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Multiple Coronary Arteriovenous Fistulae Combined with Ventricular Septal Defect*: A Case Report

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INTRODUCTION

Congenital coronary arteriovenous fistulas are all of those anomalies and malformations that result in a direct communication of a coronary artery and/or any of its branches with a cardiac chamber or extracardiac vessel resulting in a shunt of varying proportions. The majority of these fistulas form an A-V connection between a coronary artery and the right heart chambers, and rarely their is communication with the left heart chambers.

In most reported cases, the abnormal fistula involves only one coronary artery which developed alone.

This report describes a unique case in which multiple coronary fistulas associated with VSD were encountered.

CASE REPORT

A 53 year old woman was admitted because of exertional dyspnea, intermittent palpitation, easy fatigability and mild chest discomfort. She experienced frequent episodes of palpitation for 10 years, but had managed to live a relatively normal life without treatment.

During a nine month period, symptoms developed and she was medicated with seldatives and antacids, but her condition was gradually aggravated. She was then transfered to our hospital and a heart murmur was discovered on physical exam.

She had no remarkable past medical history or family history. Physical examination revealed a well developed and well nourished woman in no acute distress. Her pulse rate was 82/min and regular, and her blood pressure was 100/80mmHg.

A grade 4/6 pansystolic murmur was best heard at the cardiac apex. There is no palpable thrill noted. An electrocardiogram revealed normal sinus rhythm with a primary A-V block and left ventricular hypertrophy. The chest X-ray was within normal limits. Cardiac catheterization revealed oxygen step-up(9%) at the right ventricular level, and the Qp/Qs was 1.8. Left vetricular cineangiography in the left anterior oblique projection demonstrated an extremely dilated and tortous vessel that corresponded to the left circumflex coronary artery. A small VSD and opacification of the right ventricle(Fig. 1) was also seen.

A selective left coronary arteriogram satisfactorily opacified this vessel. The coronary sinus, right atrium, right ventricle and pulmonary artery(Fig. 2) were also visible.

A right coronary arteriogram demonstrated an enlarged vessel of the terminal branch of the posterior descending artery which terminated in a small fistulous stoma emptying into the left ventricle (Fig.

