Communications for this section will be published as space and priorities permit. The comments should not exceed 350 words in length, with a maximum of five references; one figure or table can be printed. Exceptions may occur under particular circumstances. Contributions may include comments on articles published in this periodical, or they may be reports of unique educational character. Specific permission to publish should be cited in a covering letter or appended as a postscript.

Charcot-Marie-Tooth Disease and Respiratory Failure

To the Editor:

We would like to report our experience with a patient with Charcot-Marie-Tooth disease and respiratory failure. Our observations support the findings of Chan et al (Chest 1987; 91:567-70) that Charcot-Marie-Tooth disease can involve respiratory muscles. Unlike the two cases reported by Chan et al, however, this patient did not have diabetes mellitus, thereby eliminating the possibility of diabetic phrenic neuropathy as a factor contributing to respiratory failure.

A 79-year-old woman presented to her hometown emergency room in April, 1985 with dyspnea, confusion, and lethargy. Arterial blood gas levels at FiO2 of 100 percent taken shortly after tracheal intubation were pH 7.17, Pco2 110 mm Hg and Po2 324 mm Hg. Measured serum bicarbonate level was 40 mEq/L. She was transferred to our hospital four days later after several attempts to wean her from the ventilator were unsuccessful due to persistent hypventilation. She had a 30-year history of progressive, bilateral foot and leg weakness, and had spent the last ten years in a wheelchair. In recent years she had become weak in the hands and arms. She had no previous history of pulmonary disease and had never smoked cigarettes. Examination revealed bilateral atrophic paralysis of gastrocnemius and peroneal muscles, and pes cavus deformities. Muscle atrophy and weakness were present in the intrinsic muscles of the hands. Both shoulders were weak, the trapezius muscles were ribbon-like, and the chest was moderately kyphoscoliotic. No deep tendon reflex was present. No fasciculations, cranial nerve abnormalities or peripheral nerve enlargements were present. Spontaneous tidal volume was 60 to 80 ml, vital capacity was 200 to 250 ml and minute ventilation was 1.5 to 2.0 L/min. The most negative inspiratory force she could generate varied from negative 5 to negative 12 cm H2O and did not improve with correction of mild hypokalemia and hypophosphatemia, the only identified serum chemical perturbations. Thyroid function studies, aldolase, magnesium, anti-nuclear antibody, rheumatoid factor, and creatinine phosphokinase levels were normal. An RPR was nonreactive. Chest radiograph showed reduced lung volume, subsegmental basal atelectasis and a calcified mitral annulus. Compensated mitral insufficiency was confirmed by physical examination and echocardiography. Cerebrospinal fluid was benign. A computed tomogram of the head showed only mild cerebral atrophy. Electromyographic and nerve conduction velocity studies of the lower extremities showed no motor response in the lower limbs and conduction velocities of 27 m/sec in the arms. Additional family history revealed that the patient's mother had hand atrophy. Two siblings had pes cavus, gait disturbance and atrophy of the hands. This woman could not be weaned from the respirator because of persistent hypventilation and lived two years at home with a positive pressure ventilator before dying from a cerebrovascular accident complicating a urinary tract infection.

We believe this woman had Charcot-Marie-Tooth disease because of the progressive atrophic paralysis beginning in the feet, nerve conduction velocity studies compatible with a chronic demyelinating neuropathy, absence of deep tendon reflexes, and findings in her family typical of this disease. We were not able to study this patient's diaphragm function as Chan et al did in their patients. We cannot prove that Charcot-Marie-Tooth disease directly involved any of her respiratory muscles. Kyphoscoliosis was no doubt a factor in this woman's hypventilation, but our judgment is that her respiratory failure far exceeded that which might be expected with her degree of kyphoscoliosis. We would like simply to draw further attention to a possible cause-effect relationship of Charcot-Marie-Tooth disease and respiratory failure so that more observation might be made.

Eric L. Dyer, M.D., F.C.C.P., and Alfred S. Callahan, III, M.D.,
St. Thomas Hospital,
Nashville

Erratum

To the Editor:

On reviewing our histology slides and the photomicrographs published in our article entitled, "Severe Hypoxemia in Farmer's Lung with Normal Chest Roentgenograms" (Chest 1987; 91:274), we found that a non-representative section was photographed. The published figure does not show non-caseating granulomas, as the legend states. This new photomicrograph (Fig 1) of the transbronchial biopsy sample from a representative area shows non-caseating granulomas.

We are sorry for the mistake and hope that this photomicrograph clarifies the matter.

Maqbool Arshad, M.D.,
Milwaukee

FIGURE 1. Photomicrograph of non-caseating granulomas.