foramen ovale directly made the diagnosis of paradoxical embolism. Patent foramen ovale is just an innocent passive conduit in absence of the co-existence of a venous thrombus. However, it exposes to a risk of paradoxical embolism during the whole life.³

The overall early mortality of the reported cases is high. However, therapeutic implications of such entrapped embolus through a patent foramen ovale remain controversial. Thrombolysis has already been attempted with variable results as well as cardiac surgery.⁶ Nevertheless, when surgical removal seems too hazardous, as in elderly patients, heparin treatment with echocardiographic monitoring has been suggested.⁷

The symptoms of peripheral embolism are often so minor and non specific (transient abdominal discomfort, leg cramp) that the diagnosis is often missed. Only about 2% of the patients with cardiogenic brain embolism have clinically recognized peripheral emboli.⁸ A thorough vascular examination is needed before stating that the thrombus resolved without sequellae.

In conclusion, the present case shows that paradoxical embolism may be underestimated, and illustrates that it may be the first presenting sign of malignancy. The presence of a hypercoagulable disorder associated or not with malignancy should be searched in every case of paradoxical embolism.⁹

References

KEYWORDS
Apical hypertrophic cardiomyopathy;
Atrial septal defect;
Renal failure;
Hypothyroidism;
Primary amenorrhea

Abstract
Aim: We describe a case of non-obstructive apical hypertrophic cardiomyopathy with atrial septal defect, in a 48-year-old caucasian female patient with chronic renal failure, hypothyroidism and primary amenorrhea, referred to our hospital for syncope, palpitation and shortness of breath.

Methods and results: Electrocardiogram, transthoracic echocardiogram and cardiac magnetic resonance showed classical features of apical hypertrophic cardiomyopathy. Apical hypertrophic cardiomyopathy is morphologically characterized by apical ventricular hypertrophy, and is reported to be a relatively benign prognosis compared with the other type of hypertrophic cardiomyopathy.

Conclusion: Apical hypertrophic cardiomyopathy is very rare in the West, is occasionally encountered in Japanese persons, but there have been only a few reports of its coexistence with atrial septal defect. Our present report is the first case of apical hypertrophic cardiomyopathy with atrial septal defect associated with renal failure, hypothyroidism and primary amenorrhea that could represent a multi-organ syndrome. This hypothesis was supported by the finding of the same characteristics in a sister of the patient.

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Case report

We describe a case of a 48-year-old caucasian woman, who was referred to our hospital for the evaluation of syncope, palpitations and exertional dyspnea. The patient was affected by chronic renal failure in therapy with ramipril 2.5 mg/day from around 10 years, primary amenorrhea and hypothyroidism in treatment with hormonal therapy. The family history reported arterial hypertension, coronary artery disease and sudden death. The clinical examination showed rhythmic heart sound with mild systolic murmur. Vesicular murmur was preserved and blood pressure was 120/70 mm Hg.

Electrocardiogram showed sinus rhythm, left ventricular hypertrophy with negative T-wave in leads I, aVL and V2 to V6 (Fig. 1). The transthoracic echocardiography provided the following important informations: dilated left atrial dimension (62 mm) with atrial septal aneurysm and atrial septal defect (ASD), a marked hypertrophy of distal segment of septum and apex (25 mm), normal left ventricular ejection fraction (65%), moderate mitral regurgitation (Fig. 2). By transesophageal echocardiography, a secundum-type ASD was clearly observed and Doppler color image showed abnormal flow from the left to right atrium through the intra-atrial septum. Holter 24-h electrocardiographic monitoring revealed ventricular extrasystolic (876/day) with multiple non-sustained ventricular tachycardia. The patient underwent a cardiac magnetic resonance imaging which confirm the diagnosis of apical hypertrophic cardiomyopathy (HCM); gadolinium delayed imaging revealed contrast hyperenhancement corresponding to apical left ventricle on both long- and short-axis images. The patient was treated with acetylsalicylic acid 100 mg/day, metoprolol 50 mg twice a day, ramipril 2.5 mg/day. An electrophysiologic study reproducibly induced polymorphic ventricular tachycardia, so a cardioverter defibrillator was implanted. Afterwards, her symptoms were alleviated.

Apical HCM is a hereditary myocardial disorder, caused by mutations of sarcomeric proteins. Apical hypertrophy of myocardium predominantly involves the apex of the left ventricle. Apical HCM has favorable clinical outcome. However, severe clinical manifestations, including sudden cardiac death, severe arrhythmias and apical infarction can be seen. Diagnosis of apical HCM is based on electrocardiographic (giant negative T-waves in the precordial leads), echocardiographic (hypertrophy and no intraventricular pressure gradient) and angio graphic (spade-like configuration of the left ventricle on contrast angiography) findings. As many as 25% of Japanese patients with HCM have predominantly apical involvement. Apical HCM occurs in only 1–2% of the non-Japanese population. Apical HCM is now a well-known myocardial disease, but the additional coexistence of an ASD, renal failure, primary amenorrhea and hypothyroidism is quite rare; we supposed that this combination could represent a multi-organ syndrome. This hypothesis was supported by the finding of the same characteristics in...
a sister of the patient who had remarkably similar physical examination, renal failure in therapy with ramipril, primary amenorrhea and hypothyroidism in treatment with hormonal therapy,

Figure 1  Electrocardiogram at rest demonstrates the sinus rhythm, left ventricular hypertrophy with negative T-waves in leads I, aVL and V2 to V6.

Figure 2  The echocardiographic four-chamber view showing apical hypertrophy of distal segment of septum and apex (25 mm) and atrial septal aneurysm. There is no obstruction of the left ventricular outflow tract. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.
electrocardiographic findings and ASD but not yet apical left ventricular hypertrophy. Besides, we supposed that in our patient, the prolonged therapy with ACE-inhibitor could have delayed the symptoms and cardiac concentric remodelling.

References


Mobile right heart thrombus and massive pulmonary embolism

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**KEYWORDS**
Right heart thrombus; Intracardiac mass; Echocardiography; Pulmonary embolism

**Abstract**
The current report describes a patient with pulmonary embolism, treated unsuccessfully with heparin. Transthoracic echocardiography revealed free-floating right heart thrombus. Migrating deep vein thrombus to the right heart was suspected. Transesophageal echocardiography confirmed origin of the thrombus in the inferior cava vein. Mortality rate of mobile right heart thrombus is over 40%, therefore urgent surgical embolectomy was performed with relief of symptoms.

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