

tures. Radiotherapy has not been of benefit, and the role of corticosteroids is still uncertain.

We believe that there may be an important place for radionuclide imaging in the study of retroperitoneal fibrosis. In our case the location and extent of the disease were neatly delineated in relation to the anatomy of the total abdomen in a single composite, agreeing well with operative findings. Sonography and axial tomography pose problems in spatial reconstruction and conceptualization. Moreover, they are not physiologic, showing only the mass lesions but not the more diffuse inflammatory component. Furthermore, the Ga-67 citrate scan may show additional lesions elsewhere, as might occur with metastatic neoplastic disease. And, finally, the Ga-67 citrate scan may provide a sensitive and convenient tool to monitor the patient for recrudescence of his inflammatory component.

ACKNOWLEDGMENT

We thank Dr. Marvin Gilbert for permission to report his case.

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Brain and Lung Involvement of Mycosis Fungoides Demonstrated by Radionuclide Imaging

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A patient with advanced mycosis fungoides developed neurologic and respiratory symptoms and signs following multiple courses of chemotherapy and radiotherapy. Various repeated diagnostic procedures—including cranial computerized tomography and fiberoptic bronchoscopy with transbronchial lung biopsy—failed to demonstrate an unusual involvement of the brain and lungs by mycosis fungoides. Radionuclide brain imaging and gallium imaging of the lungs demonstrated diffuse lesions confirmed at autopsy.

J Nucl Med 20: 240-242, 1979

Mycosis fungoides, a malignant skin condition with the microscopic appearance of lymphoma, is easily confused with the skin manifestations of leukemia and Hodgkin's disease (1). It may remain localized to the skin for long periods, but in some cases, it may progress to a systemic stage. According to reports in the literature, 14-94% will evolve into lymphoma, and the organs most commonly affected after the transformation are lymph nodes, spleen, liver, lungs, gastrointestinal tract, and adrenal glands (2). This report describes an unusual involvement of the brain and lungs by mycosis fungoides, documented by radionuclide imaging.

Received Sept. 11, 1978; revision accepted Oct. 10, 1978.

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CASE HISTORY

Five years before this hospital admission, a 55-year-old man developed pruritic erythematous skin plaques of the left arm and was treated with radiation therapy and topical nitrogen mustard, then and again 2 yr later. Three years after the onset of the skin lesions, diagnosis of mycosis fungoides was established by skin biopsy, and he received five courses of MOPP therapy (methotrexate, oncovin, procarbazine, and prednisolone). Since then he had received multiple courses of chemotherapy and four courses of total or partial-body electron-beam therapy for advanced skin lesions. One week before this admission he noted an increased productive cough (without hemoptysis) and leg weakness, and several days later he developed dizziness and violent jerking of the right leg that lasted about 10 min.

Physical examination revealed diffuse erythematous macules, indurated papules of varying size, and confluent papules forming large plaques on all extremities, trunk, and face. There were

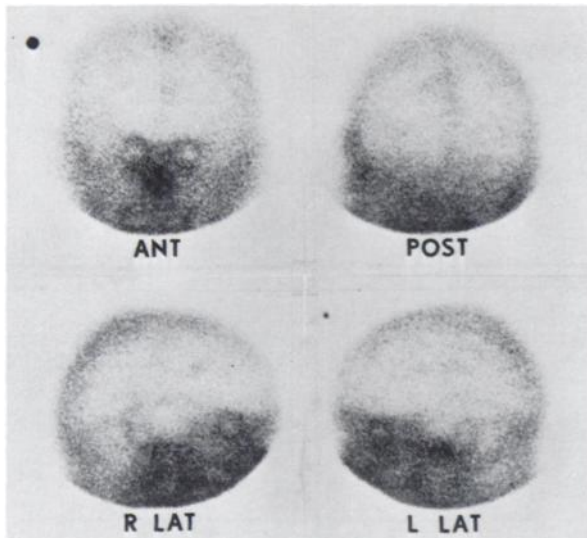


Fig. 1. Brain imaging with Tc-99m DTPA shows focal abnormal activity in right frontal lobe and ill-defined areas of activity in right and left parietal regions.

crepitant rales in both lung bases. Neurologic examination revealed slightly decreased motor and sensory function of the right leg.

Chest roentgenogram showed minimal infiltration in both lung bases. Multiple sputum examinations were negative for microorganisms, fungi, and cytology. Bronchoscopic examination also revealed no abnormality. Electroencephalography showed grade II generalized dysrhythmia. Slightly elevated cerebrospinal-fluid protein (58–73 mg/dl) was found on three examinations, but bacteriologic and serologic tests were negative.

On the day following admission, radionuclide brain studies with Tc-99m DTPA showed an ill-defined peripheral concentration of radioactivity in the left parietal region.

