Anomalous mitral arcade is a rare congenital malformation and few survive to adulthood. A 65-year-old man presented with mild dyspnea and cardiac murmur. Transthoracic and transesophageal echocardiography showed an arc-like configuration of the papillary muscles, constrained by an interconnecting fibrous band in direct continuity with the anterior mitral leaflet, and mitral insufficiency caused by the restricted mobility of the mitral valve. Multidetector computed tomography confirmed these findings. (Circ J 2005; 69: 1560–1563)

**Key Words:** Computed tomography; Congenital heart disease; Echocardiography; Elderly; Mitral insufficiency

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**Case Report**

A 65-year-old man was referred to hospital for the evaluation of a cardiac murmur and solitary pulmonary nodule. He had a history of mild dyspnea for 2 years that was tolerateable without medication, and diabetes mellitus that was controlled with oral hypoglycemic agents. Physical examination revealed a heart rate of 75 beats/min and blood pressure of 120/75 mmHg. A grade II/VI pansystolic murmur was heard maximally over the apex. There were no signs of left or right ventricular failure. The chest X-ray revealed a prominent left atrial appendage and increased peripheral pulmonary vascularity, as well as the 1.5-cm nodular mass in the left upper lung. The electrocardiogram demonstrated sinus rhythm with left atrial enlargement.

Transthoracic echocardiography showed normal left ventricular size and contraction, and mild left atrial dilation. Parasternal long- and short-axis (Fig 1A,C) and apical 4-chamber imaging (Fig 1B) showed enlarged and elongated papillary muscles in direct continuity with the anterior mitral valve leaflet, and a column of fibrous tissue forming an arc extending between the apices of the papillary muscles, connecting them with the free margin of the anterior leaflet. Commissural fusion and doming of the leaflets, calcification or significant subvalvular obstruction were not observed in the mitral and aortic valves. However, the motion of the mitral valve differed from its normal rapid opening and closing. The 2-dimensional (D) mitral valve area by planimetry and the mean diastolic transmitral pressure gradient by Doppler were 2.2 cm² and 4.4 mmHg respectively, and the pressure half-time was not prolonged. Therefore, we concluded that there was no significant mitral stenosis (Fig 2A,B). Apical 2-chamber imaging with superior tilting (Fig 1D) also showed the arc-like configuration of the papillary muscles constrained by the interconnecting fibrous band. Two-dimensional apical 4-chamber imaging (Fig 3A) demonstrated restricted mobility and thickening of the anterior and posterior leaflets, which prevented complete valve closure during ventricular systole, and color Doppler imaging (Fig 3B) showed a regurgitant jet of approximately 45 ml by the proximal isovelocity surface area method, which was compatible with moderate mitral insufficiency. TEE from the mid esophageal view at 120 degrees showed a few chordae, 5–7 mm in length, appearing from the apices of the papillary muscles and attaching to the posterior mitral leaflet, causing the mobility of the posterior leaflet to be better than that of the anterior leaflet. However, the undue restraint of the motion of the anterior mitral leaflet caused by the lack of interposition of the chordae and the restricted motion of the thickened and retracted posterior mitral leaflet prevented it occupying an optimal position for valvular closure and led to mitral insufficiency during ventricular systole. An additional study using MDCT was performed to investigate the morphology of the papillary muscles and chordae and the findings were
Anomalous Mitral Arcade in an Elderly Patient

Circulation Journal Vol. 69, December 2005

similar to those of echocardiography; that is, the mitral valve leaflets almost directly attached to both papillary muscles and there was an arc-like configuration created by the column of fibrous tissue between the elongated thick papillary muscles (Fig. 4).

Examination of a transthoracic lung biopsy of the solitary pulmonary nodule disclosed squamous cell lung carcinoma. Chemoradiotherapy was performed because the stage of cancer was inoperable.

Discussion

Since the first description by Layman and Edwards,1 several cases of anomalous mitral arcade have been reported, mainly in infants and small children, with the oldest known survivor being 36 years of age.3 The current case is the first in an elderly patient. It is characterized by extreme shortening or absence of the chordae tendineae and the presence of an abnormal bridge of fibrous tissue attaching directly to the free edge of the anterior mitral leaflet that connects the 2 papillary muscles.1–10 It is suggested that this malformation is the result of an arrest in the developmental stage before attenuation and lengthening of the collagenized chordae tendineae. The lack of interposition of chordae between the anterior leaflet and the papillary muscles causes undue restraint upon the motion of the leaflet and leads to valvular insufficiency. Unusually, congenital mitral stenosis is caused by anomalous mitral arcade in which the obstructing mechanism is related to an arcade-like thickening of the line of closure of the anterior mitral leaflet, and prominent papillary muscles that crowd the subvalvular area and the left ventricular inflow tract.4

Initially, anomalous mitral arcade was described on the basis of the postmortem autopsies of 3 infants1 but thereafter several cases were detected preoperatively by catheterization and contrast ventriculography.2–7 Although these modalities aided the diagnosis, some cases were confirmed by the surgical findings. Of the 13 reported cases1–10 echocardiographic findings were described in 4 cases, and 2 of whom were diagnosed preoperatively by echocardiography.
Echocardiography was an excellent method for detecting the anomaly in the present case; however, it cannot be easily evaluated in the single imaging plane only and in order to detect anomalies of the mitral valve apparatus and associated functional disturbances, multiple 2-D and color Doppler imaging planes are needed. In this respect, multiplane TEE with color flow mapping can provide valuable adjunctive information to precordial imaging for assessing anatomical and functional status. In the present case, MDCT imaging had a relatively higher spatial resolution than echocardiography for assessing the tensor apparatus, but had no relevance to the hemodynamic work-up of this anomaly. Anomalous mitral arcade has to be differentiated from acquired disorders, such as rheumatic valvular disease, that cause mitral stenosis and insufficiency in adult patients.

The majority of the described cases1-10 had high morbidity and mortality related to significant mitral insufficiency and stenosis. The clinical and echocardiographic features of the present case (long asymptomatic period and normalized left ventricle) suggest that the anomaly functioned relatively normal for many years, maybe because there was not significant mitral stenosis and because the reserve of the anterior mitral leaflet against the restraint by the interconnecting fibrous bridge and the relatively preserved chordae of the posterior mitral leaflet prevented any mitral insufficiency from progressing. Later in life the leaflet motion of mitral valve became more restricted, possibly because of degenerative thickening of the anterior and posterior leaflets, aggravating the mitral insufficiency and causing exertional dyspnea. In contrast to the 2 reported adult cases in which the anomalous mitral valve was replaced with a mechanical prosthesis,2,3 the current patient became asymptomatic with angiotensin-converting enzyme inhibitor and diuretics and was scheduled for follow-up.

In conclusion, even in elderly patients with symptoms of mitral insufficiency and suggestive findings on transthoracic echocardiography, anomalous mitral arcade should be suspected and assessed by TEE and, if necessary, MDCT.
References


