

# RADIONUCLIDE STUDIES IN HEMANGIOENDOTHELIOMATOSIS: CASE REPORT

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***Multiple abnormal radionuclide studies of brain, lungs, kidneys, liver, and spleen were observed in a patient with hemangioendotheliomatosis.***

Hemangioendothelioma is a very rare, locally invasive vascular tumor. This case report describes a patient with hemangioendotheliomatosis in whom radionuclide imaging studies of the brain, lung, kidney, liver, and spleen were performed. The results of these studies are reported and discussed in relation to other studies done and to the pathologic process.

## CASE REPORT

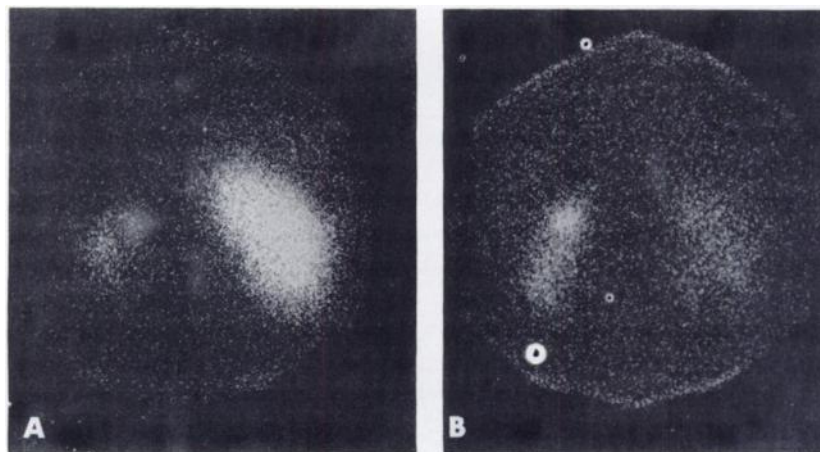
A 20-year-old man was admitted because of exertional dyspnea, palpitations, and "blackouts" of several months duration. He was known to have had severe hypertension for 6 years and had been hospitalized elsewhere for a stroke 1 year before. Previous studies had demonstrated pulmonary hypertension, a bilateral renal arterial lesion, and a cerebral arterial defect. On examination the blood pressure was 250/150 mmHg. The heart was en-

larged with an accentuated pulmonic sound. There was a systolic bruit over the lung fields posteriorly and laterally. There was no neurologic deficit. Laboratory data were not significantly altered, with a normal SMA-6 and SMA-12. An x-ray film of the chest revealed cardiomegaly with normal lung fields. The intravenous pyelogram revealed absent left renal function. Cardiac catheterization and pulmonary angiography demonstrated severe pulmonary hypertension and several areas of marked narrowing and irregularity of the secondary and tertiary pulmonary arteries.

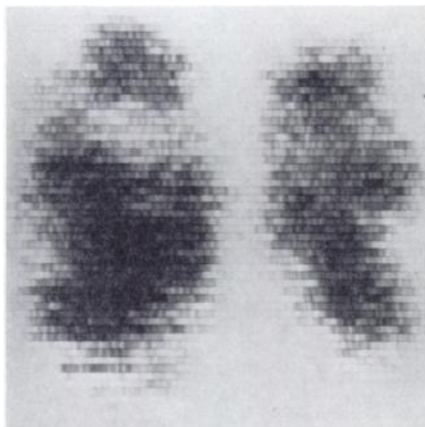
**Radionuclide studies.** Technetium cerebral flow studies showed the major cerebral arteries to be poorly outlined including the carotids. Brain scan was unremarkable. Renal scan with  $^{197}\text{Hg}$ -tagged chlormerodrin outlined a small left kidney, with only residual uptake (Fig. 1A). Renogram with  $^{131}\text{I}$ -hippurate showed very poor function of the left kidney and diminished function of the right (Fig.

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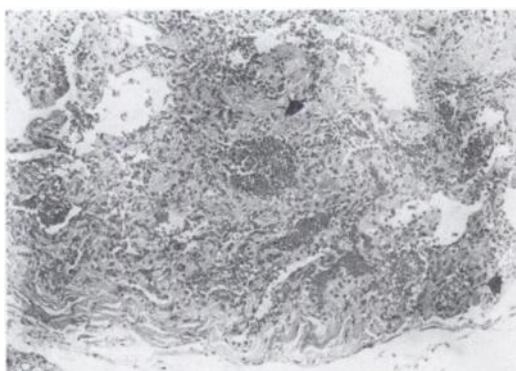
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**FIG. 1.** (A) Renal scan outlines small left kidney with residual uptake. (B) Scan taken with  $^{131}\text{I}$ -Hippuran shows poor function in both kidneys, worse in left.