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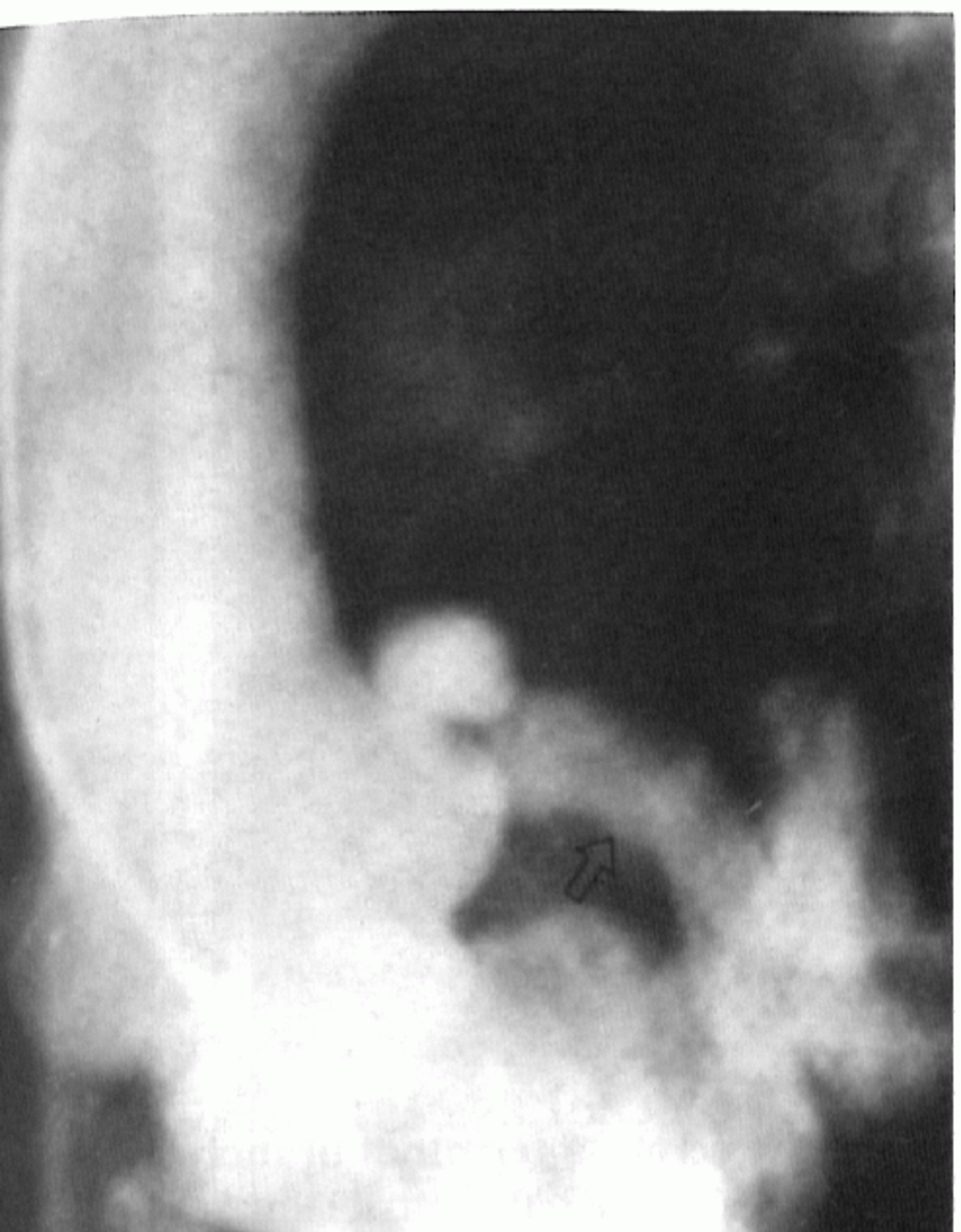


1. LAO view of left cineventriculogram reveals an emely dilated tortous left coronary artery(open ar- and a small ventricular septal defect(arrow).

3).

