Short Communications

Reduced Left Ventricular Dimension and Normalized Atrial Natriuretic Hormone Level after Repair of Aortic Coarctation in an Adult

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Summary

Although unusual in the older patient, coarctation of the aorta can be an occult cause of cardiomyopathy. This report describes a 53-year-old man with new-onset heart failure symptoms, global left ventricular (LV) dysfunction, and underlying aortic coarctation. Surgical correction resulted in reduced LV size, resolution of symptoms, and normalization of atrial natriuretic hormone levels.

Key words: aortic coarctation, atrial natriuretic hormone, congestive heart failure, neurohumoral activation

Introduction

In patients referred for evaluation of dilated heart failure, it is important to exclude reversible causes. This includes consideration of mechanical causes, such as occult aortic stenosis, that may produce a picture resembling idiopathic dilated cardiomyopathy but for which surgical correction is possible.¹ Similarly, increased afterload in the setting of coarctation of the aorta may produce heart failure.² As the fifth most common congenital heart lesion,³ aortic coarctation accounts for 5 to 8% of all congenital cardiac anomalies.⁴ Although it is occasionally diagnosed in patients over 50 years of age,⁵ little is

known about cardiac function following surgical repair at this age. We report an adult presenting in the fifth decade with new-onset dilated cardiomyopathy secondary to occult, congenital coarctation of the aorta. Following surgical coarctectomy, the patient had reduction of left-sided chamber dimensions and improvement of neurohumoral activation, as reflected in atrial natriuretic hormone levels.

Case Report

A 53-year-old man was evaluated for new-onset nocturnal dyspnea, orthopnea, and dependent edema. His medical history was notable only for gout. He was normotensive, took no regular medications, and had received regular annual medical care. He worked as the manager of a golf course, did not smoke, and denied alcohol or drug use and toxin exposures. Review of systems was negative for claudication.

Physical examination revealed blood pressure of 130/70 mmHg in both arms. The pulse was 94 beats/min and he was afebrile. Jugular venous pressure was 9 cm of H₂O. The cardiac impulse was diffuse and laterally displaced. There was a left ventricular (LV) heave, a prominent third heart sound, and an ejection click. A harsh II/VI systolic murmur was audible along the left scapular border. Bibasilar rales, mild hepatojugular reflux, and 2+ pedal edema were present. Femoral and pedal pulses were barely palpable and the lower extremity systolic pressure was 70 mmHg bilaterally.

Laboratory values were notable for creatinine 1.5 mg/dl, total bilirubin 1.8 mg/dl, aspartate aminotransferase 143 IU/l, alanine aminotransferase 341 IU/l, and International Normalized Ratio 1.7. Echocardiogram revealed four-chamber dilatation, severe systolic dysfunction, and a bicuspid aortic valve. Neither aortic stenosis nor regurgitation was present. Left ventricular wall thickness was within normal limits. Chest x-ray showed cardiomegaly, prominent pulmonary vasculature, and notching of the ribs bilaterally (Fig. 1). The electrocardiogram revealed sinus rhythm, first-degree atrioventricular block, left atrial enlargement, and nonspecific intraventricular conduction delay.

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FIG. 1 Presenting chest x-ray. The exposure shown was chosen to accentuate the rib shadows. Rib notching is present (arrows) and results from collateralization and enlargement of the intercostal arteries.

Cardiac catheterization demonstrated juxtaductal coarctation of the aorta. The gradient across the coarctation was 50 mmHg and coronary angiography showed no critical coronary artery disease. Aortography localized the narrowing to beyond the takeoff of the left subclavian artery. Extensive intercostal, suprascapular, and internal mammary arterial collaterals were present. Endomyocardial biopsy revealed moderate cellular hypertrophy and no myocarditis. The patient did not tolerate captopril secondary to hyperkalemia.

