jnm/CASE REPORT

MULTIPLE PULMONARY ARTERIOVENOUS FISTULAS DEMONSTRATED BY DYNAMIC RADIONUCLIDE PULMONARY PERFUSION SCANNING

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Dynamic radionuclide perfusion scintigraphy performed with ^{99m}Tc-labeled microspheres was instrumental in establishing the diagnosis of multiple, small-vessel, pulmonary arteriovenous fistulas in a 14-month-old patient with cyanosis. Computer analysis of the sequential distribution of tagged microspheres in the pulmonary parenchyma normally demonstrates a curve that rises rapidly to a plateau as the particles microembolize. In the case reported here, the pulmonary flow curve rose rapidly to a maximum and then fell within 2 sec to a plateau of less than 50% of the maximum count, indicating that a large proportion of the microspheres passed through the pulmonary circulation. Conventional pulmonary contrast angiography did not demonstrate any intracardiac shunting but did confirm the presence of multiple pulmonary arteriovenous fistulas.

The study of congenital cardiopulmonary shunting with dynamic radionuclide imaging and computer analysis of flow curves is now well established and has been recently reviewed by Greenfield (*1*). Right-to-left transpulmonary shunts may be identified through the use of albumin microspheres (*2*) or albumin macroaggregates (*3*) labeled with ^{99m}Tc. In the presence of an intracardiac right-to-left shunt, some of the labeled particles bypass the pulmonary circulation and appear rapidly in the liver, spleen, kidneys, and brain. This case report details the use of the dynamic perfusion pulmonary study done with ^{99m}Tc-labeled microspheres to demonstrate an extracardiac right-to-left shunt through multiple, diffuse, small pulmonary arteriovenous fistulas.

CASE REPORT

A 6-week-old child was admitted to Strong Memorial Hospital for evaluation of cyanosis. Physical examination revealed mild cyanosis of the fingers and lips while the patient was resting quietly. There were good peripheral pulses and the examination of the heart and lungs was normal. No cutaneous lesions were present. The chest roentgenogram was within normal limits. The hematocrit was 65%. Initially, the patient was suspected of having tetralogy of Fallot with pulmonary atresia. Conventional cardiopulmonary contrast angiography showed no evidence of intracardiac lesions nor any abnormalities of the pulmonary circulation. There was good flow of contrast to all segments of pulmonary parenchyma. Cardiac catheterization demonstrated normal cardiopulmonary pressures; however, oxygen saturation determinations showed the right ventricular blood to have a saturation of 62% whereas the pulmonary venous blood had a saturation of only 72%, indicating marked lack of oxygenation of blood passing through the pulmonary vasculature. A static pulmonary perfusion scan done with ^{99m}Tc-labeled microspheres demonstrated uptake of radionuclide in the brain and kidneys, indicating a right-to-left shunt. It was not possible to determine whether the shunt was intracardiac or extracardiac on the basis of the scan. The patient was thought to have pulmonary parenchymal disease of unknown etiology causing severe oxygen desaturation of pulmonary venous blood. She was managed with intermittent phlebotomy during the ensuing year.

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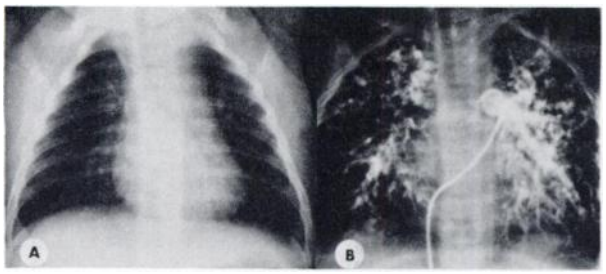

FIG. 1. (A) PA chest film done when patient was 14 months of age showing multiple small nodular densities throughout both lung fields. (B) Selected AP view from second pulmonary arteriogram confirming presence of multiple diffuse arteriovenous communications.

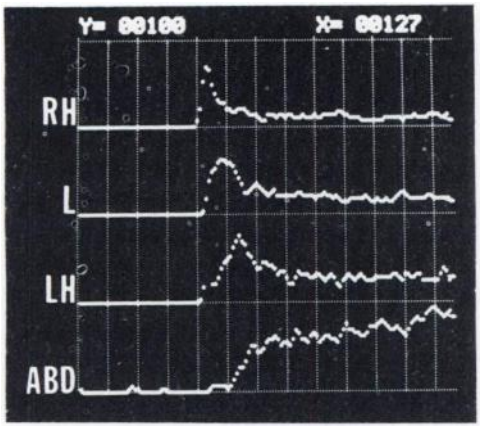

FIG. 2. Computer flow curves of right heart (RH), lungs (L), left heart (LH), and liver (ABD) demonstrating passage of microspheres through pulmonary vasculature and early systemic activity. Each vertical bar represents 2 sec.

