Upper Airway Dysfunction in Olivopontocerebellar Atrophy

Philip L. Schiffman, M.D., F.C.C.P., and Lawrence I. Golbe, M.D.

We report the findings in a patient known to have olivopontocerebellar atrophy who developed respiratory distress, inspiratory stridor, and maximum inspiratory and expiratory flow volume loops. Treatment with carbidopa-levodopa gave symptomatic relief.

(CHEST 1992; 102:1291-92)

MFVL = maximum flow-volume loop; OPCA = olivopontocerebellar atrophy

Upper airway dysfunction, identified by abnormal maximum inspiratory and expiratory flow volume loops (MFVLs), has previously been reported in patients with Parkinson's disease and other extrapyramidal disorders. This report describes a patient known to have olivopontocerebellar atrophy (OPCA) who developed symptomatic upper airway dysfunction.

CASE REPORT

The patient is 34 years old. He was well until the age of 14 years, when his family noted him to be "clumsy." He joined the army at the age of 18 and had no difficulty with marching or other physical tasks until the age of 22, when ataxic gait and tremor began. The symptoms progressed gradually, during which time the patient worked as an auto mechanic. He was discharged from the army at the age of 28 yr and became confined to a wheelchair by age 31. There is no history of sphincteric disturbance, orthostatic dizziness, temperature intolerance, alcohol or other drug abuse, malignancy, asthma, or other lung disease. There is a 5-pack-yr cigarette smoking history.

Six relatives with similar illness in three generations are known: the patient's maternal grandfather, mother, maternal uncle and aunt (there were also three unaffected members of that sibship), and two brothers (the patient also has four unaffected siblings). The familial pattern is compatible with an autosomal dominance.

An MRI scan at the age of 32 years revealed minimal cerebellar atrophy and no other abnormalities. Levels of copper, ceruloplasmin, and glutamate dehydrogenase and lipoprotein electrophoresis were normal. There are no postmortem data available on any family member.

On physical examination the patient was unable to arise from a wheelchair without maximal assistance. The blood pressure was 122/80 mm Hg, and the pulse rate was 88 beats per minute, without orthostatic drop. Mentation was intact. There was severe horizontal gaze-evoked nystagmus. Other cranial nerves, including pupillary light reflex and range of ocular gaze, were normal. There was a moderate to severe cerebellar dysarthria and severe ataxia of trunk and limbs with head titubation. Strength and sensation were intact. There were no parkinsonian features. Reflexes were 2+ and symmetric, with downgoing toes. Significant inspiratory stridor was audible across the room. No other cardiac or pulmonary abnormality was elicited. The chest roentgenogram, CBC, and arterial blood gas levels (pH = 7.39; PaCO<sub>2</sub> = 39 mm Hg; PaO<sub>2</sub> = 96 mm Hg) were normal. The MFVLs (Fig 1) were consistent with a variable extrathoracic airway obstruction. A diagnosis of laryngeal dysfunction secondary to OPCA was considered likely, and a trial of carbidopa-levodopa (25-100, three times per day) was initiated. Theophylline and albuterol were discontinued. The patient returned four weeks later without respiratory complaint. The stridor had resolved, and the MFVLs were improved. Spirometric data are presented in Table 1. Pulmonary function studies performed at another institution five months prior to referral were reviewed. They included computer-generated MFVLs also consistent with a variable extrathoracic obstruction.

DISCUSSION

Olivopontocerebellar atrophy is a chronic degenerative neurologic illness with complex and variable manifestations and controversial nosology. The most prominent and disabling element is usually cerebellar, with dystarthisms, nystagmus, ataxia, and hypotonicity. Some patients develop parkinsonian signs, with bradykinesia, rigidity, masked facies, and other signs; all are usually poorly responsive to levodopa. Mentation is usually spared. Some patients develop prominent autonomic insufficiency, with postural hypotension, sphincteric disturbances, and pupillary abnormalities, in which case the rubric of "multiple system

Table 1—Spirometry Before and After Treatment

<table>
<thead>
<tr>
<th>Data</th>
<th>Before Therapy</th>
<th>After Therapy</th>
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<tbody>
<tr>
<td>FVC, L</td>
<td>3.23</td>
<td>3.25</td>
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<td>1.90</td>
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<td>PEFR, L/s</td>
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<td>3.20</td>
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<td>FEF&lt;sub&gt;50&lt;/sub&gt;, L/s</td>
<td>1.80</td>
<td>1.80</td>
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<tr>
<td>PIFR, L/s</td>
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<td>1.65</td>
</tr>
<tr>
<td>FIF&lt;sub&gt;25&lt;/sub&gt;, L/s</td>
<td>1.18</td>
<td>1.34</td>
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<tr>
<td>FIF&lt;sub&gt;75&lt;/sub&gt;, L/s</td>
<td>0.39</td>
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atrophy” applies. Polynuropathy is also common.\textsuperscript{3} The majority of cases occur sporadically, but when OPCA is inherited, by far the most common mode of transmission is autosomal dominance, occurring in 39 percent of the cases of OPCA in one review.\textsuperscript{4}