Surgical coarctectomy with end-to-end anastomosis was performed semielectively. The patient had little postoperative hypertension.⁶ He had resolution of all physical findings of heart failure, except for the laterally displaced impulse, and he became symptom-free. All laboratory abnormalities of liver and renal function resolved. Follow-up echocardiography was performed at 2 months and showed reduction in LV and left atrial dimensions (Table I). Fractional shortening, however, remained 12–13% (cf. Fig. 2A and B). Circulating levels of atrial natriuretic hormone were followed as a measure of the neurohumoral activation associated with heart failure.⁷

TABLE I Left-sided chamber dimensions by echocardiography

	Pre-operative	Post-operative	Reference range
Left ventricular diastolic dimension	6.8	6.1	(3.3–5.8) cm
Left ventricular systolic dimension	6.0	5.3	(2.3–3.9) cm
Left atrial dimension	4.6	4.0	(1.5–4.0) cm

The postoperative echocardiogram was performed 2 months after surgical coarctectomy.

Atrial natriuretic hormone levels were grossly elevated preoperatively, but normalized after surgery (Table II). Levels of epinephrine, renin, and aldosterone were near normal values preoperatively and changed little after surgery. Angiotensin-converting enzyme and norepinephrine levels became mildly elevated postoperatively (Table II).

Discussion

Classically appearing dilated cardiomyopathy has been shown to occur with occult aortic stenosis.¹ Here we report a 53-year-old man with outflow obstruction secondary to congenital aortic coarctation. In addition to the signs and symptoms of dilated cardiomyopathy, this patient had disparate blood pressure in the upper versus lower extremities, notching of the ribs on chest x-ray, a harsh systolic murmur along the left scapula, and a bicuspid aortic valve. Following surgical correction of the coarctation, his heart failure symptoms resolved and echocardiography revealed reduced chamber dimensions. Plasma levels of atrial natriuretic hormone normalized from the grossly elevated preoperative levels, suggesting neurohumoral improvement as well.

Secondary hypertension and abnormalities of the aortic valve are commonly associated with coarctation of the aorta.

FIG. 2 Representative M-mode echocardiography. (A) The preoperative study. (B) Postoperative study performed at 2 months. Left ventricular diastolic dimensions are shown (arrows).

TABLE II Circulating neurohormone levels

	Pre-operative	Post-operative	Reference range
Atrial natriuretic hormone (pg/ml)	180	<3	(0–46)
Epinephrine (pg/ml)	84	82	(<60)
Renin (ng/ml/h)	4.6	4.4	(1.3–4.0)
Aldosterone (ng/dl)	6.5	11.0	(7.5–22.7)
Angiotensin-converting enzyme (U/l)	29	58	(8–52)
Norepinephrine (pg/ml)	549	742	(120–680)

The postoperative studies were performed at 3½ months. Values shown are plasma levels, except for aldosterone and angiotensin-converting enzyme which were measured on serum.

Bicuspid aortic valve occurs in 30 to 80% of cases.^{8–10} Other associations include increased risk of premature coronary artery disease, cerebral aneurysm, stroke, aortic rupture, and bacterial endocarditis.^{4, 10} In the evaluation of this patient, the bicuspid aortic valve detected by echocardiography provided a distinct clue. Coarctation of the aorta can be expected in 6 of 100 patients with a congenitally bicuspid aortic valve,¹¹ whereas dilated cardiomyopathy from an unrelated cause would be expected to occur at a frequency of only 1 in 10,000.¹²

The absence of hypertension and lower extremity claudication in this patient's history was also notable. Glancy *et al.*¹³ reported normal upper extremity blood pressure in up to 40% of patients with aortic coarctation. This is best explained by the extensive collateralization as well as by depressed systolic function that occur with time. In addition, Fixler¹⁴ reported that lower extremity claudication is actually uncommon in older patients with coarctation, again presumably due to long-standing, extensive collaterals.

Following surgical coarctectomy in the present case, atrial natriuretic hormone levels were the most dramatically improved of the neurohormones measured. This may be because the predominant postoperative effect was a reduction in chamber dimensions. Decreased chamber dimensions would be expected to reduce stimulation of the cardiac stretch receptors that are responsible for atrial natriuretic hormone release.⁷ In contrast, fractional shortening remained impaired postoperatively. This is consistent with results in canine surgical models of coarctation¹⁵ and may explain why levels of epinephrine and norepinephrine remained mildly elevated postoperatively.

Conclusion

Identification of reversible causes is important in evaluating new-onset cardiomyopathy. Outflow tract obstruction secondary to aortic coarctation should be considered as a potential occult cause of cardiomyopathy. In the present case, surgical correction resulted in improved LV dimension, improved left atrial dimension, and resolution of the elevated levels of atrial natriuretic hormone.

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