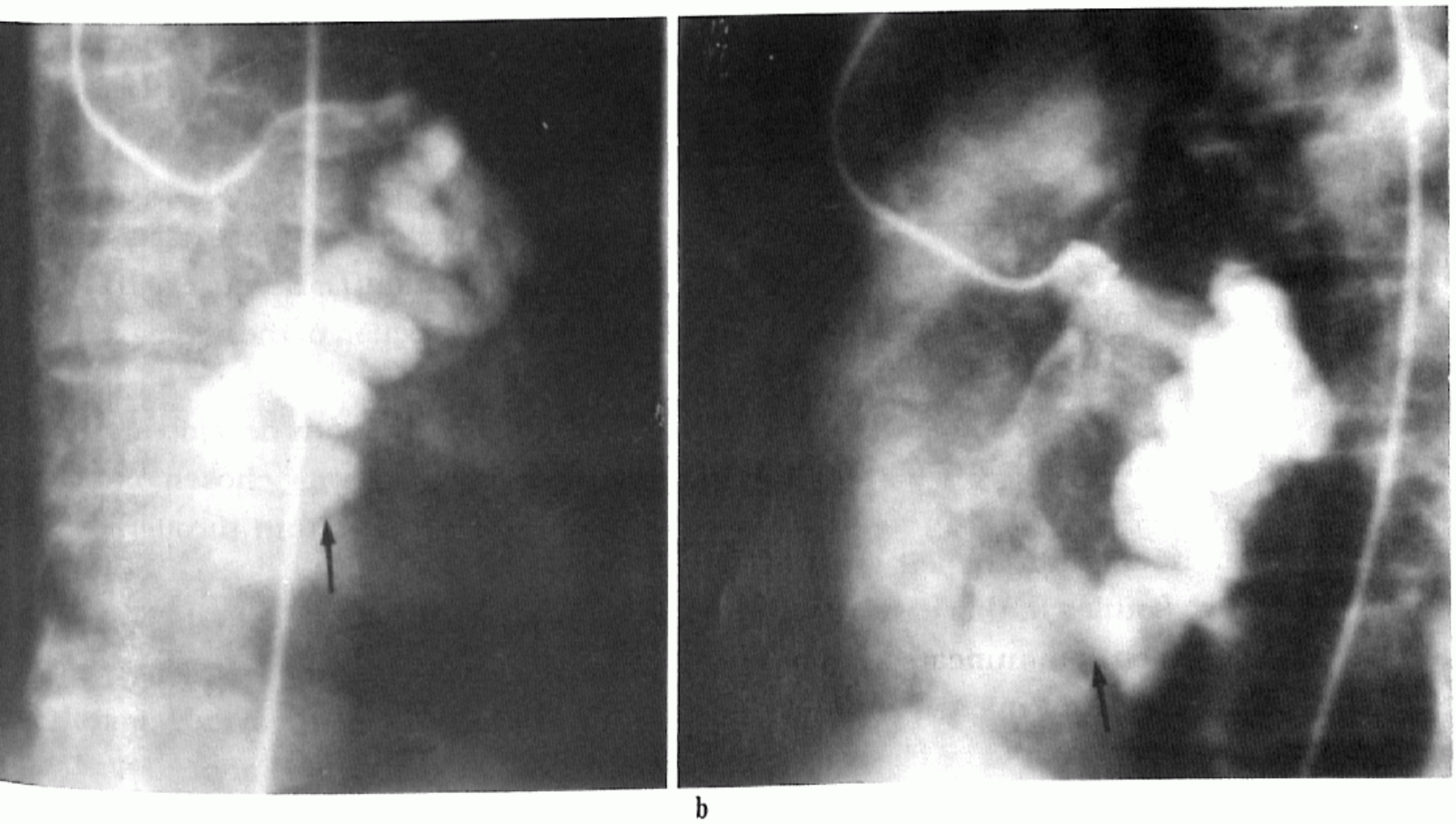
The left coronary arterial fistulous tract was ligated by transfixion sutures and the VSD was repaired by direct closure.

However, the right coronary arterial fistula was not repaired, because the 02 step-up at the ventricular level was mainly due to the left coronary arterial fistula and VSD. The patient did well postoperatively, and the murmur completely disappeared.

DISCUSSION

Coronary arterial fistulas may be formed by a well defined vessel emptying into a cardiac chamber or by a plexus of multiple fine vessels. Myocardial vessels in the adult heart are derived embryologically from both endothelial protrusions into the myocardium and coronary arteries and veins. This endothelial protrusion into the myocardium extends to the epicardial surface, forming an intertrabecular network. The outermost intertrabecular network becomes almost completely obliterated and forms a capillary network as the myocardium grows, where as the innermost intertrabecular vessels retain their communication with chambers of the heart, forming Thebesian vessels of the adult heart.

Investigators reporting earlier cases, in which sinusoidal communication existed between involved coronary arteries and the heart chamber, ascribed the



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2. a,b. Selective left coronary arteriogram reveals tortuous aneurysmally dilated left circumflex coronary artery mmunicating with the dilated coronary sinus(arrow)(A:AP B:LAO).

