An uncommon cause of dyspnoea

Une cause inhabituelle de dyspnée

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A 34-year-old woman presented to our hospital with a 9-month history of exertional dyspnoea and fatigue. She was a long-distance runner but over the preceding 9 months had developed progressive symptoms. She denied chest pain and had no previous cardiac history. Physical examination revealed normal heart sounds, absence of a bruit or murmur over the chest wall and no evidence of clubbing or cyanosis. Chest X-ray and electrocardiogram were normal. At rest oxygen saturation was 94% but decreased to 89% under stress. After an entirely normal surface echocardiogram and a reportedly normal transesophageal echocardiogram done elsewhere, we assessed for an intracardiac shunt using agitated saline contrast during transthoracic echocardiography. Agitated saline injected into the right antecubital vein revealed contrast in the right atrium within two to three cardiac cycles. After five to six cardiac cycles contrast appeared clearly in the left atrium via a pulmonary vein, suggesting the presence of an intrapulmonary shunt. Repeat agitated saline contrast with and without Valsalva confirmed an intact inter-atrial septum. The patient was referred for pulmonary angiography, which confirmed a complex arteriovenous (AV) malformation with a cluster of dominant fistulae arising from three subsegmental branches in the inferior lingular segment of right lower lobe (Fig. 1). Coil embolization was undertaken and a follow-up angiogram in this region showed successful occlusion of the feeding arteries (Fig. 2). Post-embolization at 3 months, the patient resumed normal exercise without recurrence of symptoms, using a home oximetry device to monitor oxygen saturation, which remained above 92% throughout exercise, and she resumed her hobby of long-distance running. At 3 years of follow-up, she continues to do well.
Isolated pulmonary AV malformations are uncommon. They may be seen in association with Osler–Weber–Rendu syndrome, where multiple mucosal AV malformations are present and clinical presentation is usually that of epistaxis. Diagnosis of an isolated AV malformation can be challenging, but should be suspected in cases of unexplained dyspnoea.

In all patients with exertional dyspnoea, after careful exclusion of the usual culprits such as chronic obstructive pulmonary disease, exercise-induced asthma, diastolic dysfunction and pulmonary embolism (acute or chronic), it is mandatory to perform a thorough contrast echocardiogram using an imaging window that clearly visualizes the interatrial septum in order to exclude intracardiac shunting. Presence of delayed agitated saline bubbles in the left heart is more consistent with an intrapulmonary shunt, as in the case presented. Knowledge of agitated saline bubble appearance within three cardiac cycles compared with after four cardiac cycles offers excellent discrimination in the diagnosis of an intracardiac versus an intrapulmonary shunt. It is a simple, easy, non-invasive inexpensive tool, which if used wisely clinches an important and reversible diagnosis.

Conflict of interest statement

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.acvd.2009.10.007.