Complications of Proximal Pulmonary Artery Aneurysms in Patients With Severe Pulmonary Arterial Hypertension

To the Editor,

Proximal pulmonary artery aneurysm (PAA) is defined as a dilation of the pulmonary artery trunk or its main branches. It is an infrequent disease of difficult diagnosis that is usually silent and is an autopsy finding. Occasionally, it causes severe complications such as airway compression, pulmonary artery (PA) dissection and intravascular thrombosis. Pulmonary hypertension (PH) is considered an important factor involved in its pathogenesis, and has mainly been described in patients with more severe PH such as is seen in the Eisenmenger syndrome. The incidence rate of these aneurysms in the population of patients with PH is low.

Of the 320 patients included in our pulmonary hypertension unit (mean PA pressure 56 mm Hg), we found 4 cases of proximal PAA diagnosed with chest CT after the appearance of complications. These 4 patients had severe PH (mean PA pressure 55 mmHg on catheterisation) of different aetiology (2 cases of idiopathic arterial PH and 2 cases of Eisenmenger syndrome). Before PAA diagnosis, the patients were in functional classes II-III of the WHO and in treatment with acenocoumarol and specific drugs for PH (2 patients with sildenafil and 2 patients with a combined therapy with sildenafil and subcutaneous teprostinil). Two of them came in to the emergency service with a persistent dry cough and one with compressive athelectasis. A chest CT showed an aneurysmatic dilation of the pulmonary trunk and its branches (PAA diameter 47-67 mm) with non-occlusive thrombosis in its interior and bronchial compression (Figure 1). Perfusion gamma scintigraphy ruled out perfusion defects indicating lung thromboembolism. Thrombi are relatively frequent in PAA and make it necessary to carry out a differential diagnosis with chronic thromboembolic PH. The hypercoagulability study was normal and other causes associated with PAA were ruled out. In all patients specific treatment for PH was intensified, compressive symptoms disappeared and anticoagulation was maintained due to the risk of thrombosis in the PAA region. The patients have been reviewed every 3 months without recurrence of clinical symptoms seen (follow-up: 2-3 years). In the follow-up CT carried out, the persistence of aneurysms without complications was confirmed. The fourth patient was admitted with chest pain; a chest X-ray showed dilation of the main PA (Figure 2); chest CT showed an aneurysm of the PA trunk and its main branches of 46.6 mm in diameter and a flap of the intima at the origin of the left PA. The lobular arteries arose out of the true artery lumen and were compressed by the false lumen of the aneurysm. No recent factors that could have precipitated dissection were found, except for the PH itself. The possibility of surgical treatment was ruled out due to the high risks of the intervention, as also was the possibility of percutaneous repair due to the risk of occlusion of the lobular arteries. Anticoagulant treatment was suspended and specific PH treatment intensified. In the follow-up CT, it was seen that the dissection had not progressed. The patient has worsened clinically and is currently on the waiting list for cardiopulmonary transplant.

In our experience, intensification of specific PH treatment improves airway compression, which was the most frequently seen complication in our patients. Maintenance of anticoagulation, although controversial, is usually necessary due to the risk of PAA thrombosis, although it may be necessary to suspend it in cases of haemoptysis, progressive

Figure 1. Chest computed tomography: aneurysmatic dilation of the trunk and main branches of the pulmonary artery.
Letters to the editor

We read with interest the article by León et al. who studied a sample of workers aged 25 to 64 years recruited during their routine medical check-up exams undertaken by the prevention services of FREMAP during 2003 and estimated an overall prevalence of the metabolic syndrome (MS) of 13.4%, higher in men (15.9%) than in women (5.2%). What really caught our attention, though, was that the greatest prevalence of MS in the various autonomous regions in Spain corresponds to men in Extremadura: 22.15% (95% confidence interval [CI], 21.7%-22.5%), whilst the prevalence in women is also the highest in Spain: 9.1% (95% CI, 8.1%-10.2%). This high prevalence of MS in Extremadura, by far the highest in Spain, is surprising. As the authors point out, the prevalence not only of MS, but also of other cardiovascular risk factors, like hypertension, diabetes and obesity, and even ischemic heart disease, is greater in regions of southern Spain, including Extremadura. Our group (GERIVA: Grupo de Estudio del Riesgo Vascular de Extremadura [Group for the Study of Vascular Risk in Extremadura]) recently published a study analyzing the prevalence of MS according to various different criteria in the province of Caceres (also in the region of Extremadura). The study involved the general population over the age of 24 years, and was based on a population sample of 1314 subjects (55.4% women), with a mean age of 52.8 (18) years. The prevalence of MS according to the ATP-III (2001) criteria, which we understand to be those used for the diagnosis of MS by León et al, was 18.6% (95% CI, 16.5-20.8), and was similar in men (18.3%; 95% CI, 15.2-21.4) and women (19%; 95% CI, 16.2-21.8). This prevalence is similar or just slightly higher than that of other similar studies carried out in Spain and with which we compared our findings. Thus, a prevalence, especially in men, of 22.15% would seem high, both in absolute terms and in comparison with the prevalence seen in the other autonomous regions.

BIBLIOGRAFÍA


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