A Case Showing a Rare Evolution from Hypertrophic Obstructive Cardiomyopathy to “Dilated” Cardiomyopathy Demonstrated by Echocardiography

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SUMMARY

A 20-year-old woman whose echocardiograms showed a rare evolution from hypertrophic cardiomyopathy (initially with obstruction and 4 years later without obstruction) to dilated cardiomyopathy over an 8-year observation period is described.

Additional Indexing Words:
Hypertrophic obstructive cardiomyopathy Hypertrophic nonobstructive cardiomyopathy Dilated cardiomyopathy Evolution of the left ventricle Echocardiogram

HYPERTROPHIC cardiomyopathy and dilated cardiomyopathy are believed to be different entities and the evolution from a hypertrophic to a dilated left ventricle even in the “endstage” of hypertrophic cardiomyopathy (HCM) has been reported to be extremely rare.1)-4) We report a case whose echocardiogram revealed a rare evolution from hypertrophic obstructive cardiomyopathy (HOCM) to “dilated” cardiomyopathy (“D” CM) over an 8-year interval.

Case Report

A 20-year-old woman initially noted palpitations 8 years ago (February, 1974) when she was diagnosed as having HOCM at a nearby hospital. At 18 years of age she started to be troubled by palpitations starting from 16 weeks of pregnancy but she refused to have an induced abortion. Prior to this admission she had 2 pregnancies in which a caesarean section was necessary in one; the other terminated in a spontaneous abortion. At the age of 20 she became pregnant again and was admitted to this hospital for the third
time at 30 weeks of pregnancy (July 2nd, 1982). At admission her blood pressure was 122/60 mmHg and the pulse was 56. Moist rales were audible in both lower lung fields posteriorly. Peripheral edema (+) was noted.

A chest X-ray revealed moderate cardiomegaly with a cardiothoracic ratio of 0.66 and congestion of the pulmonary vasculature which was compatible with “D” CM (Fig. 1-B). A chest X-ray taken at the age of 13 years showed a prominent left ventricular border with a cardiothoracic ratio of 0.52 (Fig. 1-A).

The electrocardiogram (Fig. 2-B) showed a sinus rhythm, a PQ interval of 0.21 sec, left atrial overloading, intraventricular conduction disturbance and inverted T waves in leads I-III, aVF and V₄₋₆.
An electrocardiogram (Fig. 2-A) recorded on February 12th, 1974, disclosed a regular sinus rhythm at a rate of 60/min, prominent tall R waves in $V_6$ (5.6 mV) and deep S waves in $V_1$ (4.2 mV) with ST-T changes indicating left ventricular hypertrophy and strain.

Echocardiographic findings included thinning of both the interventricular septum (IVS=10 mm) and posterior wall of the left ventricle (LVPW=15 mm) with hypokinesis and dilatation of the cavity (LV diastolic dimension: LVDd=65 mm, EF=0.21) suggesting "D" CM (Fig. 3-D).

An echocardiogram recorded on February, 1974, showed markedly thickened IVS and thickened LVPW, asymmetric septal hypertrophy (ASH), systolic anterior movement (SAM) of the mitral leaflet and a small left ventricular cavity indicating HOCM (Fig. 3-A). These findings were supported by a systolic ejection murmur and left ventriculogram (Fig. 4).

On June 9th, 1980, the echocardiogram revealed the same degree of increased thickness of both IVS (27 mm) and LVPW (23 mm) with EF of 0.66 and disappearance of the SAM and a narrow cavity (LVDd=40 mm) (Fig. 3-B). On March 11th, 1982, the echocardiogram showed a moderately thickened IVS (20 mm) and LVPW (21 mm) and a moderately dilated cavity (LVDd=53 mm, EF=0.40) compared to the last examination (Fig. 3-C).

