Communications

Systolic Click Syndrome Without a Click: Hints on Diagnosis

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Barlow et al[1-4] in the mid-1960s described billowing of mitral valve leaflets in the mid-systolic click – late systolic murmur syndrome. Shortly thereafter Read et al[5] characterized myxomatous transformation of the floppy mitral valve leaflets as a possible forme fruste of Marfan's syndrome. Since that time it has been found to be a common disorder, sometimes with no clear click or murmur.[6]

How, then, can the family physician make the diagnosis? He sees many anxious females with complaints of atypical chest pain, palpitations and tachycardia, whose physical examination, chest x-ray, SMA-12, urinalysis, and electrocardiogram fall within normal limits.

Recently, Bon Tempo et al[7] and Salomon et al[8] have shown a way out of this dilemma. They demonstrate thoracic cage anomalies in the majority of patients with prolapsing mitral valve leaflets even in the absence of clicks or murmurs by phonography. The most common abnormalities seen are pectus excavatum, "straight back" (loss of normal thoracic kyphosis), and scoliosis.[8] Seventy-five percent of patients with floppy mitral valve leaflets are found to have a smaller than average anteroposterior to transverse thoracic ratio.[7] Therefore, the family physician would do well to consider echocardiography when such thoracic abnormalities are observed.

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Case Report

A 34-year-old, white woman presented with an eight-year history of tachycardia and palpitations associated with shortness of breath and intermittent chest pain which began during her last pregnancy. The pain was substernal without radiation, squeezing in quality, aggravated by physical activity and certain movements, such as bending over, and relieved by rest and reduction of physical activity. She had repeatedly sought medical advice but no diagnosis had been made and she had been treated primarily with diazepam over the years. The patient denied syncope, hemoptysis, edema, rheumatic heart disease, nausea, and vomiting. Past physical examinations, electrocardiograms, routine chemistry and thyroid tests were within normal limits. Past medical history was noncontributory except for a mild iron deficiency anemia and the fact that she had been frequently sick as a child.

On physical examination the temperature was 36.8 C, the pulse rate 100 beats per minute and the respirations 12 per minute. The blood pressure was 110/80 mm Hg without paradox.

On examination she was a slender but well-developed and well-nourished woman who was somewhat teary and agitated. The skin was without lesions. Head, eyes, ears, nose, and throat were unremarkable. The neck was supple without thyromegaly. Chest examination disclosed a straight back with flattening of the normal dorsal kyphosis. The lungs were clear. The heart was not enlarged; there was a regular tachycardic rhythm without click or murmur. The second sound was split physiologically. An S-4 was present. Peripheral pulses were full. The abdomen was soft without masses or organomegaly. The extremities showed no clubbing or edema; the metacarpophalangeal joints were somewhat hyperextensible. The pelvic examination was normal. The neurological examination was unremarkable except for mild anxiety.

The urine was normal. The hematocrit reading was 35.2 percent; the white blood cell count was 5,700/cu mm, with 57 percent neutrophils, 39 percent lymphocytes and 4 percent eosinophils. The SMA-12 was unremarkable.

The electrocardiogram showed a tachycardia of 110 to 120 beats per minute and a slight voltage irregularity. No murmurs were demonstrated by phonocardiography. The echocardiogram revealed mild prolapse of the posterior mitral leaflet and redundant but nonprolapsing anterior mitral leaflet.

The patient was started on propanolol hydrochloride (Inderal), 10 mg, three times a day, and has done well.

Because thoracic cage abnormalities raised the question of Barlow's syndrome to the family physician, the patient is no longer denied an explanation, evaluation and specific treatment for her condition, to say nothing of prophylaxis against subacute bacterial endocarditis.

References

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