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Two women with connective tissue disease developed a characteristic steroid-induced myopathy. Reduced maximal transrespiratory pressures indicated reduced respiratory muscle strength. Gradual steroid dosage tapering resulted in prompt clinical improvement and marked increases in respiratory muscle strength, maximal inspiratory pressure increasing by 33 percent in one patient and by 70 percent in the other. This reversible steroid-induced respiratory muscle weakness may be of great significance in reconsidering long-term steroid therapy in patients with underlying lung disease.

SYSTEMIC corticosteroid therapy is known to cause many unwanted side effects, among which muscular weakness and wasting have been observed since its earliest days.1,2 Experimental steroid myopathy* was shown to share many of the features of its clinical counterpart, which was extensively described by Affit et al.4 A more precise definition of the biochemical abnormalities of this myopathy, a better correlation between clinical and laboratory findings, and a useful approach for early detection, differential diagnosis, and management were provided by Askari et al.7

Steroid-induced myopathy in the respiratory muscles, to our knowledge, has never been reported. We were fortunate to make observations on respiratory muscle strength in two patients with steroid myopathy and complaints of dyspnea and on the changes caused by progressive tapering of the steroid therapy.

CASE REPORT

Case 1

In January 1978 this 58-year-old woman developed typical skin lesions of dermatomycosis and was followed up as an outpatient in the dermatology department. Clinical and biochemical investigation was negative, and spirometric test results were within normal limits.

Corticosteroid treatment (50 mg of prednisone daily) was started in March 1978 and over the following years continuously adjusted and tapered.

In March 1986, however, steadily worsening disease activity (reappearance of pathognomonic dermatologic features) necessitated an increase in prednisone dose, up to 60 mg daily. For the first time the patient developed rapidly progressing muscle weakness and was referred to us because of prominent dyspnea. Her nutritional status was normal. Muscle enzyme values remained normal, but 24-h creatine excretions were clearly elevated (Fig 1). Pulmonary function test results revealed both notably decreased maximal inspiratory pressure, Pimax, being 52 percent predicted and maximal expiratory pressure, Pmax, being 40 percent predicted. After gradual reduction of the prednisone dose to 7.5 mg daily, the creatinuria substantially decreased, and both Pmax and Pmax increased by 33 percent.

Case 2

In May 1986 the diagnosis of "unidentified connective tissue disease" was established in this 50-year-old woman with chronic pyelonephritis and secondary hypertension. Spirometric test results were normal. Prednisone treatment (50 mg daily for four weeks, gradually tapered to 30 mg) resulted in rapid regression of skin lesions and she was followed up as an outpatient in the division of nephrology.

After ten weeks of treatment, she complained of general fatigue, dyspnea on exertion, and muscle weakness. Serum enzyme values remained normal, but spirometric study results now showed clearly

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Corticosteroid-induced Myopathy (Janssens, Decramer)
reduced volumes (VC, 74 percent; FEV1, 70 percent). Physical disability considerably progressed with distinct facial mooning, severe girdle muscle wasting, osteoporosis, and lumbar vertebral collapse. The nutritional status, however, remained normal.

She was then seen in our outpatient clinic because of prominent dyspnea. Maximal transrespiratory pressures were clearly reduced, Pimax and PEmax being 35 percent and 41 percent predicted, respectively. The 24-h creatinuria was sixfold increased and showed a net decrease during prednisone dose reduction (Fig 1). This was associated with pronounced clinical improvement, normalization of spirometric values, and increased respiratory muscle strength, the Pimax increasing by 70 percent and PEmax remaining relatively constant.

**DISCUSSION**

This report provides, to the best of our knowledge, for the first time consistent evidence of reduced respiratory muscle strength caused by corticosteroid treatment.

Indeed, in both patients, diagnosis of steroid myopathy was firmly documented. First, in one patient, typical complaints of muscle weakness occurred concomitantly with the increased prednisone dosage. The other patient gradually developed steroid myopathy during seven months of high-dose prednisone treatment. In both patients, greatly increased creatinuria appeared in the presence of normal serum muscle enzyme levels. Second, underlying disease was in partial or complete remission, and especially in the second patient muscle weakness appeared simultaneously with other disabling nonmyopathic side effects of corticosteroid therapy. At the time of diagnosis, both patients were in a normal nutritional status and did not feel sick. Finally, in both patients muscle strength improved and creatinuria decreased after steroid dosage reduction, which confirms the diagnosis of steroid myopathy.7

These cases also clearly indicate that considerable reductions in respiratory muscle strength may be present with normal or only moderately reduced lung volumes on spirometric study.8 At the time of diagnosis in our patients, Pimax were only 52, respectively, and 35 percent predicted, and PEmax averaged about 40 percent predicted, indicating severe muscle weakness. The vital capacity, however, was normal in the first patient and slightly reduced in the second. After steroid dosage reduction, Pimax improved to 70 (59 percent predicted), whereas PEmax showed a 33 percent improvement in one patient and remained relatively unchanged in the second.

