

Letters to the Editor

Lung Disease Secondary to Morphea *en Coup de Sabre*, a Form of Localized Scleroderma

Afectación pulmonar por esclerodermia localizada tipo morfea en coup de sabre

To the Editor:

Morphea *en coup de sabre* is a type of localized scleroderma characterized by patches of localized sclerotic skin over the face or scalp and accompanied by marked abnormalities of the tissues derived from the underlying mesenchyma. It has always been distinguished from diffuse scleroderma because there is no systemic component. We report the case of a man with morphea *en coup de sabre* with secondary dermal involvement and unilateral interstitial lung disease in the same location.

The patient was a 35-year-old man who had been admitted to hospital 5 years previously for hemiatrophy of the body (left side) and a cutaneous disorder. At that time, he reported only muscle weakness on the left side and had no respiratory symptoms. Physical examination revealed slight left-sided hemifacial and hemilingual muscle atrophy, hair loss on the left hemithorax and telangiectasias, a left frontal linear lesion that appeared to affect the skin and subcutaneous tissues, as well as extensive muscle atrophy predominantly in left lower and distal regions. The only significant findings of the laboratory tests were titers of 1/320 for antinuclear antibodies, 90 mg/dL for C3, and 15 mg/dL for C4. Tests for antibodies against extractable nuclear antigens, anti-Ro, anti-La, anti-Sm, Scl-70, anti-Jo1, anti-PM1, antineutrophil, antimitochondrial, and anticentromere cytoplasmic antibodies were negative, and kidney and liver function was normal. Additional tests (chest radiograph and lung function tests) were also normal. These findings led to a diagnosis of localized scleroderma, specifically morphea *en coup de sabre*. Eighteen months later, the patient was referred to the pulmonology department because of dyspnea on exertion and abnormalities detected by chest x-ray, consisting of loss of volume in the left hemithorax and ipsilateral interstitial involvement with a reticular pattern of septal thickening. High-resolution computed tomography (HRCT) confirmed the presence of an interstitial infiltrate in the left hemithorax (Figure A). The remainder of the parenchyma was normal. Lung function tests revealed mild to moderate restriction, with a forced vital capacity (FVC) of 3030 mL (68%), a forced expiratory volume in 1 second (FEV₁) in 1 second of 2550 mL (67%), a FEV₁/FVC ratio of 83%, and total lung capacity of 4190 mL (66%). The distance covered in the 6-minute walk test was 542 m, with a minimum oxygen saturation of 95%. After 1 year, the patient's breathlessness had increased. A chest x-ray showed a

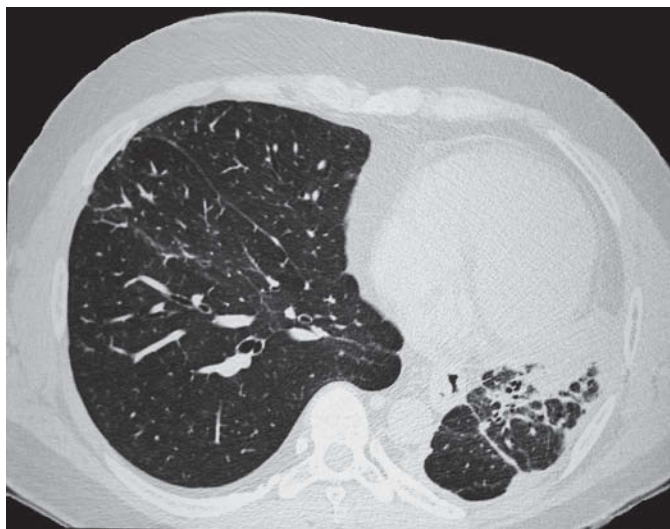
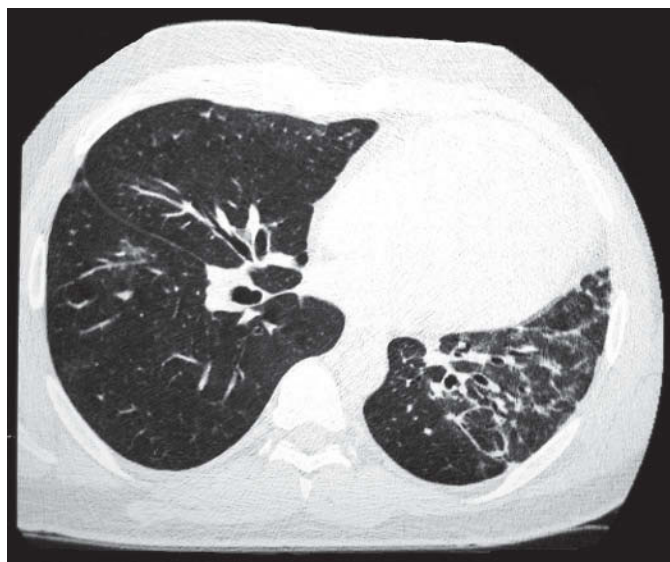


Figure. A. Mild interstitial disease in the left hemithorax. B. Progression of the disease after 1 year.

greater loss of volume in the left hemithorax, and HRCT revealed that the interstitial pattern had progressed significantly (Figure B). These findings were confirmed by the lung function tests: FVC of 2430 mL (55%), FEV₁ of 1800 mL (48%), FEV₁/FVC ratio of 74%, and total lung capacity of 3010 mL (48%). The distance covered in the

6-minute walk test was 500 m, with a minimum oxygen saturation of 87%. After confirmation of the progression of the lung disease, the dosage of corticosteroids was increased and azathioprine was added. The lung disease was subsequently seen to stabilize.

