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Tremor and Other Hyperkinetic Movements

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

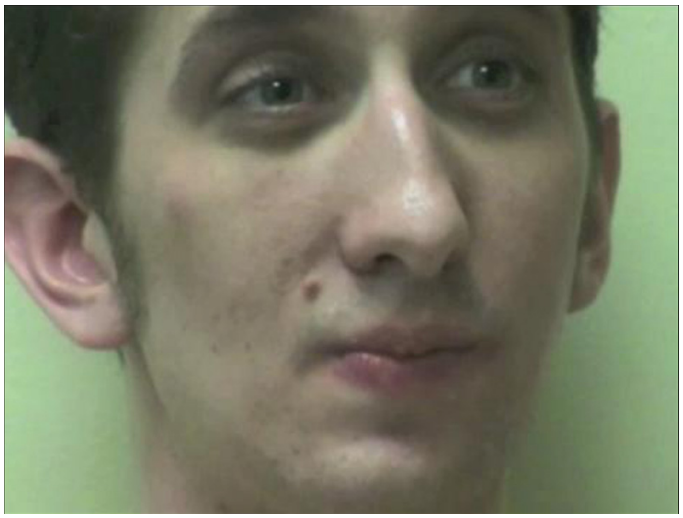
Table 1. Cases of Progressive Supranuclear Palsy-like Syndrome After Aortic Surgery

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 descending aortic aneurysm	Descending aortic aneurysm dissection following initial repair	Dysarthria, dysphagia, gait Imbalance	Anarthria, further gait imbalance	6 weeks	Right occipital infarct
3[1]	56/M	Resection of AA and AVR		SNGP, mild gait unstability, dysarthria	Marked unsteady gait, dysarthria, SNGP	After 3-4 months	WNL
4[1]	45/F	Resection of acute aortic dissection		SNGP, transient memory deficits	Marked unsteady gait, dysarthria, SNGP	After 2 months	WNL
5[1]	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[1]	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[1]	57/M	Repair AV and ascending aorta		SNGP	Unsteady gait, SNGP, dysarthria	5 months	Tiny lacunar infarct caudate head
8[1]	50/M	AAA repair and AVR		SNGP, unsteady gait	SNGP, unsteady gait, dysarthria	2 months	Head CT old R cerebral infarct
9[1]	45/F	Resection of AAA, AVR		Mild dysarthria and dysphagia, probable SNGP	Dysarthria, dysphagia, drooling, gait unst, SNGP	2 months	WNL
10[2]	65/M	AA repair	Hypotension	Dysarthria, dysphagia	Reduced vertical gaze and gait instability	6 months	Hypoxic-ischemic bilateral striopallidal lesions
11[4]	64/M	AAA repair and AVR		SNGP, balance difficulty, dysarthria	Not provided	2 years	None performed, CT WNL
12[4]	41/F	Repair of patent ductus arteriosis		SNGP, dysarthria, gait difficulty	Progessive gait difficulty	5 years	MRI WNL, MRA narrow P1 segment of L PCA
13[4]	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

Table 1. Continued

Case	Age/ Gender	Procedure	Complications	Initial Signs and Symptoms	Later symptoms	Time Course	MRI findings
14[4]	46/M	Resection of malignancy from right atrium	3 minutes circulatory arrest	SNGP, dysphagia and drooling	Not provided	4 months	WNL
15[4]	45/F	AVR		SNGP, emotional lability	Not provided	10 months	None performed, CT WNL
16[4]	40/M	Aortic dissection repair		SNGP	Not provided	10 months	Increased signal L posterior thalamus and L medial temporal lobe
17[4]	52/M	Repair thoracoabdominal aneurysm	Post-operative hypotension followed by hypertension	SNGP	Not provided	6 months	Diffuse signal changes, no evidence of infarction
18[4]	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[4]	70/M	Aortic aneurysm repair and AVR	Post-operative septic shock	SNGP, gait difficulty, dysarthria	Not provided	18 months	Mild diffuse atrophy
20[4]	56/M	Aortic aneurysm repair and AVR		SNGP, dysarthria	Not provided	4 months	Mild periventricular white matter lesions
21[5]	54/M	AAA repair	Hypoxia	Absent volitional saccades	Dysphagia, bradykinesia, and wide-based gait	12 months	Chronic microvascular disease L parietal lobe
22[6]	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[6]	37/M	Aortic root repair and AVR		Blurred vision, dysphagia, imbalance	Slow small amplitude saccades	2 months	WNL
24[6]	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.



Video 1. **The Patient at Initial Visit.** The patient is unable to initiate horizontal or vertical saccades. However, tracking his cellphone with auditory cues enabled the patient to look in all directions. Mild facial masking and dystonia are also present.

palsy is due to brainstem injury.[4] 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.[5] 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.[6] 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.[4] 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.

Video 2. **The Patient at Initial Visit.** The patient demonstrates facial masking and a mild quizzical stare. He demonstrates a saccadic gaze palsy, with inability to look left or vertically on command. Rightward eye movements are possible, but attempts at saccades demonstrate severe slowing.

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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