Letters

Progressive Supranuclear Palsy-Like Syndrome After Aortic Aneurysm Repair: A Case Series

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Abstract

The syndrome of progressive supranuclear palsy-like syndrome is a rare complication of ascending aortic aneurysm repair. We report two patients with videos and present a table of prior reported cases. To our knowledge there is no previously published video of this syndrome. The suspected mechanism is brainstem injury though neuroimaging is often negative for an associated infarct. We hope our report will increase recognition of this syndrome after aortic surgery, especially in patients with visual complaints.

Keywords: Progressive supranuclear palsy, supranuclear gaze palsy, aortic aneurysm repair

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Introduction

We present and demonstrate by video two unusual patients with a progressive supranuclear palsy-like syndrome following ascending aortic aneurysm repair. While previous patients have been reported with the disorder, to our knowledge no video has been published. The rarity of the condition and the availability of videos of both patients may be of value to readers of the journal.

Cases

A 25-year-old man with a family history of medial arterial dissections developed severe dysarthria and severe saccadic gaze palsy after a complicated repair of an ascending aortic aneurysm (Video 1). He noticed his impaired vision in the days following the repair. He was completely unable to generate saccades, and navigated by turning his head to fix a target and then tracking it. Mild parkinsonism with masked facies was also present. He also had a slight delay in reopening his eyes after forcefully closing them, suggestive of dystonia. Brain magnetic resonance imaging (MRI) revealed hyperintensities in the splenium of the corpus callosum and left frontal lobe. He ultimately succumbed 2 years later to a chronic bacterial infection of the chest cavity.

Our second patient, a 53-year-old man with hypertension and hyperlipidemia, underwent two sequential repairs of a dissecting ascending aortic aneurysm. After his first repair, he had right occipital infarct seen on MRI and several transient ischemic attack-like episodes. During the following 6 weeks he developed prominent dysarthria, dysphagia, and gait imbalance with a left homonymous hemianopia. After his second repair for progressive dilatation of the proximal descending thoracic aorta, he had progression of his neurologic symptoms, becoming nearly anarthric and also requiring a walker to walk. He had no response to Sinemet 25/100 mg three times daily. A horizontal and vertical saccadic gaze palsy was present; however, there was some preservation of ability to generate rightward voluntary eye movements (Video 2). He had bilateral dysmetria on the finger-to-nose test. No rest tremor or bradykinesia was found, though he had mildly spastic tone. Brain MRI was significant for a right occipital infarct only.

