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Journal

Proceedings of the UCLA Department of Medicine, 16(1)

Authors

Ching, Wendell

Zakzook, Sami

Publication Date

2012-08-29

CLINICAL VIGNETTE

From Adenoma to Alien Hand

Wendell Ching, M.D., Sami Zakzook, M.D.

A 64-year-old male was referred for pre-operative evaluation prior to excision of a left parotid pleomorphic adenoma. He was a right-handed smoker with hypertension, hyperlipidemia and diabetes mellitus. He had a high school education, no personal or family history of any mental disorder and no history of alcohol and drugs use. An asymptomatic right carotid bruit was noted and carotid ultrasound documented a 90% right internal carotid and 70% left internal carotid stenosis. He underwent an uneventful right carotidendarterectomy.

Two months later the patient returned for pre-operative evaluation for his parotid surgery. He reported several days of cough and dyspnea. Chest x-ray revealed mild cardiomegaly, mild pulmonary congestion and small bilateral pleural effusions and the parotid surgery was postponed again. An echocardiogram showed a depressed ejection fraction of 45% and hypokinesis of his left ventricle. Left and right heart catheterization revealed triple vessel coronary artery disease as well as 60% right renal artery stenosis. Angioplasty was performed on the right renal artery and the patient was prepared for coronary artery bypass surgery (CABG).

Bypass surgery was uneventful except for post-operative atrial fibrillation. He was anticoagulated with heparin. On post operation day 9, he developed involuntary movement of his right hand. He reported that his right hand “went on an adventure of its own”, was “flapping around and I had no control over it”. His right hand and forearm felt swollen with a different skin texture. His hand seemed weak and he used his left hand to help move his right hand, which he described as “was not mine”. The episode lasted about 10 minutes. Six hours later he had a similar episode lasting 30 minutes. He initially thought the nurse was stroking the right side of his face and was surprised to see it was his own right hand that was touching his face. His systolic blood pressure was in the 120-130’s and his anticoagulation was therapeutic on the heparin drip during this time.

Initial examination was remarkable for right-sided pronator and parietal drift, mild weakness bilaterally in index finger and 5th finger opposition with thumb.

He remained awake, alert, oriented and appropriate. Brain CT scan showed stable cerebral and cerebellar atrophy without any acute changes. MRI/MRA scan revealed a recent, small, cortical infarct in the left inferior parietal lobe just near the angular gyrus, normal intracranial MRA and an approximate 60% stenosis of the left internal carotid artery. Trans-esophageal echocardiogram did not find a thrombus or a patent foramen ovale.

Four days after the initial episode he underwent successful left carotidendarterectomy and he did not have any subsequent incidents or any other neurologic episodes.

He eventually underwent successful resection of his parotid adenoma... four years later.

Discussion

Alien hand syndrome (AHS) is a perplexing, dramatic, and uncommon clinical diagnosis in which patients experience their limb performing purposeful or seemingly purposeful acts without their intention or control with some patients denying ownership of the affected hand. It was first described in 1908 by Goldstein in patients with corpus callosum tumors showing inability to transfer functions between hemispheres¹⁻³. Its hallmark is the patient's perception of alienation and loss of control over one or both upper limbs coincident with observable complex goal-directed involuntary movements³.

AHS has been associated with other conditions affecting the corpus callosum, such as contralateral frontal strokes, corpus callosum infarction, anterior communicating artery rupture, corpus callosotomy, combination of a posterior corpus callosum lesion and contralateral thalamic sensory lesion, bifrontal penetrating cerebral injury, callosal tumors, and seizures^{3,4}. More recently, it has also been described in patients with Alzheimer's disease⁵, corticobasal ganglionic degeneration³, Cruetzfeldt-Jakob disease⁶ and Multiple Sclerosis⁷.

In 1992 Feinberg reviewed the clinical characteristics and neuroanatomy of 20 reported cases of AHS and classified AHS into two distinct syndromes. The “frontal alien hand syndrome” results from damage to the supplementary motor area, anterior cingulate gyrus, medial prefrontal cortex of the dominant hemisphere, and anterior corpus callosum. The symptoms of frontal alien hand syndrome always occur in the dominant hand, with prominent motor phenomena including reflexive grasping, groping, and compulsive manipulation of tools. This case is an unusual presentation of this category. The second type or “callosal alien hand syndrome” requires only a callosal lesion and is characterized primarily by intermanual conflict, with the non-dominant hand usually affected⁸. In 2000 a third type of AHS was described as “Posterior or Sensory AHS,” involving posterior cortical or sub-cortical areas. These patients have multiple disorders of sensation and sensory processing, with feelings of estrangement from the non-dominant upper limb. Some patients have sensory, optic and cerebellar ataxia, “triple ataxia” of the non-dominant arm^{9,10}.

In corticobasal ganglionic degeneration, the movement abnormality is slightly different as the affected limb can drift or levitate and assume odd postures¹¹.

AHS is usually persistent but has been reported as a paroxysmal phenomenon attributable to seizures¹¹.

Our patient was at high risk for a cardiovascular event. He had a small left inferior parietal lobe stroke but the corpus callosum was not affected and this stroke should not have caused his transient symptoms. The left internal carotid stenosis would put him at risk of hypoperfusion the anterior and middle cerebral artery circulation watershed area including the peri corpus callosum area. Lesions in these areas have been described to cause AHS⁸. This case is clinically consistent with a transient ischemic attack manifested as AHS, which was eliminated after the left carotidarterectomy. It might be considered as a transient form of “frontal AHS “. In reviewing the literature we found one similar case reported by Andre in 1996¹². We believe this is the second reported case of transient AHS.

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Submitted on August 29, 2012