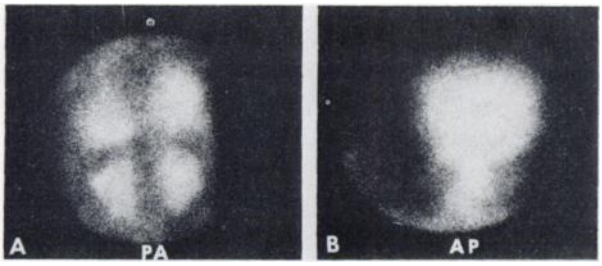

FIG. 3. (A) Static posterior scan showing microspheres in lungs, kidneys, and spleen. (B) Static anterior view of skull showing microspheres trapped in circulation of head and neck.

At age 14 months, the child was readmitted for re-evaluation of polycythemia and cyanosis. Physical examination of the heart and lungs was still normal. At this time, however, the chest roentgenogram showed multiple, tiny, nodular densities less than 2 mm in size throughout both lung fields (Fig. 1A). Dynamic pulmonary perfusion scintigraphy was performed with ^{99m}Tc-labeled microspheres and the flow pattern to the heart, lungs, and liver was analyzed by computer (Fig. 2). The curves indicate extremely rapid transit of the radionuclide through the lung fields (less than 2 sec) with early accumulation of activity in the systemic circulation. The flow curve for the lungs showed the normal rapid rise but then sharply declined to plateau at less than 50% of the maximum count. Normally, the curve should plateau at the maximum since virtually all the tagged particles lodge in the pulmonary microcirculation. The shape of the curve demonstrates that only part of the microspheres caused pulmonary microembolization while the remainder passed through the pulmonary vasculature into the systemic circulation. Quality-control procedures ruled out the presence of free, unlabeled ^{99m}Tc. Static images documented the presence of tagged microspheres in the liver, spleen, brain, and kidneys (Fig. 3A and B). The results were interpreted to indicate a marked pulmonary parenchymal arteriovenous shunt at the arteriolar level. Coexistent intracardiac right-to-left shunt could not be ruled out by the dynamic perfusion study. Pulmonary contrast angiography confirmed the presence of multiple diffuse small arteriovenous communications with a right-to-left atrium transit time of 0.7 sec (Fig. 1B). No intracardiac shunting was demonstrated. The diagnosis of multiple, pulmonary small-vessel arteriovenous fistulas was made and a trial of steroid therapy was undertaken with no significant improvement in the condition of the patient. She is currently being managed with intermittent phlebotomy.

DISCUSSION

Utzon (*4*) recently reviewed pulmonary arteriovenous fistulas in children and concluded that the spectrum of the pathologic communications between pulmonary arteries and veins may range from one or a few isolated arteriovenous fistulas to innumerable telangiectatic anastomoses. He identifies the latter condition as the "disperse telangiectatic type," which appears to be the lesion of the patient in this report. The right-to-left intrapulmonary shunt results in reduced oxygenation of the blood in the systemic circulation, which in turn results in cyanosis and polycythemia. The patient generally presents with dyspnea and cyanosis. Digital clubbing and cutaneous telangiectasia may also be present. These patients are usually suspected of having congenital defects of the heart on the basis of the initial symptomatology. Conventional cardiopulmonary contrast angiography, however, rules out intracardiac shunting and often demonstrates the arteriovenous communications. Cardiac catheterization will show normal cardiac and pulmonary pressures with marked desaturation of pulmonary venous blood.

In the case reported here, initial cardiac catheterization did show pulmonary venous blood desaturation with normal cardiac and pulmonary pressures; however, pulmonary angiography initially failed to show any evidence of intracardiac or intrapulmonary shunting. Eighteen months later the dynamic perfusion pulmonary study was done and showed that part of the labeled microspheres traversed the pulmonary vasculature and lodged in other systemic organs. This finding could not be accounted for on the basis of intracardiac shunting and therefore the diagnosis of pulmonary arteriovenous shunting at the arteriolar level was made and confirmed by conventional pulmonary arteriography. Clinicians should be aware that the presence of a right-to-left shunt demonstrated by static perfusion scan does not localize the site of the lesion and that computer-assisted dynamic perfusion pulmonary scintigraphy is a valuable tool that helps in the evaluation and diagnosis of this unusual group of patients.

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ERRATUM

Regarding the abstract, "Evaluation of Learned 'Puffing Response' of Monkeys with In-113m Labeled Smoke," by G. D. Robinson, Jr., and R. K. Siegel, included in the Proceedings of 21st Annual Meeting (*J Nucl Med* 15: 528, 1974), C. A. Johnson was inadvertently omitted as the third author.