The syndrome of saccadic gaze palsy with parkinsonism is a rare and devastating complication of ascending aortic aneurysm repair.1,2 Although some patients have been reported with infarcts in the pons, substantia nigra, centrum semiovale, frontal subcortex, striatum, corona radiata, internal capsule, and basal ganglia,3 the saccadic gaze...
| Case | Age/Gender | Procedure | Complications | Initial Signs and Symptoms | Later symptoms | Time Course | MRI findings |
|------|------------|-----------|---------------|---------------------------|----------------|-------------|--------------|
| 1    | 25/M       | AVR and resection of infected graft | Graft infection | Vision difficulty | Dysarthria, dysphagia, unsteady gait | 2 months | Small T2/FLAIR hyperintensity in splenium of corpus collosum and another in frontal lobe |
| 2    | 53/M       | AAA repair and repair dissecting ascending aortic aneurysm | Descending aortic aneurysm dissection following initial repair | Dysarthria, dysphagia, gait imbalance | Anarthria, further gait imbalance | 6 weeks | Right occipital infarct |
| 3    | 56/M       | Resection of AA and AVR | SNGP, mild gait instability, dysarthria | Marked unsteady gait, dysarthria, SNGP | After 3-4 months | WNL |
| 4    | 45/F       | Resection of acute aortic dissection | SNGP, transient memory deficits | Marked unsteady gait, dysarthria, SNGP | After 2 months | WNL |
| 5    | 52/M       | Resection of acute aortic dissection | SNGP, unsteady gait | Marked unsteady gait, SNGP, dysarthria, dysphagia, several partial seizures | 3-4 months | Subtle T2 signal abnormality mesial temporal lobes |
| 6    | 44/M       | Resection of AA, AVR | SNGP | Unsteady gait, dysarthria, dysphagia, SNGP, dystonic pharyngeal movements | Several weeks | MRI WNL, MRA with mild anomalous irregularities of MCA |
| 7    | 57/M       | Repair AV and ascending aorta | SNGP | Unsteady gait, SNGP, dysarthria | 5 months | Tiny lacunar infarct caudate head |
| 8    | 50/M       | AAA repair and AVR | SNGP, unsteady gait | SNGP, unsteady gait, dysarthria | 2 months | Head CT old R cerebral infarct |
| 9    | 45/F       | Resection of AAA, AVR | Mild dysarthria and dysphagia, probable SNGP | Dysarthria, dysphagia, drooling, gait unstead, SNGP | 2 months | WNL |
| 10   | 65/M       | AA repair | Hypotension | Dysarthria, dysphagia | Reduced vertical gaze and gait instability | 6 months | Hypoxic-ischemic bilateral striopallidal lesions |
| 11   | 64/M       | AAA repair and AVR | SNGP, balance difficulty, dysarthria | Not provided | 2 years | None performed, CT WNL |
| 12   | 41/F       | Repair of patent ductus arteriosus | SNGP, dysarthria, gait difficulty | Progressive gait difficulty | 5 years | MRI WNL, MRA narrow P1 segment of L PCA |
| 13   | 44/F       | Several repairs of aortic dissection and AVR | SNGP, dysphagia | Not provided | 10 years | Periventricular small vessel changes, MRA narrow P1 segment of PCA |
| Case | Age/Gender | Procedure | Complications | Initial Signs and Symptoms | Later Symptoms | Time Course | MRI findings |
|------|------------|------------|---------------|---------------------------|---------------|-------------|-------------|
| 14<sup>4</sup> | 46/M | Resection of malignancy from right atrium | 3 minutes circulatory arrest | SNGP, dysphagia and drooling | Not provided | 4 months | WNL |
| 15<sup>4</sup> | 45/F | AVR | | SNGP, emotional lability | Not provided | 10 months | None performed, CT WNL |
| 16<sup>4</sup> | 40/M | Aortic dissection repair | | SNGP | Not provided | 10 months | Increased signal L posterior thalamus and L medial temporal lobe |
| 17<sup>4</sup> | 52/M | Repair thoracoabdominal aneurysm | Post-operative hypotension followed by hypertension | SNGP | Not provided | 6 months | Diffuse signal changes, no evidence of infarction |
| 18<sup>4</sup> | 59/M | Aortic dissection repair | Difficulty weaning from cardiopulmonary bypass | SNGP, transient diplopia, R lower facial weakness | Not provided | 2 months | Nondiagnostic, diffusion negative |
| 19<sup>4</sup> | 70/M | Aortic aneurysm repair and AVR | Post-operative septic shock | SNGP, gait difficulty, dysarthria | Not provided | 18 months | Mild diffuse atrophy |
| 20<sup>4</sup> | 56/M | Aortic aneurysm repair and AVR | | SNGP, dysarthria | Not provided | 4 months | Mild periventricular white matter lesions |
| 21<sup>5</sup> | 54/M | AAA repair | Hypoxia | Absent volitional saccades | Dysphagia, bradykinesia, and wide-based gait | 12 months | Chronic microvascular disease L parietal lobe |
| 22<sup>6</sup> | 52/M | AA resection and aortic valve repair | | Slurred speech, unsteady gait | Unsteady gait, absence of saccades | 3 months | Small acute infarcts R cerebellar hemisphere and both sensory motor cortices |
| 23<sup>6</sup> | 37/M | Aortic root repair and AVR | | Blurred vision, dysphagia, imbalance | Slow small amplitude saccades | 2 months | WNL |
| 24<sup>6</sup> | 70/F | AVR, aorta resection, aortic arch replacement | | Blurred vision, trouble tracking objects | Small slow horizontal volitional saccades | 8 months | WNL |

Case number with superscript = reference number (see list of references).
MRI = Magnetic Resonance Imaging, AAA = ascending aortic aneurysm, AA = aortic aneurysm, AVR = aortic valve replacement, SNGP = supranuclear gaze palsy, WNL = within normal limits, MRA = Magnetic Resonance Angiogram, CT = Computer Tomography.
Saccadic gaze palsy is characterized by slow, hypometric saccades and absent quick phases of optokinetic nystagmus, with intact vestibular ocular reflexes. Vertical saccades may be affected in isolation, though both vertical and horizontal saccades are typically impaired. As in most cases (summarized in Table 1), our patients’ MRIs did not reveal a brainstem injury, likely because of insufficient imaging resolution. Importantly, one of our cases did have an occipital infarction as evidence of posterior circulation ischemia. The mechanism of injury in this syndrome remains unclear, although a perioperative ischemic stroke from embolism, hypothermia protocol, hypotension, hyperviscosity, or cardiopulmonary bypass is possible. A possible location of embolism may be in the posterior thalamo-subthalamic paramedian artery, a branch of the proximal posterior cerebral artery, which supplies an area of the rostral midbrain that is crucial to generation of vertical saccades. Because it is difficult to unify the constellation of supranuclear gaze palsy with dysarthria, dysphagia, and gait imbalance into a single infarct, multiple embolic infarcts are likely. Many cases involved surgery of the ascending aorta, suggesting multiple micro-emboli to the posterior circulation that may have been too small to generate symptoms in the anterior circulation. This syndrome is usually permanent, and symptomatic treatment is rarely successful, although treatment with levodopa, dopamine agonists, and anticholinergic agents has been attempted.

**Discussion**

Whether or not this devastating syndrome can be prevented is still uncertain, as is the possibility that more limited forms of the syndrome may be more common, perhaps overlooked in the immediate postoperative state. To improve accurate diagnosis, dynamic eye movements such as saccades should be assessed in any patient with visual complaints after aortic artery surgery.

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