Scleroderma is a connective tissue disease of unknown origin and is classified as either systemic or localized, according to whether there is internal organ involvement or not.¹ Localized scleroderma is in turn separated into 4 variants: linear scleroderma, localized morphea, generalized morphea, and morphea *en coup de sabre*.² The latter is characterized by linear sclerotic lesions that affect one side of the body, occasionally including the face and the scalp. The skin and underlying tissues are involved. Although the distinction between systemic and localized scleroderma is restricted to the presence or absence of internal organ disease, in some cases organ involvement has been demonstrated in localized scleroderma, though patients were asymptomatic and involvement was mild.³ There was marked involvement in the case we report, however, and it progressed over time. The literature describes a few cases of extrapulmonary disease secondary to localized scleroderma,^{4,5} due to the involvement of muscles and subcutaneous tissue of the chest wall. However, after reviewing the literature, we conclude that this is the first case of interstitial lung involvement in localized scleroderma *en coup de sabre*.

The implication of this case is that we should perform series of additional tests on patients with localized scleroderma (whatever the

variant) in order to detect any internal organ disease that might be present—given that, despite normal results on the occasion of the first examination, our patient developed subsequent lung involvement. We should, therefore, be vigilant, taking a more aggressive approach to care to avoid complications like those described in the case we report.

References

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Comments on “Anesthesia in Thoracic Surgery in Catalonia: Results of a Survey Carried Out in 2003”

Comentarios a propósito del artículo “Actividad anestésica en cirugía torácica en Cataluña. Resultados de una encuesta realizada durante 2003”

To the Editor:

I read with interest the article by Vilà et al¹ entitled “Anesthesia in Thoracic Surgery in Catalonia: Results of a Survey Carried Out in 2003,” and I would like to make a few comments from the viewpoint of a thoracic surgeon.

The surgical volume in the aforementioned work is scant, with the result that it is difficult to draw conclusions. The caseload of the study comprises only 42 major pulmonary resections (lobectomies and pneumonectomies). These are the interventions that most accurately represent the volume and quality of a specific group of surgeons, and they have been analyzed in some detail in a benchmarking study carried out by 9 groups of thoracic surgeons.² A study published in 2006 by the Bronchogenic Carcinoma Cooperative Group of the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR)³ analyzed 2994 pulmonary resections performed in 19 thoracic surgery units. Morbidity, mortality, and survival were studied in terms of the number of interventions carried out annually by each unit. The units were divided into 3 groups according to their volume: low volume (≤ 40 cases per year), medium volume (between 41 and 54 cases per year), and high volume (≥ 55 cases per year). The authors concluded that the short-term and long-term results were not affected by the number of interventions performed. The findings were interpreted as the result of performing highly standardized procedures and based on very similar criteria in specialized

services. These data provide a certain guarantee as to the homogeneous quality of thoracic surgery teams in Spain.

Nevertheless, the beneficial effect of a large surgical volume both for a specific surgeon and for a group of surgeons has been recognized,⁴ as has the fact that the procedures are carried out by specialized surgeons.⁵ This has been a constant concern for those surgeons, like me, who are members of the National Committee of Thoracic Surgeons, and this concern has been voiced in a new forthcoming Training Program for Thoracic Surgeons that requires a medical resident to perform a considerably larger number of procedures while training.

One of the most serious problems facing thoracic surgery in Spain is the disperse nature of the work. This circumstance is favored by current legislation and by the decentralized way in which the country is governed. The work by Vilà et al¹ mentions 27 different centers for the procedures performed. Clearly, many of these centers are privately run, and we must not forget the volume of work carried out by thoracic surgeons on private patients or by general surgeons who also perform thoracic procedures, especially less complex ones. At present there are 10 thoracic surgery units or services in Catalonia,⁶ and 7 of these are accredited to provide training. These are the centers with the greatest and most complex experience in thoracic surgery performed in Catalonia. These data may dispel some of the doubts the authors raise in the discussion section of their study. Nevertheless, I feel that it would be wise to review the accreditation criteria for thoracic surgery units where residents are trained and to audit those that are currently accredited.

Finally, I think that studies analyzing surgical volume—such as that mentioned above—always generate better options for rational resource planning and enable us to optimize results. Therefore, they should be encouraged.