native pathogens on bronchoalveolar lavage, as well as the complete clinical and radiographic response to directed antimicrobial therapy support the pathogenicity of \textit{R. henselae} in this patient. To our knowledge, bronchial polyps and interstitial lung disease with associated hilar adenopathy have not been associated previously with BA; however, this clinical picture has been reported with pulmonary \textit{Mycobacterium avium} complex in AIDS patients.\textsuperscript{a}

The agent of BA has been recently identified as \textit{R. henselae}, a rickettsial organism. Tick bites have been epidemiologically linked to transmission of infection, yet this association remains unproven.\textsuperscript{b} Antibiotics reported effective for BA include erythromycin, doxycycline, rifampin, and chloramphenicol.\textsuperscript{b} Because of the potential for intracellular localization of \textit{R. henselae} coupled with slow growth characteristics, protracted administration of antibiotics capable of intracellular penetration has been recommended.\textsuperscript{5} Clarithromycin, a 6-0-methyl derivative of erythromycin that demonstrates unusually high tissue and polymorphonuclear cell penetration, may be a particularly active antibiotic in the treatment of BA.

This case serves to expand the clinical presentation of BA as well as the differential diagnosis of interstitial lung disease with hilar adenopathy and bronchial polyps in HIV-infected patients. Effective antimicrobial regimens should include clarithromycin.

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REFERENCES


Inclusion Body Myositis as a Cause of Respiratory Failure\textsuperscript{*}

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Inclusion body myositis (IBM) is a slowly progressive myopathy that has not been reported to affect respiratory muscles. It is often refractory to treatment and a muscle biopsy specimen is necessary for the diagnosis. This is a report of a patient with IBM who quickly progressed to respiratory muscle failure requiring intubation.

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Inclusion body myositis (IBM) is a form of inflammatory myopathy that is characterized by a protracted course, involvement of distal muscles, and unresponsiveness to steroid therapy. The most severely affected muscles are those of the limbs and, to our knowledge, there are no reports of respiratory muscle involvement. Extramuscular involvement of this disease has been described infrequently and usually involves the cardiovascular and the gastrointestinal systems. We describe a patient with respiratory failure secondary to IBM.

CASE REPORT

A 69-year-old woman was admitted to the ICU with respiratory failure. She had a 10-day history of progressive shortness of breath, cough, lethargy, and weakness. There was no history of fever, sweats, or chills. The patient was hospitalized in 1987 with a similar presentation. At that time, she had complained of a 2-week history of progressive dyspnea and slowly worsening limb weakness for the preceding five years. The electromyogram (EMG) at that time was compatible with a myopathy; however, the creatine kinase (CK) and liver function test results were normal. She was found to have an elevated partial thromboplastin time (PTT) that was shown to be due to a lupus anticoagulant. Her antinuclear antibody (ANA) titer was 1:320. Her platelet count was normal as were the results of her thyroid function tests. A Tension test was negative. The chest radiograph was normal. She required mechanical ventilation and was treated as having mixed connective tissue disease and received large doses of intravenous methylprednisolone sodium succinate (Solu-Medrol). Her hospital course was complicated by multiple infections that were, in part, attributed to the large doses of steroids. As there was no clinical improvement, the steroid therapy was discontinued. The patient underwent a muscle biopsy from the left quadriceps that showed isolated myofiber degeneration, atrophic fibers, and no inflammatory infiltrates. She was discharged from the hospital 1 year later with a diagnosis of idiopathic myopathy. She remained well until the present hospital admission without worsening of the myopathy or further respiratory problems.

The physical examination showed the patient to be in moderate respiratory distress. She had petechiae on both calves and poor air entry in the lungs, which were otherwise clear. The neurologic examination showed her to be alert and oriented, with distal muscle...
The tuhulofilaments showed chest downgoing. Laboratory tests revealed normal respiratory gases with a pH of 7.40, a Pco2 of 49 mm Hg, and a Paco2 of 55 mm Hg on room air. The CK, lactate dehydrogenase, and liver function test results were normal. The chest radiograph was normal. She refused to perform a tidal volume maneuver. The negative inspiratory force was −12 cm H2O. The EMG was again consistent with myopathy.

Her respiratory status quickly deteriorated requiring intubation. The low platelet count was thought to be secondary to immune thrombocytopenia and responded to intravenous immunoglobulin therapy with normalization of the count. A muscle biopsy specimen revealed occasional fibers containing small round or angular vacuoles situated centrally or peripherally. Vacuoles showed granular rimming with the modified Gomori stain (Fig 1). Electron microscopy showed the vacuoles to contain membranous whorls and rare tubofilaments (Fig 2). The features were those of IBM. The patient's course worsened after she developed an upper gastrointestinal tract bleed and sepsis; she died due to complications of the sepsis.

**Figure 1.** Longitudinal frozen section of muscle biopsy specimen showing slit-like rimmed vacuole (modified Masson Gomori trichome, × 500).

**Figure 2.** Electron micrograph of a cytoplasmic vacuole showing membranous whorls (A) and tubofilaments (B) (× 10,000).