She delivered a baby boy on July 15th, 1982, at 32 weeks of pregnancy. After that, although the number of premature contractions decreased, congestive heart failure worsened markedly and she died 1 month after delivery following the sudden onset of ventricular tachycardia and ventricular fibrillation. A chest X-ray taken one day before her death revealed marked cardiomegaly with a cardiothoracic ratio of 0.76, prominent pulmonary vasculature and pleural effusion (Fig. 1-C). Necropsy was not done because of
Fig. 3. M-mode echocardiograms. A: Recorded on February 12, 1974, illustrating asymmetric septal hypertrophy (ASH), systolic anterior movement (SAM) of the mitral leaflet and a small left ventricular cavity (LVC). B: Recorded on June 9, 1980, showing prominent hypertrophy with disappearance of ASH and SAM. C: Recorded on March 11, 1982, revealing thinner ventricular wall and greater LVC compared with the former examinations. D: Four months later, disclosing a significantly thinner wall and dilated LVC with poor contraction.

refusal by her family. Past and family history had not been clarified because she had been a daughter-in-law.

DISCUSSION

Maron et al\(^2\) initially reported the existence of a minority of patients with HCM who are characterized by supraventricular arrhythmias and severe congestive heart failure. Fujiwara et al\(^3\) presented the case of a patient who had progressed from HOCM to “D”CM-like features in the end stage, documented by echocardiography and at necropsy.
In our case, serial echocardiograms showed an interesting chronological evolution: the initial HOCM pattern changing into HCM with the disappearance of ASH and SAM 5 years later; a thinning left ventricular wall, although still thicker than normal, and dilatation of the left ventricle 2 years later; and finally typical features of "D"CM in the end stage.

This rare evolution has been ascribed to (1) transmural myocardial infarction due to narrowing of the small intramural coronary arteries, (2) spasm of major and/or intramural coronary arteries, (3) thromboembolism in the major coronary arteries, (4) relative ischemia due to ventricular hypertrophy, outflow tract obstruction and arrhythmias, (5) disarray, and (6) complicating myocarditis.2)–4)

Although we did not perform invasive studies because the admission of our patient had always been connected with pregnancy and autopsy was refused, a definite myocardial infarction can be excluded from the clinical standpoint. Therefore, it seems plausible that myocardial ischemia secondary to the above-mentioned mechanisms other than myocardial infarction could bring about massive transmural fibrosis, leading to the thinning and dilatation of the left ventricle. As she had experienced three pregnancies, the possibility of peripartum or postpartum cardiomyopathy should also be considered. Peripartum and postpartum cardiomyopathy are widely known clinical syndromes of myocardial disease of unknown etiology that present toward the end of pregnancy or in the early puerperium.5)–6)

Diagnostic criteria proposed by Walsh et al.6) consist of; (1) absence of the history, symptoms and physical findings of heart disease prior to the puerperium; (2) appearance of signs and symptoms of heart disease between the
first and 20th week of the puerperium; and (3) inability to establish an etiologic basis for the heart disease.

Several speculations have been made as to the relationship between this disorder and pregnancy; (1) a form of cardiomyopathy associated with pregnancy; (2) some type of postinfectious myocarditis developing during the gravid or postgravid period; (3) some relation to toxemia, malnutrition, prolonged delivery, shock or bleeding.

Although our patient showed rapid deterioration of her cardiac status during and after each pregnancy, she was not diagnosed as having postpartum cardiomyopathy because she was known to have HOCM diagnosed at age 13. Moreover, her pregnancies were not considered to have been complicated by toxemia, prolonged delivery or viral myocarditis. However, the possibility cannot be denied that unknown factors resulting in postpartum cardiomyopathy might have been responsible for the evolution from HCM to "D"CM in the present case.

According to Walsh et al,6 subsequent pregnancies and deliveries aggravate postpartum cardiomyopathy and 2 of their 6 patients who developed congestive heart failure following delivery were reported to exhibit overt congestive heart failure in spite of digitalis.

The present case also showed rapid progression from HCM to "D"CM-like features shortly after her second delivery and then died suddenly.

As necropsy was not allowed for this patient, the final "D"CM-like features were not clarified from the pathological point of view. However, the slow progression from HOCM to HCM in 8 years and then the rapid progression from HCM to "D"CM is quite impressive.

REFERENCES