Pneumonia Preceding Respiratory Failure*

A Rare, Easily Misleading Clinical Manifestation in Adult Arnold-Chiari Malformation

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A 47-year-old woman was admitted for bilateral lower lobe pneumonia with respiratory distress. Two episodes of respiratory failure developed despite improvement of pneumonia after antibiotic chemotherapy. Loss of consciousness and quadriplegia accompanied the last episode of respiratory failure. Arnold-Chiari malformation type 1 was diagnosed and a suboccipital craniectomy was performed. The neuromuscular and respiratory disorders greatly improved after operation. We believe that ACM 1 should be considered when an adult develops unexpected respiratory failure after improvement of the primary pulmonary condition. This disease is potentially treatable by surgical management, and if it is misdiagnosed, will be fatal.

(Chest 1991; 99:1294-95)

ACM 1 = Arnold-Chiari malformation, type 1

Arnold-Chiari malformation is defined as a condition where part of the cerebellar tissue is depressed caudally toward the medulla and spinal cord into the cervical canal and is associated with medulla and fourth ventricle herniation into the cervical canal. These cases can be divided into two major types. Arnold-Chiari malformation type 1 consists of variable displacement of the medulla oblongata through the foramen magnum into the cervical canal, but the cerebellar herniation is much more prominent. The lower cranial nerves are then displaced downward and have an ascending course to their points of exit.1 About one third of these patients may have syringomyelia, and this type of Arnold-Chiari malformation occurs mainly in adults.2

Respiratory depression as the early clinical manifestation is very rare in ACM type 1 and mostly is the result of postoperative conditions.3 We report an adult patient, with previously undiagnosed ACM 1, who presented with a bilateral lower lobe pneumonia followed by two unexpected respiratory failures.

CASE REPORT

A 47-year-old woman was admitted with complaints of productive cough, fever and respiratory distress for three days. She also had had swallowing disturbance and numbness over the tips of fingers for more than two months. There was no other important past medical history.

Physical examination showed a slightly overweight woman breathing with effort but alert and oriented. There were moist crackles

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Figure 1. A computerized tomographic scan with intrathecal iopamidol injection of the posterior fossa and upper cervical region revealed dilated ventricles and bilateral tonsillar herniation down to C1-2 level. The medulla and high cervical cord were compressed and deformed.

over both lower lung fields. Neurologic examination showed intact cranial nerves, except for the absence of gag reflexes. Motor, sensory and deep tendon reflexes of the extremities were intact and symmetric.

Laboratory test results were within normal limits, except a white blood cell count of 7100/cu mm but with a left shift. Arterial blood gas values revealed hypoxemia with hypocapnia (pH: 7.35, PaO2, 63 mm Hg, and PaCO2, 32.8 mm Hg). Chest roentgenogram showed bilateral lower lung infiltrations but with normal diaphragmatic movement. She was treated for bilateral pneumonia with antibiotic and oxygen therapy.

Despite improvement of the clinical condition and chest roentgenographic lesions, an episode of apnea developed one week after admission. This condition was attributed to sputum suffocation, and further evaluation was not done. After mechanical ventilatory support for one week, she recovered spontaneous breathing but was still tachypneic with a respiratory rate around 20 to 30 per minute. Unfortunately, recurrent apnea with loss of consciousness preceded by three episodes of generalized tonic-clonic seizures developed 12 days after the first episode of apnea. After ventilator support, her consciousness returned but quadriplegia remained. Neurologic examination showed absence of muscle power over all four limbs, with bilateral positive Babinski's sign. A computerized tomographic scan with intrathecal iopamidol injection of the posterior fossa and upper cervical region revealed dilated ventricles and bilateral tonsillar herniation down to C1-2 level. The medulla and high cervical cord were compressed and deformed. There was no evidence of syringomyelia (Fig 1).
ACM 1 was diagnosed, and the patient received suboccipital craniectomy; decompressive laminectomy at C1-2 level and duroplasty. She had a smooth postoperative course and was weaned from the ventilator two days after the operation. There was no further respiratory impairment and muscle power of her limbs improved gradually. Three months after operation, she could walk with support, and the swallowing problem resolved.

**DISCUSSION**

ACM 1 usually has the clinical presentations of pain (mainly headache), weakness, numbness of the limbs, unsteadiness and loss of balance. 

Although respiratory failure may occur after operation in ACM 1, it is rarely seen as a clinical manifestation before operation. 

Up to now, there were only a few reports.

Bullock et al presented two patients with respiratory failure as the initial manifestation of ACM associated with syringomyelia. Both of these patients had bilateral paralysis of the diaphragm confirmed by roentgenogram. This was explained by the phrenic nerve palsy due to the compression of syringomyelia. But Fish et al described two patients with ACM 1 without associated with syringomyelia and paralysis of diaphragm who developed respiratory arrest before the diagnosis of ACM 1. Therefore, respiratory failure alone may occur in ACM 1 whether it is associated with syringomyelia or not. The compression of the brain stem in ACM 1 can result in central respiratory failure even if the phrenic motor neuron or phrenic nerve are spared. This was consistent with our patient's manifestation.

Bokinsky et al described one patient with ACM 1 associated with syringomyelia who developed acute respiratory failure with total loss of hypoxic drive and minimal loss of hypercapnic drive. This finding was explained by the impairment of peripheral chemosensitivity due to the ninth cranial nerve dysfunction that interrupted peripheral chemoreceptor afferents. In our patient, despite absence of gag reflex, the hypoxic drive seemed intact as evidenced by hypoxemia with tachypnea and hypcapnia.

The pneumonia in our patient might have been either community acquired or resulted from aspiration due to a swallowing disturbance. Although it might have aggravated the respiratory distress, it was not the cause of the respiratory failure. Physicians should look for underlying neuromuscular disorders such as ACM 1 in patients who develop unexpected respiratory failure. ACM 1 is curable by surgical treatment, but if it is missed, it may be fatal.

**REFERENCES**


**Exercise-Induced Ventricular Tachycardia in Association with a Myocardial Bridge**

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Sustained ventricular tachycardia induced by exercise is uncommon. This is a report of a patient who has a positive exercise test at a high level of exercise. The electrocardiogram has classic ischemic ST depressions. Following the appearance of these ST depressions, the patient developed ventricular tachycardia at a rapid rate. Workup that included an echocardiogram and cardiac catheterization revealed myocardial bridging of the left anterior descending artery as the only structural abnormality. Electrophysiologic studies revealed the patient to have ventricular tachycardia only with isoproterenol (Isuprel) infusion. (Chest 1991; 99:1295-96)

IHSS = idiopathic hypertrophic subaortic stenosis; PTCA = percutaneous transluminal coronary angioplasty

Sustained ventricular tachycardia induced with exercise is uncommon. Little is written in the literature regarding this entity. When exercise-induced ventricular tachycardia does occur, it is usually associated with coronary artery disease. However, certain conditions such as mitral valve prolapse, aortic stenosis, idiopathic hypertrophic subaortic stenosis (IHSS), and the prolonged QT syndrome have been associated with exercise-induced ventricular tachycardia. In other patients, no structural abnormalities are found. We report a case of exercise-induced ventricular tachycardia in a patient with a myocardial bridge causing systolic compression of the left anterior descending artery.

**CASE REPORT**

A 41-year-old man presented with atypical chest discomfort. He is a nonhypertensive, nondiabetic who was referred for a stress test shortly after his mother underwent percutaneous transluminal coronary angioplasty (PTCA) of a tight proximal left anterior

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