**DISCUSSION**

Inclusion body myositis is an inflammatory muscle disorder that was initially thought to be due to a viral etiology although that remains unproven. There is a 2:1 male preponderance, and the onset is usually after age 50 years. The average duration from onset of symptoms to diagnosis has ranged from 5 to 19 years. This disease is usually characterized by a slow but progressive course, distal muscle weakness, and resistance to immunosuppressive therapy. The weakness and atrophy can be asymmetric, with selective involvement of the quadriceps, iliopsoas, triceps, and biceps muscles. Early loss of the patellar reflex can occur due to the quadriceps weakness and a neurogenic disease is often suspected. This disorder can clinically mimic idiopathic polymyositis and in one report accounted for almost one third of the adults with "polymyositis unresponsive to therapy." The occurrence of respiratory failure due to IBM is distinctly unusual.

While dysphagia has been noted to occur in 40 percent of those affected, our patient did not complain of any swallowing difficulties and there was no clinical or radiologic evidence of aspiration. Lotz et al reported a 55 percent incidence of cardiovascular signs and symptoms such as hypertension, ECG changes compatible with ischemia, and rhythm disturbances in their 40 patients. Other diseases such as malignancy and diabetes have been associated with IBM, but to our knowledge, respiratory muscle weakness had not been reported. The positive ANA, immune thrombocytopenia, and antiphospholipid antibody present in our patient give evidence of disturbed immune regulation. Both elevated ANA titers and thrombocytopenia were reported in patients with IBM, and we have found no reported association with lupus anticoagulant. Serum CK levels can be up to 10-fold elevated or normal. Electromyographic findings are those of a myopathy, although there is occasionally evidence of a neuropathy. A muscle biopsy specimen is important in excluding primary neuopathic disease and in diagnosing IBM and distinguishing it from other myopathies that may cause respiratory failure such as polymyositis and systemic lupus erythematosus. Not only is the natural history of IBM different, but since it is refractory to treatment with corticosteroids and cytotoxic agents, the correct diagnosis can spare the patient the side effects associated with these medications. It is important to obtain the biopsy specimen from muscles where the inflammation is active and the biopsy site should be correlated with activity on the EMG.

The inflammation may be distributed in a patchy fashion as likely was the case in our patient and occasionally multiple or repeated biopsy specimens are needed to establish the diagnosis. Review of first biopsy specimen did not show any evidence of IBM. It is likely that the patient had IBM when she was first admitted to the hospital in 1987. However, for the above-mentioned reasons, the diagnosis was not made on the initial muscle biopsy specimen. A low index of suspicion, as IBM is thought to evolve slowly, and the lack of reported respiratory muscle involvement contributed to the lack of earlier recognition in the present case.

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Late Cardiac Strangulation due to an Iatrogenic Pericardial Defect*

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A 14-year-old boy developed broad posterolateral myocardial infarction. During cardiac surgery at age five, a small pericardial window had been made. Autopsy revealed an extensive left-sided pericardial defect and necrosis of the left ventricular free wall, which had herniated and strangulated through the enlarged pericardial defect.

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The presence of the pericardium is not generally considered necessary to preserve life, but it does provide protection for the heart. In this article, we present the case of a 14-year-old boy who died of cardiac strangulation due to herniation of the left ventricle through an acquired pericardial defect which had been made 9 years before.

CASE REPORT

A five-year-old Japanese boy underwent an operation for patent ductus arteriosus. To prevent cardiac tamponade, a pericardial window, 10 × 15 mm in size, was made over the left ventricle.

At 14 years old, however, he suddenly developed chest pain. Acute heart failure occurred. The ECG showed marked ST elevation in leads 1, 2, aVL, and V₅-V₆ and ST depression in leads 3, aVF, and V₁-V₄ (Fig 1), which later changed to a poor R wave in leads 1, aVL, and V₅-V₆ and a high R wave in leads V₁ and V₄. The chest x-ray film demonstrated pulmonary congestion and a bulge of the left cardiac border, which disappeared later. The white blood cell count was markedly elevated with the peak value of 18,800/mm³ as well as serum creatine kinase level of 5,760 U/L, but viral titers were unremarkable. Echocardiography revealed severe dilatation (LV/Db: 1.8 mm) and akinesia of the left ventricular free wall. Thallium 201 myocardial perfusion scintigraphy showed a large defect in the posterolateral wall of the left ventricle. A diagnosis was made of acute posterolateral myocardial infarction; the patient died of intractable heart failure.

Autopsy revealed an extensive left-sided pericardial defect and myocardial necrosis of the left ventricular free wall (Fig 2). Post-mortem coronary angiography and pathologic examination showed no significant coronary stenosis. There was no inflammatory cell infiltration.

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DISCUSSION

This patient suffered from acute myocardial infarction as confirmed by the clinical and autopsy findings. However, the infarct zone was not explained by the coronary distribution but instead corresponded to a large pericardial defect. The extensive myocardial necrosis was not caused by either coronary occlusion or myocarditis. We concluded that cardiac herniation through the pericardial defect had produced myocardial infarction.

There are some patients with congenital pericardial defect who are discovered by chance and usually remain asymptomatic. It is quite common to resect a part of the pericardium and to leave it open during cardiac surgery. However, there have been seven cases of sudden death due to cardiac strangulation caused by herniation through a congenital pericardial partial defect.²³ Moreover, intrapericardial