Cor triatriatum Masked by Coexisting COPD in an Adult*

Wing-Hung Leung, M.R.C.P.; Cheuk-Kit Wong, M.R.C.P.; Chu-Fuk Lau, M.R.C.P.; and Chun-Ho Cheng, M.R.C.P.

Cor triatriatum presenting in adulthood is extremely rare. We describe a case of adult cor triatriatum in which the diagnosis was initially masked by the concomitant existence of COPD. Cardiac catheterization revealed only slightly elevated pulmonary wedge pressure despite severe pulmonary arterial hypertension. Both the primary lung disease and cor triatriatum greatly accentuated the pulmonary vascular disease which led to a reduction of pulmonary blood flow. Consequently, pulmonary venous obstruction was masked and was not reflected by measuring pulmonary wedge pressure. The diagnosis was made by two-dimensional echocardiography and left ventriculography.

*From the Department of Medicine, University of Hong Kong, Queen Mary Hospital, Hong Kong.

We report an unusual case of this anomaly in an adult in which COPD masked the manifestation of this disorder.

CASE REPORT

A 53-year-old Chinese woman first presented in 1982 with exertional dyspnea and palpitations. She was a nonsmoker. Physical examination revealed diffuse expiratory wheezing, atrial fibrillation, left parasternal heave and loud pulmonary component of second heart sound (P₂). There was no cardiac murmur. Chest roentgenogram showed normal cardiac size and increased pulmonary vascular markings (Fig 1, left). Arterial blood gas values showed hypoxemia with normal PaCO₂. Lung function tests documented moderately severe airflow obstruction with FVC 1.47L (predicted: 2.24L); FEV₁, 0.81 L (predicted: 1.83 L); FEV₁/FVC, 55%; and PEF, 3.10 L/s (predicted: 6.74 L/s). The airflow obstruction was only partially reversed with inhalational bronchodilators. Therefore, her symptoms were attributed to COPD and cor pulmonale. Other relevant investigations included normal thyroid function and serum alpha-antitrypsin level. She was treated with inhalational bronchodilators. A trial of oral corticosteroid showed no improvement in the degree of airflow obstruction. Digoxin and verapamil were used for the control of atrial fibrillation. She remained stable until October 1986 when she complained of gradual deterioration in exercise tolerance and the appearance of bilateral ankle edema. She was given diuretics in addition. During the past six months, she required hospital admission three times because of shortness of breath. In the latest admission, physical examination revealed atrial fibrillation, left parasternal heave, loud P₂, and tricuspid regurgitation. A repeat chest roentgenogram showed progressive cardiomegaly with bulging in the left atrial border, double right atrial border, pulmonary venous and pulmonary arterial hypertension.¹²

FIGURE 1. Posteroanterior chest views: (left) normal cardiothoracic ratio and increased pulmonary vascular markings on initial presentation; (right), bulging left atrial border, double right atrial border, pulmonary venous congestion and progressive cardiomegaly.
venous congestion as well as a more dilated proximal main pulmonary artery when compared with the previous chest x-ray (Fig 1, right). Arterial blood gas values showed compensatory respiratory acidosis with hypoxemia (pH, 7.38; PaCO₂, 8.54 kPa; PaO₂, 7.2 kPa; actual bicarbonate, 37.9 mmol/L, and base excess, +10.6, on room air). A repeated lung function test confirmed progression of the airflow obstruction with FVC, 1.39 L (predicted: 2.12 L); FEV₁, 0.68 L (predicted: 1.73 L); FEV₁/FVC, 49 percent; and PEF, 2.20 L/s (predicted: 6.38 L/s). In view of the chest roentgenographic findings, two-dimensional echocardiogram was performed which revealed an abnormal membrane present in the left atrium dividing it into superior and inferior chambers in both the long axis and apical four chamber views (Fig 2). The right atrium, right ventricle, and the pulmonary artery were dilated. The left ventricle was small with satisfactory systolic function. The diagnosis of cor triatriatum in addition to COPD was made. Cardiac catheterization was performed. There was severe pulmonary hypertension (100/40, mean 60 mm Hg) and the mean pulmonary capillary wedge pressure was 15 mm Hg. The mean gradient between the pulmonary capillary wedge pressure and left ventricular end-diastolic pressure was 10 mm Hg by planimetry. The cardiac index was 2.1 L/min/m² and the pulmonary vascular resistance was 19 Wood units (1520 dynes.cm⁻².m⁻¹). By using the Gorlin formula, the calculated area of the left atrial membrane orifice was 0.54 cm² which gave an orifice diameter of 8 mm. There was no evidence of intracardiac shunting. In view of the risk of pulmonary angiography in this patient, left ventricular cineangiography in the right anterior oblique view was performed which showed a small sized left ventricle with good systolic contraction. The catheter was manipulated during the contrast injection so that ventricular ectopics occurred. The regurgitation of the contrast medium induced by ventricular ectopics entered the left atrium outlined the cor triatriatum. Despite the high operative risk in view of the poor lung function, surgical excision of the left atrial membrane was offered. However, the patient refused operation and she was then managed conservatively.