Five days after admission, the patient experienced sudden weakness of the left arm, and computerized transmission tomography (CTT) showed an ill-defined area of decreased density in the left parietal lobe, interpreted as possible vasculitis, cerebritis, or embolism. Repeat radionuclide studies two days later demonstrated the ill-defined lesion in the left parietal region and also a well-defined focus of radioactivity in the right frontal lobe and ill-defined focal activity in the right parietal region (Fig. 1). On the following day a second CTT revealed ill-defined areas of low density in both hemispheres and a diffuse demyelination was considered most likely.

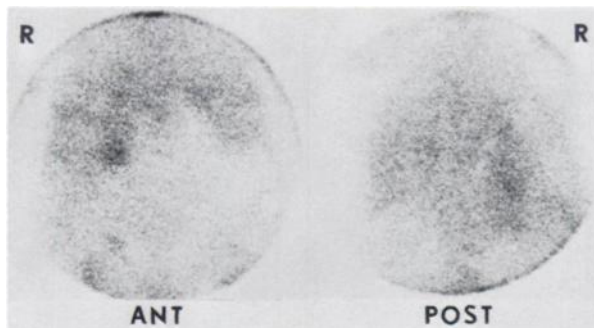


Fig. 2: Gallium-67 imaging demonstrates diffuse abnormal activity in both lungs and a focal increased activity in right lung base.

Two weeks after hospital admission the patient suffered from mental deterioration, increasing somnolence, and nocturnal fevers as high as 103°F (39°C). Gallium-67 citrate imaging (to rule out an occult abscess) showed diffuse abnormal activity in both lungs and a focal area of increased activity in the base of the right lung (Fig. 2). Transbronchial biopsy of the right lung base revealed nonspecific interstitial fibrosis. Liver biopsy was normal and bone-marrow biopsy revealed atypical histiocytic proliferation.

The patient continued to deteriorate clinically, with increasing confusion. About 1 wk before death, repeat radionuclide and CTT studies demonstrated no significant differences from the previous second studies. Necropsy sections showed multiple foci of malignant lymphomatous cells in the lungs and in all lobes of the brain. The cells were predominantly perivascular, but direct invasion of brain tissue was also observed. No microorganisms were found, and there were no acute or chronic inflammatory reactions.

DISCUSSION

Mycosis fungoides, a disease whose exact nature and place in nosology remains controversial, is generally accepted as a disease of the lymphoreticuloendothelial system. It was first described in 1806 by Alibert, who initially termed the disorder *pian fungoïde* because it resembled yaws. Traditionally, it has been divided into three stages: first stage, erythematous premalignant; second stage, plaque; and the third or tumor stage, which is the terminal phase in the evolution of the cutaneous lesions. When the internal organs are involved, the pathologic picture may be that of lymphosarcoma, reticulum-cell sarcoma, Hodgkin's disease, or leukemia (2). The reported wide range of incidence of its transformation into a lymphoma is probably due to the confusion from pathologic interpretation. According to some, it is likely that the pure cutaneous form of mycosis fungoides can convert to a lymphoma 33% of the time (2).

There are no consistent diagnostic laboratory findings, but the distortion or destruction of nodal architecture observed on lymphangiography suggests a transformation of mycosis fungoides into a malignant lymphoma. Radionuclide brain imaging has been an effective method for the study of the central nervous-system lesions, with an overall detection rate of 83–90% (3,4). Data from the literature indicate that the radionuclide studies are slightly less sensitive than CTT for the detection of metastatic disease (5), although Baker et al. (6) have reported an 8.5% false-negative rate for CTT in patients with metastatic cerebral lesions. In our case, CTT demonstrated ill-defined areas of low density in both hemispheres; such areas were considered probably due to diffuse demyelination, whereas radionuclide studies showed multiple focal concentrations of radioactivity consistent with metastatic lesions. However, one could not totally exclude demyelinating cerebral disease or multifocal leukoencephalopathy occurring in a patient with an abnormal immune system.

Gallium-67 citrate imaging has been helpful in the evaluation of lymphomas, with overall true-positive and true-negative rates of 53 and 90%, respectively (7). On the other hand it is clear that the abnormal accumulation of Ga-67 is nonspecific and occurs in non-neoplastic lesions. As was present in our case, diffuse radiogallium concentration in both lungs, with minimal or absent radiographic changes, has been reported in pneumoconiosis (8), disseminated erythematous (8), pneumocystis carinii pneumonia (9), bleomycin toxicity (10), and acute radiation pneumonitis (11).

Our case demonstrates the relatively unusual combination of abnormal concentration of Ga-67 in both lungs, due to the involvement of lymphoma (mycosis fungoides), and positive findings in the brain by radionuclide imaging (Tc-99m DTPA).

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