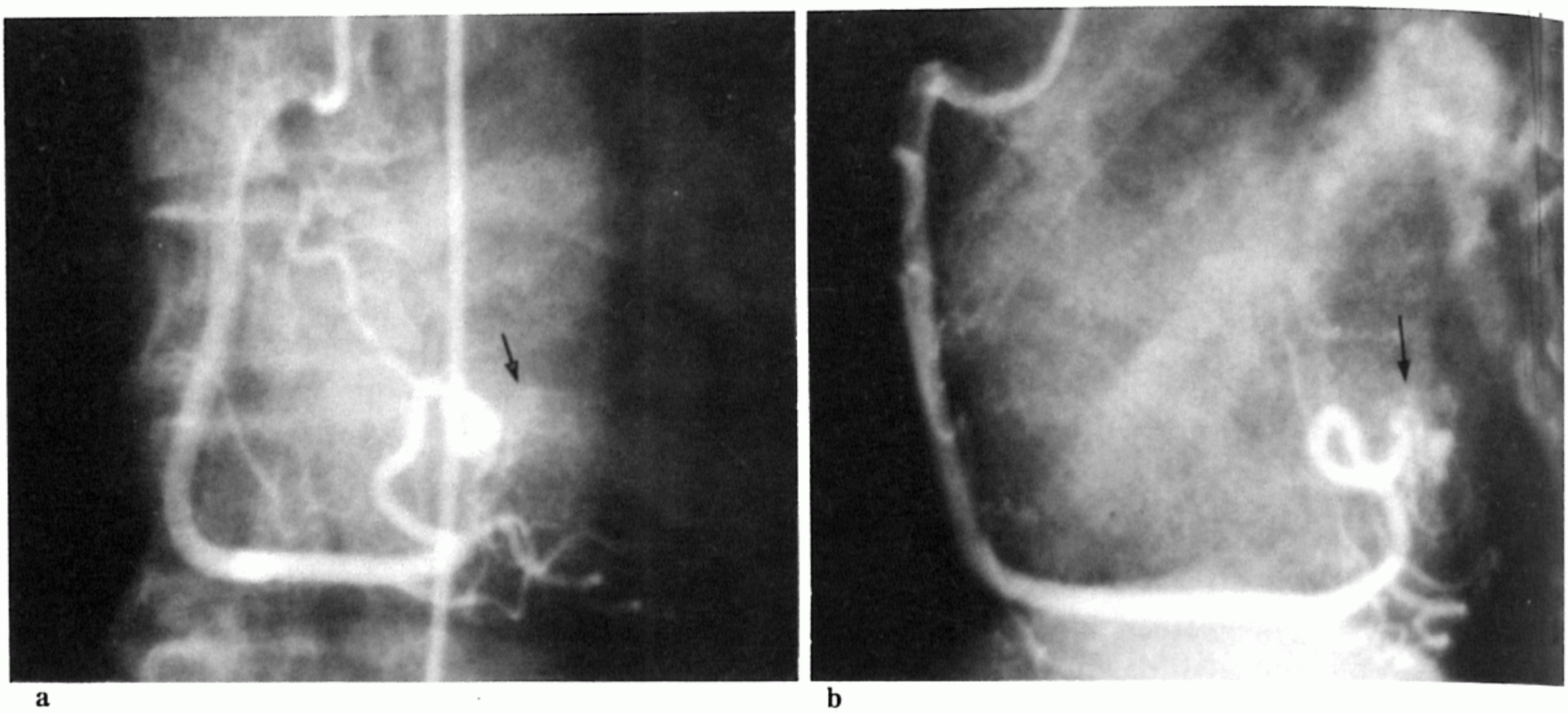

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Fig. 3. a,b. Selective right coronary arteriogram reveals an enlarged vessel of its terminal branch of the posterior desoending artery which terminated in a small fistulous stoma emptying into the left ventricle(arrow)(A:AP B:LAO).

anomaly to persistence of the outermost intertrabecular space, there by permitting communication between the coronary artery and the heart chamber through the Thebesian vessels(3).

The coronary artery involved is usually extremely dilated and tortous. The dilatation starts from the origin of the involved vessel and extends to the point of communication. However, the size of the orifice of the communicating vessel is relatively small in comparison with the dilated coronary artery. The branches distal to the point of communication are relatively small.

The recipient coronary sinus or vein, which also becomes aneurysmally dilated and thin-walled, serves as a site of blood clot formation.

