before the primary lesion was identified. Thus, despite the absence of an autopsy, we feel that this case represents the first reported example of MFH arising in the lung.

The histologic findings of the lesion on the right, as obtained by bronchoscopy, was identical to that on the left, and included the storiform pattern which is common in primary lesions and less apt to occur in metastatic MFH. This tends to favor multicentric simultaneous origin of the tumor masses rather than metastasis from one lesion to another. Other sarcomas of the lung have occasionally been multiple, and MFH has been previously reported as a multifocal neoplasm. The slow, progressive growth of the tumor mass in the right lung of our patient is consistent with the biologic behavior of primary MFH in other parts of the body. Surgical extirpation is the treatment of choice. Local recurrence is more common than distant metastasis, but cases of metastatic MFH have been reported. The usual pattern is that of multiple, small, rounded pulmonary nodules which are seen on the chest roentgenogram, and the workup commonly reveals evidence of tumor in other organs. This contrasts with the clinical course of our patient.

The light and electron microscopic findings in our pulmonary neoplasm are as described for MFHs in other locations. By electron microscopy rather than a dual fibroblastic and histiocytic differentiation, there is a wide range of differentiation of the undifferentiated mesenchymal cells, resulting in fibroblasts, histiocytes, and intermediate cells. The consensus seems to be that MFH is a mesenchymal sarcoma with an undifferentiated mesenchymal cell origin with potential to originate from supporting structures of various organs. The addition of the lung as another primary site for MFH supports the concept that this tumor may arise in virtually any part of the body. Our ultrastructural findings of undifferentiated cells, along with fibroblasts and histiocytes in different degrees of differentiation, further support the hypothesis that MFH represents a mesenchymal tumor of a primitive or stem-cell origin.

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Cardiac Dysfunction in a Patient With Familial Hypokalemic Periodic Paralysis*

Lynn D. Kramer, M.D.; John P. Cole, M.D.; John C. Messenger, M.D.; and Myroil H. Elledad, M.D.

A 19-year-old white man with familial hypokalemic periodic paralysis developed evidence of cardiac dysfunction during a episode of flaccid paralysis. This consisted of elevated total creatinine phosphokinase (CPK), an increased myocardial fraction of CPK (myocardial band), alteration in the lactic dehydrogenase isoenzyme pattern, severe bradycardia, and evidence of left ventricular dysfunction. These findings, in conjunction with selected cases from the literature, suggest the possibility that cardiomypathy may be a heretofore unrecognized complication of this disorder.

The voluntary muscle dysfunction associated with the affection of familial hypokalemic periodic paralysis (FHPP) is well documented in the literature. The pathologic substrate of this disorder is not clearly defined however.

Research interest in this disorder has centered primarily about the metabolic and histopathologic abnormali-

*From the Department of Medicine, Memorial Hospital Medical Center, Long Beach, CA.
Reprint requests: Dr. Cole, 1021 Pacific Avenue, Long Beach 90813
ties of skeletal muscle. Little attention has been paid to the possibility of myocardial histopathology or alterations in cardiac function. Although there have been reports of transient cardiac dysfunction in FHoPP, these have been relatively sparse and poorly documented. In this paper, we report the occurrence of an elevated total creatinine phosphokinase (CPK) level with an associated rise in the myocardial band (MB) isoenzyme in conjunction with marked bradycardia and hammocking of the mitral valve in a patient with FHoPP.

CASE REPORT

A 19-year-old white man came to the emergency room with his third episode of flaccid tetraparesis and muscular cramping. He had awakened from sleep following a day of heavy exercise and large carbohydrate intake. His history and review of systems were unrevealing.

His family history disclosed that his father and four male relatives had experienced similar episodes throughout their lives, decreasing in frequency with advancing age. The patient also had a brother who had died at 16 years of age from a heart condition and has an older brother who suffers from a similar “heart condition.” Neither of these siblings had FHoPP by history.

