due to either direct pressure or thrombosis.
- The signs and symptoms can mimic many other neurological diseases.
- Locked-in-Syndrome is a rare complete paralytic (except for eyes) condition; however, cases of complete recovery have been documented.
- Such condition needs urgent cerebral angiography if thrombolytic therapy is to be contemplated.

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References