Our observations may have several important implications to clinical medicine. First, steroid treatment may contribute to worsening respiratory muscle function, already compromised in chronic obstructive pulmonary disease.8 Second, impaired respiratory muscle function induces a relatively ineffective cough, predisposing to respiratory tract infections, and promotes lung atelectasis consequent to the absence of deep breaths and sighs, which normally overcome closure of lung units.9 A critical reassessment of the effects of systemic corticosteroids on the respiratory muscles in all respiratory patients seem warranted.

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De novo Circumscribed Pulmonary Lobar Cystic Lymphatic Anomaly in a Young Boy*

A Possible Sequela of Bronchopulmonary Dysplasia

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This report describes a massive pulmonary lymphatic cystic anomaly affecting the right lower lobe of a nine-year-old boy. A year earlier, only an ill-defined small infiltrate could be seen in the affected lobe radiologically. The pathogenesis of this highly unusual lesion is discussed, taking into consideration the possible role of three months of mechanical ventilation in the neonatal period.

(Chest 1989; 95:1162-63)

Diffusely anomalous pulmonary lymphatic drainage in children is seen in the neonatal period. It is referred to as lymphangiectasis and tends to be uniformly fatal soon after birth. It may be isolated or associated with cardiovascular malformations and, in particular, an anomalous pulmonary venous return. Occasionally, it may be part of a generalized lymphatic dysplasia affecting many sites.1,3

Apparently primary but localized pulmonary lymphatic anomalies have been reported on rare occasions in adults.4,5 They have been called lymphangiomata or lymphangiectasis. Such lesions, to our knowledge, are exceptional in children,6,10 which makes the lesions unlikely to be included in the differential diagnosis of circumscribed cystic pulmonary lesions. This report proposes to document such a case and discuss its pathogenesis.

Case Report

A nine-year-old boy was admitted to St. Christopher's Hospital for Children with the chief complaint of fever and hemoptysis. Born 12 weeks prematurely, he had required mechanical ventilation for the first three months of life and had developed necrotizing enterocolitis, which was treated surgically. At eight years of age, he was incidentally found to have a small infiltrate in his right lower lobe, but was lost to follow-up until he developed low-grade fever and hemoptysis a year later. At this time, his PPD test and cultures of sputum were negative and his chest x-ray film and CT showed a round, well-defined, large right lower lobe mass (Fig 1). Contrast medium was administered during the study and failed to demonstrate any arteriovenous connection. A right lower lobectomy was performed.

Pathologic Study

At the posterior basal segment of the resected lobe, a spherical mass extended from the pleura into the pulmonary parenchyma (Fig 2). The visceral pleural surface covering the mass contained multiple dark red cysts measuring 0.2 to 0.5 cm in diameter. The cut surface of the mass showed it to be 4.5 cm in diameter, round and well demarcated. It had a spongy consistency and was made of numerous cystic spaces filled with blood-stained fluid.

Histologic examination revealed a network of channels, most of which centered around the bronchioles. Some were present in the septa and in the subpleural region. There was no capsule separating the lesion from the surrounding parenchyma. The channels varied in size, and their walls were of variable thickness. The inner surface of the channels was lined by flattened endothelial-looking cells (Fig 3). The walls were composed of connective tissue in which, in some areas, groups of smooth muscle fibers were present. Focal aggregates of lymphocytes could be seen in the walls of the channels and in the parenchyma adjacent to the lesion. Some channels were filled with red blood cells; others contained eosinophilic fluid. The surrounding pulmonary tissue was the site of a mild peribronchial lymphocytic infiltration. Acute intra-alveolar hemorrhage was prominent in a few areas.

Discussion

Diffuse or localized dysplastic development and increased intralymphatic pressure are essentially the two pathogenetic mechanisms traditionally invoked to explain the presence of lymphatic anomalies in children.

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Circumscribed Pulmonary Lobar Cystic Lymphatic Anomaly (Karmazin et al)