The general physical examination revealed a young, white man in no distress. The exam was unremarkable with the exception of a grade 2/6 pansystolic murmur best heard at the apex and intermittent bradycardia with pulse rates varying from 35 to 80 beats per minute. The neurologic exam exhibited flaccid tetraparesis with sparing of the facial and oropharyngeal muscles. Sensory testing was intact. No percussion myotonia was present. Deep tendon reflexes were absent throughout.

The admission laboratory data disclosed a serum potassium level of 1.5 mEq/liter and a CPK value of 545 IU (normal below 50) of which 4 percent was MB band. The remainder of the admission laboratory test results were within normal limits including T₄, magnesium, aldolase, CBC, electrolytes, and 12-factor automated blood chemical analysis. He developed a severe sinus arrhythmia with rates as slow as 35 beats per minute without escape phenomena.

This sinus arrhythmia remained a constant feature of the hospital course although bradycardiac episodes following resolution of the paresis were associated with cardiac rates of not less than 40 beats per minute. On the second day of hospitalization, the CPK fell to 372 IU, MB band detectable qualitatively, SGOT of 33 IU (normal less than 25), and a HBD of

Figure 1. Carotid pulse tracing demonstrating abnormal systolic time intervals as referred to in text.

Figure 2. Echophonocardiogram demonstrating hammocking of mitral valve and pansystolic apical murmur.
Evaluations of his cardiac status included normal chest roentgenogram, pyrophosphate heart scan and His bundle recording. The phonocardiogram (Fig 1) revealed a faint pansystolic murmur that was recorded best at the apex without significant change after administration of amyl nitrate. The echocardiogram (Fig 2) disclosed pansystolic hammocking of the mitral valve that was unchanged by administration of amyl nitrate. The left ventricular ejection time (LVET) (Fig 1), before amyl nitrate, was shortened at 341 msec (normal, 345 to 425 msec), and following amyl nitrate, increased to 408 msec. The pre-ejection period (PEP/LVET) ratio was increased to 0.49 (normal, 0.345 ± 0.038) indicating probable left ventricular dysfunction. These tests were done both on admission and after resolution of the paresis without significant alterations.

The treadmill ECG (Fig 3) demonstrated a downsloping pattern at rest without any ST segment depression. During the first minute of exercise, the patient had 1 mm ST segment depression and increased his ST segment depression to 2 mm at three minutes of exercise, with the T-wave becoming upright at peak exercise. There was no evidence of ST segment depression beyond seven minutes of exercise. The patient exercised nine minutes to a heart rate of 180 beats per minute. This was 90 percent of the predicted maximum, and the patient stopped due to fatigue. During this test, central venous potassium levels were measured which demonstrated a potassium value of 3.2 mEq/liter prior to exercise, 4.8 mEq/liter at peak exercise, and a return to the resting value at 8 and 30 minutes postexercise. The T-wave changes before, during, and after exercise correlated with the serum potassium level in a more exaggerated fashion than would be expected.

A 24-hour Holter monitor showed a significant phasic sinus arrhythmia with frequent episodes of marked sinus bradycardia, with rates as low as 40 beats per minute. There were intermittent, nonspecific T-wave inversions occurring primarily with activity.

Figure 3. Electrocardiographic tracings recorded during treadmill stress test using the Memorial Hospital protocol. Note marked ST segment depression during intermediate stages with resolution near peak exercise load.

Discussion

These studies demonstrate several interesting features in a patient with FHOPP that have not heretofore been reported. Cardiac dysfunction, as illustrated by the episodes of severe bradycardia during the paresis, the less severe bradycardia in the resting state, the shortened LVET, the abnormal but diagnostically equivocal stress ECG, hammocking of the mitral valve, nonspecific T-wave abnormalities on the two-hour Holter monitor, elevated total CPK level with myocardial fraction (MB band) present, and the alteration in the LDH3:LDH2 ratio are of particular interest in this case.