**DISCUSSION**

Cor triatriatum usually presents in infancy and childhood. Rarely, less severe cases of obstruction may allow survival to adulthood. In a review of 36 patients by Niwayama, ten were greater than 12 years old. Our patient, age 53, represents one of the oldest reported living patients. Adult patients usually present with symptoms of pulmonary congestion, atrial fibrillation, and occasionally, cerebral embolism.

In our patient, the initial diagnosis of cor triatriatum was masked by the coexisting pulmonary problem. The diagnosis of COPD was substantiated by the predominant obstructive pattern in lung function tests and the presence of chronic compensated respiratory acidosis. The large difference between pulmonary artery diastolic pressure and the pulmonary wedge pressure suggested the presence of pulmonary vascular disease. The severe pulmonary vascular disease with resultant severe pulmonary hypertension in this patient was related to the presence of both cor triatriatum and COPD, but the extent to which each contributed is difficult to assess. The fact that clinical manifestations of pulmonary congestion like orthopnea, paroxysmal nocturnal dyspnea and pulmonary edema were not present despite the small orifice area of the anomalous membrane was due to the reduced pulmonary arterial blood flow. Consequently, pulmonary venous obstruction was masked and could not be assessed by measuring pulmonary wedge pressure. Moreover, the gradient across the left atrial membrane may also be underestimated. Lang et al. have also reported a child with cor triatriatum masked by coexisting primary pulmonary hypertension in which cardiac catheterization revealed high pulmonary arterial pressure and normal pulmonary wedge pressure. For these reasons, surgical excision of the left atrial membrane was entertained in our patient despite the poor pulmonary function.

Two-dimensional echocardiography, which was the key to the diagnosis in this patient, is generally considered to be the optimal noninvasive method of identifying the anomaly. The use of the venous phase of the pulmonary angiogram is the classic angiographic diagnostic method. However, in view of the underlying lung problem in our patient, pulmonary angiography was not performed, as there is a significant risk in such group of patients. Instead, the left atrial membrane was demonstrated by contrast regurgitating into left atrium during left ventriculography.

**REFERENCES**

Interrupted Aortic Arch in an Asymptomatic Adult*

Cheuk-Kit Wong, M. B.; Chun-Ho Cheng, M. B.; Chu-Buk Lau, M. B.; Wing-Hung Leung, M. B.; and Fu-Luk Chan, M. B.†

Isolated interrupted aortic arch is a rare congenital cardiac disorder believed at one time to be incompatible with life once the ductus arteriosus closed. To our knowledge, only 12 cases have been reported in the literature and mostly in children. The diagnosis was made in all of them by catheterization. We report the first adult patient with an asymptomatic interrupted aortic arch diagnosed by computed tomography.

 Interruption of the aortic arch is a rare anomaly.1,4 Although commonly associated with serious complex cardiac lesions with a poor prognosis,5,9 cases of isolated interruption have been reported with symptoms of exertional claudication and paresthesia in childhood.2,3 The site of the interruption has been used for classification.9 Type A interruption occurs distal to the subclavian arteries, type B occurs distal to the left common carotid artery, and type C just distal to the origin of the innominate artery. The genesis of aortic interruption and the more commonly encountered aortic coarctation have been investigated by various workers.8,7 CT appearance of aortic coarctation has recently been described.8,10 We report the first adult patient with an asymptomatic type A aortic interruption diagnosed by CT.

CASE REPORT

A 33-year-old man presented to our medical unit for investigation of hypertension. On physical examination he was well built, with blood pressure of 160/100 mm Hg. The pulses were equal over both upper limbs, and a radial-femoral delay was noted. Both femoral pulses were equal but weak. The blood pressure measured 70/50 mm Hg over both thighs by Doppler measurement. The heart was mildly enlarged with a left ventricular impulse. The auscultatory findings were normal. Fundoscopy showed grade 2 hypertensive changes. An ECG confirmed left ventricular hypertrophy with a strain pattern. A chest roentgenogram showed bilateral rib notching. An echocardiogram showed concentric left ventricular hypertrophy with a normal tricuspid aortic valve. He was diagnosed as having coarctation of the aorta and given antihypertensive drugs. The patient was reluctant to undergo cardiac catheterization. A CT of the thoracic aorta was performed on a General Electric CT 9800 scanner utilizing dynamic scanning with table incrementation during rapid injection of contrast medium by a power injector (Fig 1 and 2). The aorta was found to be interrupted at the level of the

*Department of Medicine, University of Hong Kong.
†Department of Diagnostic Radiology, Queen Mary Hospital, Hong Kong.