ISOLATED RIGHT SUPERIOR VENA CAVA DRAINING INTO LEFT ATRIUM DIAGNOSED BY RADIONUCLIDE ANGIOCARDIOGRAPHY

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The first adult case of a right-to-left shunt due to the congenital isolated right superior vena cava draining into the left atrium is reported. The diagnosis was made by a simple technique using intravenous radionuclide angiocardiography.

Intravenous radionuclide angiocardiography using a short-lived radionuclide, a gamma scintillation camera, and a video data recording system has recently been developed and used as a practical clinical test for various congenital and acquired cardiovascular diseases (*1–6*).

This report is of a case with a right-to-left shunt caused by an isolated right-sided superior vena cava draining into the left atrium. This congenital anomaly is very rare and, to our knowledge, is the first to be reported in an adult and diagnosed by simple and noninvasive isotopic angiocardiography.

CASE REPORT

A 34-year-old black woman was admitted to the University of Cincinnati Medical Center for evaluation of vague complaints of occasional palpitations, dyspnea on exertion, and two pillow orthopnea which had lasted 1 year.

The patient had an apparently normal gestation and delivery. There was no cyanosis or respiratory distress after birth. The patient sustained a cerebral concussion and pelvic fracture from an automobile accident during her childhood. She had normal growth, development, and activity throughout her childhood. There was no history of rheumatic fever, hypertension, hemoptysis, or seizure disorder. Subsequently, she carried five normal pregnancies without difficulty. About 3 years ago, the patient was hospitalized for depression and suicidal tendencies.

Physical examination revealed a thin woman in no distress. Her blood pressure was 100/70 mmHg; pulse 80/min; temperature and respiration normal. There was a suggestion of mild acrocyanosis but no clubbing. Mucous membranes were normal in color. The fundi were normal. The neck veins were not distended. No bruits were heard over the great vessels. The lungs were clear. There was no cardiac enlargement, murmur, gallop, hepatosplenomegaly, or pedal edema. Pelvic and neurologic examinations showed no abnormalities.

Admission chest x-ray revealed a normal cardiac silhouette, normal vascularity, and clear lung fields (Fig. 1). The EKG revealed minor, nonspecific S-T and T-wave changes without evidence of specific chamber enlargement. The hematocrit was 57%, hemoglobin 18 gm%; WBC, platelet count, and RBC morphology were normal. Evaluation just before this admission had revealed a serum pH of 7.35, pCO_2 of 34 mmHg, and pO_2 of 50 mmHg, which rose to 89 mmHg following positive pressure breathing of 100% oxygen at 15 cm of water pressure for 30 min. Pulmonary function tests including

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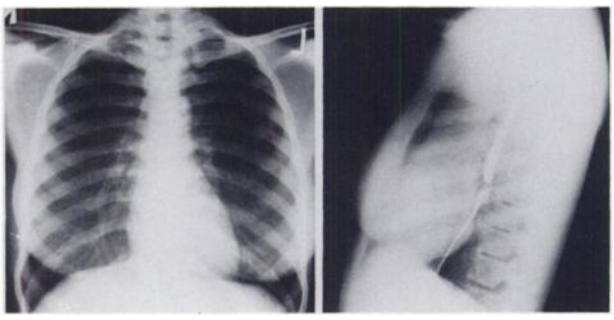

FIG. 1. P-A and left lateral chest x-ray on admission showing no abnormality.

carbon monoxide diffusion were normal. Red cell mass (by ^{51}Cr) was 1,953 ml (normal 1,353 ± 258 ml — 2 s.d.) and plasma volume (^{125}I-IHSA) was 2,239 ml (normal 2,227 ± 392 ml — 2 s.d.). Other findings including intravenous pyelogram, hemoglobin electrophoresis, and liver function studies were within normal limits.

A radionuclide angiocardiogram and lung scan were done to search for an intracardiac or intrapulmonic shunt lesion.

PROCEDURE AND FINDINGS

Radionuclide angiocardiography. These studies were carried out using a Nuclear-Chicago (Pho/Gamma III) scintillation camera with a 4,000-parallel-hole, high-sensitivity collimator.

A dose of 3 mCi of ^{99m}Tc-macroaggregated albumin was injected as a bolus into the right antecubital vein. The flow of activity was viewed on the persistence scope, and the data were simultaneously recorded on the Data-Store/Play Back Accessory System (Nuclear-Chicago 3122). Sequential 2–4-sec exposure scintiphotographs were obtained when the recorder was played back. Blurred focus was used to reduce background activity on the scintiphotographs (Fig. 2).

With the patient in the left anterior oblique position, the flow of activity was seen passing through the right subclavian vein and entering the superior vena cava (Fig. 2, A-1). The bolus then proceeded downward and to the left and by an anomalous route entered the left atrium (Fig. 2, A-2). It then entered the left ventricle (Fig. 2, A-3) and filled the thoracic aorta, subclavian, and carotid arteries (Fig. 2, A-4). The pulmonary circulation was completely bypassed, and no activity was localized in the lung. Instead, intense activity was seen in the systemic circulation—liver, spleen, and brain.

