



## Letters to the Editor

### Primary pulmonary hypertension in Sjögren's syndrome: a rare association

#### *Hipertensión pulmonar primaria en síndrome de Sjögren: una asociación poco frecuente*

To the Editor:

Primary Sjögren's syndrome (SS) is rarely associated with pulmonary hypertension (PH). In connective tissue diseases, the development of PH is usually due to interstitial lung involvement.<sup>1</sup>

We present the case of a 40-year-old female who came to the Rheumatology department in 2002 with xerophthalmia of 5 years' evolution to which xerostomia and arthralgia had been added in the last 6 months, with no other extraglandular manifestations. Additional tests allowed us to diagnose SS: Positive ANA, positive anti-SSA, positive anti-SSB, abnormal Schirmer test, chronic inflammation in minor salivary gland biopsy information and compatible salivary gland scintigraphy. In September 2002, she had a routine echocardiogram, where data for slight PH (32 mm Hg) first appeared: that was later confirmed by right side heart catheterization carried out in December 2002. In November 2003, the patient presented dyspnea on exertion. The echocardiogram estimated a PH of 46 mm Hg. A thoracic CT eliminated pulmonary fibrosis and thromboembolic events. Currently, she has arthralgias that disappear with regular painkillers, with stable dyspnea.

Sjögren's syndrome is a common autoimmune disease, with glandular and extraglandular involvement. Pulmonary involvement is generally due to an interstitial pulmonary pathology and involvement of the lower airway.

Pulmonary hypertension in association with connective tissue disease occurs more frequently in systemic sclerosis (particularly in CREST syndrome), in lupus erythematosus and mixed connective tissue disease.<sup>1</sup> Association with SS is rare. According to published data, only 41 cases of SS with PH have been reported<sup>2</sup> (3 of them in Spain<sup>3-5</sup>) and only one SS paediatric case.<sup>6</sup> In a study published in 2009, 79 cases of arterial PH diagnosed from 1,892 patients with connective tissue disease were analysed. It was estimated that the PH prevalence was 4.2% in these patients and only 3.8% of these presented SS.<sup>7</sup> The specific mechanisms that cause PH are unknown, but some anomalies have been described that could participate in remodelling of pulmonary arteries and of small precapillary arterioles and in vasoconstriction of pulmonary arterioles. Histopathological similarities of idiopathic PH and that associated

to connective pathologies suggest common pathophysiological mechanisms. The anomalies (mainly described in idiopathic PH) lead to a reduction in vasodilator factors of endothelial origin (nitric oxide, prostacyclin), excessive production of growth factors (endothelin-1, vascular endothelial growth factor and platelet-derived growth factor angiopoietin-1), changes in potassium channels and increased serotonin transporter expression. More rarely, PH associated to connective pathologies can be due to a thromboembolic mechanism or a veno-occlusive disease, or to hypoxia caused by pulmonary fibrosis.<sup>8</sup> Some of these anomalies have led to new therapeutic targets.

We present a new case of this rare association between SS and PH, with the absence of interstitial lung disease without radiological evidence of thrombotic/thromboembolic events. The presence of dyspnea should alert the clinician to the possible existence of PH. The availability of efficient PH treatments justifies screening and the early diagnosis of PH in connective pathologies.

### References

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