Although stridor and symptomatic upper airway obstruction is thought to be unusual in OPCA, recent reports of nocturnal stridor\textsuperscript{\textdagger} and sleep apnea\textsuperscript{7} have brought attention to this potential problem. The former was partially improved by a surgical procedure and the latter pharmacologically with trazodone. A single case of respiratory failure secondary to vocal cord dysfunction that required tracheostomy has also been described.\textsuperscript{8}

The patient reported herein had severe upper airway dysfunction that, left untreated, would probably have led to respiratory failure. His presentation was similar to that of a previously reported 66-year-old man with Parkinson's disease who was also initially diagnosed as having asthma, but whose condition was unresponsive to bronchodilators and eventually improved when treated with carbidopa-levodopa and trihexyphenidyl.\textsuperscript{9} Although most of the present patient's nonrespiratory symptoms were secondary to cerebellar disease, parkinsonian manifestations are known to occur late in OPCA, and the MFVLs were consistent with the abnormal type-B pattern described by Vincken et al\textsuperscript{10} in patients with extrapyramidal disorders. Therefore, a trial of carbidopa-levodopa therapy was initiated. With treatment, the patient's respiratory symptoms and stridor resolved, his inspiratory flow rates improved, and a tracheostomy was avoided. The improvement has been sustained, and the carbidopa-levodopa therapy has been continued. While it is impossible to state absolutely that the patient's improvement is a result of the therapy, the prolonged and progressive nature of the respiratory symptoms prior to treatment make spontaneous resolution unlikely. Under similar circumstances, in a patient with OPCA exhibiting stridor, a trial of carbidopa-levodopa would be warranted.

**REFERENCES**


**Thrombosed Pulmonary Artery Aneurysm**

**A Rare Cause of a High-Probability Lung Scan**

Cheryl L. Fields, M.D., F.C.C.P.; Thomas M. Roy, M.D., F.C.C.P.; and Miguel A. Ossorio, M.D., F.C.C.P.

The high-probability ventilation-perfusion lung scan is accepted as supportive of pulmonary embolism and often negates further diagnostic evaluation; however, there are processes that mimic the clinical presentation and radiographic findings of pulmonary emboli, including a unilateral segmental or greater perfusion defect. We present the findings in a patient whose presentation and ventilation-perfusion scans over a three-month course were suggestive of pulmonary embolism, yet pulmonary angiography revealed a thrombosed pulmonary artery aneurysm. The interpretation of a unilateral segmental perfusion defect as high probability does not secure the diagnosis of pulmonary embolism and should not preclude further evaluation for alternative etiologies.

(Chest 1992; 102:1292-94)

For the past decade the ventilation-perfusion scan has played an important role in the evaluation of the patient with a suspected pulmonary embolism.\textsuperscript{1} By current criteria a scan with one or more segmental or greater perfusion defects and normal ventilation has a recognized high probability for pulmonary embolism, and no further evaluation is indicated;\textsuperscript{2} however, this perfusion pattern, especially when unilateral in presentation, may be mimicked by nonthromboembolic processes that may necessitate further diagnostic evaluation.\textsuperscript{4} We present the findings in a patient in whom the clinical diagnosis of pulmonary embolism was suggested by multiple high-probability perfusion scans, yet who was found to have a ruptured pulmonary artery aneurysm. This process has not been reported previously as a cause of high-probability ventilation-perfusion scans.

**CASE REPORT**

A 36-year-old white woman was admitted with a one-week history of dyspnea. She denied chest pain, cough, or hemoptysis. The patient had quit smoking three months earlier after a 7-pack-year-history. She denied illicit drug use. A chest roentgenogram revealed a right midfield infiltrate. An arterial blood sample measured a pH of 7.46, a PaO\textsubscript{2} of 77.5 mm Hg, and a PaCO\textsubscript{2} of 32.4 mm Hg, with an alveolar-arterial gradient of 31.7 mm Hg. The differential diagnosis included pulmonary embolism. A ventilation-perfusion scan was obtained and interpreted as high probability for pulmonary embolism (Fig 1). The patient received anticoagulative therapy and was discharged on warfarin therapy.

Two months later, the patient was admitted to a different hospital with a three-day history of right-sided pleuritic chest pain, dyspnea, and a nonproductive cough. Although the patient claimed compliance with her warfarin sodium (Coumadin) therapy, her prothrombin time at admission was 11.5 s, with a control of 13.4 s. A chest roentgenogram displayed decreased vascularity in the right hemi-

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