**FIG. 2.** Lung scan demonstrates multiple defects, both central and peripheral.



**FIG. 3.** Multilayered proliferating endothelial cells penetrate arterial wall, as well as surrounding alveoli, resulting in narrowed lumen (arrows).



**FIG. 4.** Arteriography shows narrowed and beaded renal artery.

1B). Lung scan with  $^{99m}\text{Tc}$ -macroaggregated albumin demonstrated multiple peripheral and central defects (Fig. 2). Ventilation studies with  $^{133}\text{Xe}$  gas showed symmetrical and equal single breath, equilibration, and "washout" phases. Liver and spleen scans with  $^{99m}\text{Tc}$ -sulfur colloid outlined a patchy left lobe of the liver but were otherwise normal.

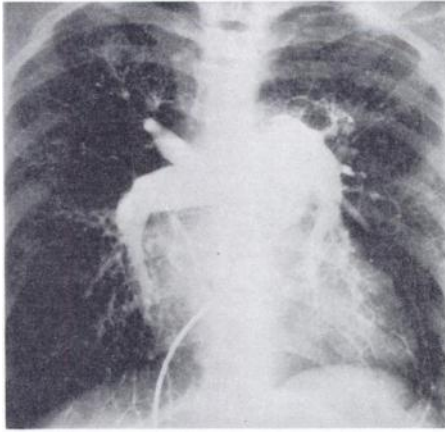
**Pathology.** An open-lung biopsy specimen showed moderate hypertrophy of muscular layers of the pulmonary arteries with restricted lumens in several areas. An area of obstruction of the pulmonary artery by proliferating endothelial cells penetrating the arterial wall and extending into the surrounding alveolar septae was demonstrated (Fig. 3). A diagnosis of hemangioendotheliomatosis with secondary pulmonary hypertensive changes was made.

#### DISCUSSION

Hemangioendothelioma is a very rare neoplasm of vascular origin. It is an intergrade between the more common but benign hemangioma and the relatively uncommon hemangiosarcoma (1). Involvement of the skin and viscera, particularly the liver and spleen, has been well demonstrated in hemangioendotheliomatosis (2). Skin lesions were not present in our patient but the liver scan findings which appear nonspecific are probably compatible with hepatic involvement. Although the more malignant hemangiosarcoma has been noted currently in the liver in increasing numbers of workers exposed to vinyl chloride (3,4), there is at present no known relationship of vinyl chloride to hemangioendotheliomatosis. Our patient had no history of unusual exposure to this chemical substance either.

The multiple abnormal radionuclide dynamic and static imagings in this patient are not specific in any finding to indicate a diagnosis of hemangioendothelioma. Nonetheless, they do indicate a multisystemic vascular disease involving both the pulmonary and systemic circulations. The cerebral flow studies indicating major cerebral and carotid artery defects, the multiple defects on lung scan, the impaired renal functions, and the abnormality on liver scan are all suggestive of widespread arterial occlusive disease. All of these defects were confirmed with radiographic contrast studies except those in the liver. The renal arteriographic studies demonstrated narrowed and beaded renal arteries indistinguishable from fibromuscular hyperplasia (Fig. 4). It is probable that multicentric origins of the hemangioendothelioma produced the multiple arterial lesions. The presence of multilayered atypical endothelial cells extending through arterial walls and into adjacent tissue and venous channels is the histologic finding characteristic of this disease (2).

The lung perfusion scan and pulmonary angiogram are difficult to distinguish from Takayasu's arteritis in the lung (5). The peripheral defects on scanning and the normal  $^{133}\text{Xe}$  ventilation scan are indistinguishable from multiple pulmonary emboli although the central perfusion defects tend to favor



**FIG. 5.** Pulmonary arteriography demonstrates narrowed and sometimes cut-off vessels at various levels.

cystic changes. However, the angiographic findings revealed areas of irregular narrowing of arteries at various levels which were inconsistent with pulmonary embolism and suggested some form of arteritis (Fig. 5).

Radionuclide procedures provided a method for assessment of multiple areas of the circulation with no patient morbidity. It is significant that the radionuclide studies indicated areas of circulatory defects that were more diffuse than those apparent on angiographic study. This may reflect the more "functional" distribution of radiopharmaceuticals that are distributed by the circulation unaltered by injection

artifacts. This is particularly applicable when diffuse types of vascular disease are suspected clinically. Radionuclide scanning can also indicate the most productive areas of selective study with contrast radiography and for biopsy consideration. These studies would also provide a method of following the course of diffuse arterial disease and assessing the possible response to therapy. There is, however, no specific treatment established for hemangioendothelioma at present.

#### ACKNOWLEDGMENT

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