Congenital coronary arterial fistulas usually arise from a single artery, most frequently the right coronary artery, and usually communicate with the right ventricle, right atrium or pulmonary artery(4). the fistula between the right coronary artery and left ventricle is quite rare. One case was reported in Korea, which revealed a marked dilatation of the right coronary artery(diameter 3cm) and a large fistulous stoma(diameter 1.5cm) into the left ventricle(5).

One of the two fistulas in this case was the right coronary artery which communcated the left ventricle. The stoma of the fistula opened into the left ventricle but was small enough to be closed during the systolic phase by ventricular contraction after which mild dilatation of the right coronary artery was noted.

The majority of patients in the younger age group with this anomality are asymptomatic and may reach the age of 30 to 40 years without difficulty. However most of them eventually will become symptomatic later in life as a result of the long-standing hemodynamic abnormality, complications of the disease, the development of coronary arteriosclerosis, or hypertension(6).

If symptoms occur, most frequent symptoms are chest pain, dyspnea, palpitations and easy fatigability. Chest pain is probably due to myocardial ischemia secondary to the shunt, that is, a large amount of oxygenated blood that normally would pass through the myocardial capillaries is shunted instead through the fistula(6).

Pulmonary hypertension, congestive heart failure and subacute bacterial endocarditis are the most common complication(7). Because of these complications, surgical correction of the anomalies is the treatment of choice.

In this case, palpitations developed at age 43, and surgical intervention was chosen because of the significant left to right shunt through the coronary arterial fistula and VSD.

In most cases, the congenital coronary arterial fistula develops alone. But infrequently several combined anomalies are reported; Patent ductus arteriosus, Tetralogy of Fallot, Mitral stenosis etc(8-10).

To the best of our knowledge, this is the first case

of a congenital coronary arterial fistula combined with an interventricular septal defect, written in the English literature. We propose that, in a congenital coronary arterial fistula, a complete cardiac catheterization and a cardiac angiogram should be performed for further evaluation of combined cardiac anomalies.

REFERENCES

1. Yu YJ, Han MC, Park JH: Congenital coronary arteriovenous fistulae. The journal of the Korean radiological society 1982;18:744-750
2. Oh YH, Kim H, Zeon SK, Suh SJ: Congenital coronary artery fistula. The journal of the Korean radiological society 1986;22:1083-1086
3. Reddy K, Gupta M, Hamby RI: Multiple coronary arteriosystemic fistulas. Am J Cardiology 1974; 33:304-306
4. McNamara JJ, Gross RE: Congenital coronary artery fistula. Surgery 1969;65:59-69
5. Lee BH, Yu SJ, Moon ES, Kim SH, Choi YH: A case report of congenital coronary artery fistula to the left ventricle. The journal of the Korean radiological society 1987;23:420-423
6. Kimbiris D, Kasparian H, Knibbe P, Brest AN: Coronary artery-coronary sinus fistula. Am J Cardiology 1970;26:532-539
7. Steinberg I, Baldwin JS, Dotter CT: Coronary arteriovenous fistula. Circulation 1958;17:372-390
8. Schaffer AB, St Ville J, Mackler SA: Coronary arteriovenous fistula with patent ductus. Am Heart J 1963;65:758
9. Busch Vw, et al: Late deferioration in tetralogy of Fallot. Arch Internal Med 1978;138:1423
10. King SB, Schonmaker FW: Coronary artery to left atrial fistula in association with severe atherosclerosis and mitral stenosis. Chest 1975; 67:361

<국문 요약>

심실 중격 결손증과 동반된 다발성 관상 동정맥루 1예 보고*

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선천성 관상동맥루는 매우 드문 심질환으로서, 국내에서도 몇 예가 보고된 바가 있으나(1, 2) 다발성 관상 동맥루는 보고된 바가 없으며 세계적으로도 희귀한 증례이다.

최근 저자들은 53세의 여자에서 우측 관상동맥이 좌심실과, 그리고 좌측 만곡 관상동맥이 우심실과 연결된 다발성 관상동맥루와 동반된 심실중격 결손증 1예를 경험 하였기에 보고하는 바이다.