Next, the course of the inferior vena cava was visualized by injection of 10 mCi of ^{99m}Tc-sodium-pertechnetate into the right saphenous vein with the patient in the supine position. Activity was seen in the inferior vena cava (Fig. 2, B-1, B-2); then the flow passed through the right atrium, right ventricle, and pulmonary outflow tract which showed the normal U-shaped configuration (Fig. 2, B-3). Both lungs were visualized leaving a nonvisualized area between the right ventricle and the left lung, which is the location of the left ventricle (Fig. 2, B-4). There was no early filling of the left ventricle or aorta from the right ventricle as one might expect in a septal defect with right-to-left shunt.

A third study was done to determine the course of the left subclavian vein. Ten mCi of ^{99m}Tc-sodium-pertechnetate was injected into the left antecubital vein with the patient in the left anterior oblique position. Again, the flow followed the same direction as seen in the first study—namely, right-sided superior vena cava, left atrium, left ventricle, and aorta (Fig. 2, C-1, C-4). Frames C-4 and A-4 in Fig. 2 may not appear to match exactly, but this slight difference was a result of minimal rotation of the patient's position. The radionuclide angiocardiographic findings indicated that the right-sided superior vena cava drained into the left atrium, and there was a normal inferior vena cava emptying into the right atrium. There was no evidence of persistent left superior vena cava.

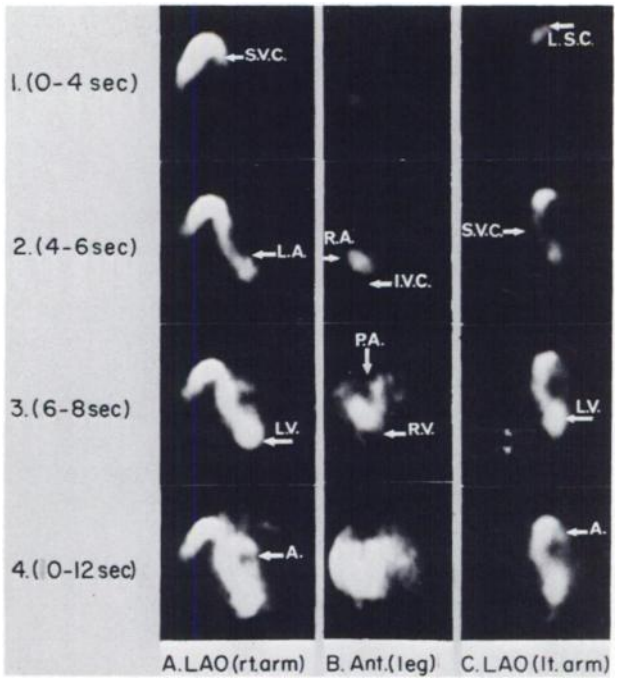

FIG. 2. (A) Sequential scintiphotos following injection of 3 mCi ^{99m}Tc-MAA into right antecubital vein. Notice activity in left atrium, left ventricle, aorta, and absence of activity in lungs. (B) Injection of 10 mCi ^{99m}Tc-pertechnetate into right saphenous vein shows right ventricle and both lungs. (C) Injection into left antecubital vein shows same sequence of activity as in A.

Cardiac catheterization finding. Cardiac catheterization from the right antecubital vein was performed to confirm the anomaly. When the catheter was advanced toward the heart, the catheter tip entered the superior vena cava and then entered the left atrium.

The atrial septum was not crossed despite considerable probing, and normal pulmonary veins were identified. Oxygen saturation at this point was 89 vol%. Further manipulation caused the catheter tip to advance into the left ventricle, which was in its normal, posterior position.

DISCUSSION

The discovery of this congenital cardiac anomaly, proven by radionuclide angiocardiography, was strik-

ing for several reasons. The patient had grown and developed normally without experiencing physical difficulty until recently, and had tolerated well the large right-to-left shunt that was estimated to be one-third of the total venous return (*7*). No murmur was ever audible and there was no evidence of cardiac or pulmonary disease on routine chest x-rays.

This is the fourth case, to our knowledge, to be reported, and the first in an adult. All four were female. The three cases reported previously were female children, aged 10, 2, and 3 years, respectively. In the first case, surgical intervention was not thought to have been warranted because the disability was too slight (*8*). Two other patients underwent surgical correction with good results (*9,10*). Surgical correction of this anomaly has been suggested to our patient, but she was reluctant to submit to surgical intervention because of her relatively minor discomfort at this time.

Anomalies of the great veins are not uncommon. The most frequent consists of bilateral superior venae cavae with a persistent fetal left superior vena cava that enters the right atrium through the sinus of the coronary veins; thus no physiologic derangement results (*11*). In 7.5% of these cases, the left superior vena cava terminated in the left atrium (*12*). The vast majority of the latter had associated significant cardiac abnormalities. Long-term survival in a relatively healthy individual with a large right-to-left shunt from an isolated anomalous drainage of the inferior vena cava to the left atrium has been reported (*13*). A case of a single, left-sided superior vena cava draining into the left atrium with left ventricular hypertrophy and no associated anomalies has been reported in a 15-year-old boy (*14*). Anomalous drainage of a right superior vena cava into the left atrium as a completely isolated defect, however, is a very rare anomaly resulting in a large right-to-left shunt.

The embryology of this malformation is not clear. However, malposition and aberrant development of the right horn of the sinus venosus seems the best explanation for this anomaly (*9*).

This is the first such case diagnosed by a simple radioisotope technique, which is available to many institutions with a nuclear medicine unit.

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