Spontaneous pneumomediastinum associated with laryngeal lesions and tracheal ulcer in dermatomyositis

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ABSTRACT

We described a 41-year-old woman with dermatomyositis, interstitial lung disease, and cutaneous vasculopathy who developed a pneumomediastinum. The routine bronchoscopy investigation found pale lesions in the larynx, that extended to the tracheobronchial tree, and deep ulcers in the membranous wall of the trachea. The histopathology examination revealed an inflammatory process that was diagnosed secondary to the vasculitis, but no infections. Superior and inferior airway lesions in the same patient with dermatomyositis is a very rare condition. The association of dermatomyositis with deep mucosal ulcers and pneumomediastinum is not clear, but a bronchoscopic examination should be used to improve evaluation.

Keywords: dermatomyositis, bronchoscopy, pneumomediastinum diagnosis.

INTRODUCTION

Dermatomyositis (DM) is a general inflammatory connective tissue disease of unknown cause involving mainly the muscles and the skin. Pulmonary complications are frequent and may lead to death.[1] The usual respiratory manifestations are interstitial pneumonitis, infection, dysfunction of the respiratory muscle, drug-induced disease, and pneumomediastinum (PM).

Spontaneous pneumomediastinum (SPM) has been described in DM as a rare complication, which carries a poor prognosis.[2] Several reports have described mortality rates ranging from 37.5%–52.5%, following the onset of PM. The precise mechanism has not been clarified, and it is believed that PM could be related to interstitial pneumonitis.[3,4] The rupture of the alveoli adjacent to vessels due to vasculitis may be a cause.[1,3,5] Kono et al.[6] first described bronchial necrosis as an airway manifestation in DM. They hypothesized that vasculopathy was a possible cause of necrosis and may occur after PM.

We report in this study an association between laryngeal lesions, tracheal and bronchial ulcers, and PM in a woman with DM.

CASE REPORT

A 41-year-old woman with DM and progressive interstitial lung disease with severe cutaneous activity, despite the use of corticosteroids and immunosuppressive agents, evolved with subcutaneous emphysema. The clinical examinations and laboratory evaluations showed the following: serum creatine kinase (CK) and aldolase at normal levels. The chest x-ray and computerized tomography revealed PM. Fiberoptic bronchoscopy was performed to evaluate the airways, and during the inspection was revealed: pale symmetrical lesions in the mucosa of the false vocal cords in the larynx, deep mucosal ulcerations in the posterior wall of the trachea ranging in size from 0.5–1.0 cm, and two flat, pale mucosal lesions surrounded by a hyperemic halo (may

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correspond to points of possible initial ulcer) situated in the membranous wall of the trachea and posterior wall of the main right bronchus with diffuse mucosal hyperemia of the tracheobronchial tree as well.

Biopsy of all visible lesions (more than one procedure) was performed with flexible biopsy forceps. The culture of the fragments was negative for fungus, mycobacterium, virus, and unspecified infection. The histological examination showed an unspecified inflammatory disease with predominant polymorphonuclear infiltrate.

The patient died due to sepsis that did not respond to antibiotic treatment.

DISCUSSION

Bradley[3] described the first case of PM associated with DM in 1986. In the English and French literature[4,7] 25 cases have been reported. Most of the patients had interstitial lung disease and cutaneous vasculitis (periungual infarct or ulcerous lesions). PM occurred during treatment with corticoids, and the patients had normal CK levels, like our patient. Kono et al.[6] reported on a patient with DM who had ulcerative skin lesions, white plaques on the bronchial mucosa at the carina and the main, lobar, and segmental bronchi in the lungs, immature lung disease, and PM. A biopsy showed subepithelial necrosis of the bronchial wall with epithelial squamatization, which are reminiscent of the macroscopic signals in our case.

The mechanism of SPM in DM has not yet been explained. It is known that the majority of patients reported on in the literature had interstitial lung disease and just a few patients had cutaneous vasculopathy solely. In almost half of the cases, CK levels were normal and most patients were treated with systemic corticotherapy. Kono et al.[6] assumed that necrosis of the bronchial wall attributable to vasculopathy could be the plausible mechanism.

This report provides a description of laryngeal lesions associated with tracheobronchial ulcers in DM, suggesting that manifestation of disease in the superior airway is possible. A differential diagnosis of infectious diseases is essential for establishing the appropriate treatment for immunocompromised patients, because diseases may differ even with similar clinical picture. In our case, the bronchoscopic analysis associated with the absence of specific signals in the histological examination suggested the hypothesis of vasculitis of the tracheobronchial mucosa as the cause of the ulcers, which could be a possible origin of the PM corresponding to the activity of interstitial disease.

CONCLUSION

The possibility of diffuse compromise of the airways caused by the pathological activity of DM must be considered. The relation between the airway lesion and PM in DM needs to be evaluated in further studies. A careful bronchoscopic examination is a powerful tool for the investigation of these patterns.

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