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Cardiovascular involvement in systemic sclerosis: A case of atrial septal aneurysm associated with intracranial aneurysm

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Introduction

There is increasing concern about vascular and cardiac involvement in systemic sclerosis (SSc). This is a systemic infiltrative disorder that commonly affects the cardiovascular system and this case is presented to draw attention to the possibility of a pathophysiological connection between SSc and cardiovascular aneurysm formation. No data exist on the incidence or natural history of aneurysm in patients with SSc and to the best of our knowledge, there is no other report to date of atrial septal aneurysm associated with intracranial aneurysm in SSc.

The heart is one of the major organs involved in SSc (1,2), the involvement of which can be manifested by myocardial disease, conduction system abnormalities and arrhythmias, or pericardial disease.

Representative is the myocardial involvement by patchy fibrosis (secondary to both repeated ischemia and immune-inflammatory damage) that leads to ventricular diastolic dysfunction (3,4), whereas right ventricle overload and failure may complicate pulmonary hypertension.

SSc is frequently linked to vascular and/or parenchymal lung disease (5), determining symptom occurrence, particularly dyspnoea and fatigue. Several vasodilator approaches (prostacycline or NO/endothelin) may counteract the microvascular dysfunction at peripheral and cardiopulmonary level and fight

the sequelae of pulmonary hypertension (5) which is secondary to damage in the lung.

There has been an increased awareness of left ventricular abnormalities in SSc patients, but the occurrence of cardiac aneurysms seems to be a rare complication of SSc, only few cases are illustrated in the literature (6).

Vascular abnormalities such as fingertip ulcers and Raynaud's syndrome as well as involvement of other organs including kidney and the gastrointestinal tract, are prominent features of the disease. Macrovascular involvement in SSc has received relatively little attention, the prevalence of peripheral large vessel disease is increased in SSc, on the other hand, the association with vascular aneurysm is poorly understood, few reports are sporadically described (7-9).

Case report

We hereby describe an atrial septal aneurysm associated with an intracranial aneurysm in a 56-year-old woman presenting with increased esophageal motility disorders and rapidly progressive diffuse cutaneous scleroderma with induration of all extremities and Raynaud's phenomenon.

During routine TTE, an hypertrophic cardiomyopathy and an asymptomatic atrial septal aneurysm (ASA) without shunting at

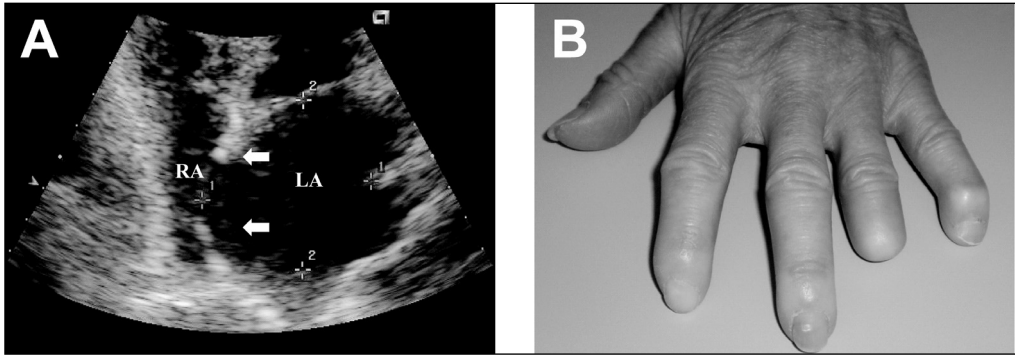


Figure 1

two-dimensional color-Doppler echocardiography, were detected.

The ASA was protruding far away into the right atrium with oscillation, and maximum displacement of ASA was evaluated between 10 and 12 mm (Fig A). Initial pulmonary artery hypertension at 45 mmHg, although a treatment with Bosentan since two years for a severe form of Raynaud's phenomenon (Fig. B), was measured. Serum BNP concentration was moderately increased (274 ng/L) indicating an overload of the right and/or left ventricle. An incidentally found irregular intracranial aneurysm (10 x 6 mm; not shown) during a magnetic resonance imaging performed for aspecific malaise/migraine confirms the possible mechanism whereby systemic sclerosis may result in aneurysm formation.

Conclusion

There is evidence of generalized endothelial inflammation in patients with SSc (10), and this is thought to be a precondition for aneurysm formation (11); endothelial injury to vessels, followed by platelet adhesion and leukocyte activation, plays a primary role in aneurysm formation. These events cause complementary activation and release of prostaglandins and lysosomal enzymes, with additional mechanical disruption of the vessel wall. Hemodynamic stress at the sites of vessel disruption may result in focal wall and

aneurysm formation. Inflammatory reactions and endothelial damage seem to be common features in the formation and growth of intracranial aneurysms, too.

SSc is also characterized by inflammation and fibrosis of many organs. As previously described, the underlying mechanism appears related to microcirculatory impairment causing focal ischemic injury and irreversible fibrosis (12,13). The histological alterations are primarily found on the intima of the vessel wall, increasing its collagen content (14). Immune complex deposition in the disease is extensive and leads to vascular damage (15). Most of the effect is on small vessels and capillaries.

Although the occurrence of atrial septal aneurysm associated with an intracranial aneurysm as in this case, may be pure coincidence, an inflammatory process could be suspected to be a cause for vascular aneurysm formation and the same process could be postulated for other cardiovascular structures leading to aneurysm formation.

A better knowledge and awareness of cardiovascular involvement is necessary because it conveys a major risk for mortality; a meticulous search for aneurysms seems to be important in patients with SSc. Vascular and cardiac involvement in SSc is often manifest, and nearly always present when accurately searched.

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