Cardiac dysfunction has been described in relation to the disorder, but these reports share common problems. Most reports are in the older literature, usually German, and those in the English literature are brief notations citing transient cardiac enlargement, bradycardia, or apical systolic murmur. The serum CPK and LDH values are generally reported as normal in the disorder, even in the face of permanent myopathy, although not invariably so. Various drug-induced hypokalemic myopathies have shown a more frequent occurrence of increased serum muscle enzymes including aldolase. Our patient demonstrated an elevation in total CPK and in the MB band on quantitative and qualitative analysis, but no elevation in aldolase was noted. These findings have not been previously reported in this disorder.

The reversal of the LDH3:LDH2 ratio has been discussed in the past in relation to myocardial disease. Our patient had reversal of this ratio while the other LDH isoenzymes remained in normal proportions. Wieme and Herpol reported a similar alteration in LDH3:LDH2 ratio in muscular dystrophy, but they found LDH2 was also elevated. The origin of the elevated enzymes in this case is therefore assumed to be the myocardium and strongly suggests the possibility of an
underlying cardiomyopathy in conjunction with the generalized skeletal muscle involvement.

The literature would seem devoid of microscopic evaluation of cardiac muscle in this disorder. Ionesescu et al. have demonstrated low calcium-binding activity in the sarcoplasmic reticulum of skeletal muscle in this disorder. In view of the similarities in the structure of the sarcoplasmic reticulum in the two types of muscle, the possibility of a similar cardiac muscle dysfunction is intriguing.

The characteristic myopathic vacuoles are present in the diaphragm, yet the number of cases of respiratory distress are few. Campa and Sanders have demonstrated that the postjunctional membrane, at the motor end-plate, retains at least some potential toward depolarization, and that through repetitive nerve stimulation, a temporary recovery in muscle function can be obtained. The recovery in muscle function occurring during the repetitive stimulation may explain not only the infrequency of respiratory failure but also the lack of cardiac dysfunction that is observed.

The etiology of this apparent cardiomyopathy is not clear at this time, and the prospect of more widespread involvement in this disorder requires further evaluation.

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Reexpansion Pulmonary Edema*

Vijay K. Mahajan, M.D., F.C.C.P.; Morris Simon, M.D.; and Gary L. Huber, M.D., F.C.C.P.

A case of pulmonary edema following reexpansion of a collapsed lung due to pneumothorax is described and illustrated. The importance of recognizing this relatively uncommon phenomenon is stressed. The development of such edema can be prevented by avoiding application of sudden and excessive negative pleural pressures during the evacuation of a pneumothorax or a pleural effusion. The edema generally occurs in a lung that has been collapsed for more than three days. The importance of the duration of pulmonary collapse in the causation of edema is demonstrated in this patient.

A number of noncardiac causes of pulmonary edema have been recognized. A rare noncardiac type of pulmonary edema is its occurrence following reexpansion of a lung after collapse due to pneumothorax or pleural effusion. Because of the rarity of its occurrence, we report the findings in a patient who on one occasion developed radiologically evident pulmonary edema in the reexpanded lung following the evacuation of a prolonged pneumothorax, while after another briefer episode of pneumothorax, edema did not develop.

CASE REPORT

An 18-year-old white woman experienced a sudden sharp pain in her right hemithorax three days prior to the present admission. There was clinical and radiologic evidence of rightsided pneumothorax. The underlying lung was completely collapsed (Fig 1). No other significant abnormality was detected on clinical and laboratory examination.

A No. 22F Foley catheter was introduced into the right pleural cavity through the second intercostal space, and the pneumothorax was evacuated by applying suction at a negative pressure of 15 cm H₂O. A repeat chest x-ray film obtained one-half hour later demonstrated complete reexpansion of the previously collapsed right lung; however, at this time, there was an ill-defined patchy consolidation in the lower zone of the right lung that extended throughout the right lung over the next 72 hours (Fig 2). The pulmonary infiltrate progressively decreased within the next 48 to 72 hours (Fig

*From the Departments of Medicine and Radiology, Thorn- dike Laboratory and Beth Israel Hospital, Harvard Medical School, Boston.
Reprint requests: Dr. Huber, 330 Brookline Avenue, Boston 02215

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