Coexistent Primary Lymphosarcoma of the Lung Incidentally Present for Five Years in a Patient Treated for Achalasia

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The case to be reported is unique in that while the patient was being observed and treated over a period of eight years for cardiospasm and achalasia of the esophagus, a primary lymphosarcoma of the lung developed insidiously. The tumor was first noted on Roentgen examination five years prior to the patient's demise, and for two years the first changes noted on repeated x ray's of the chest were not considered neoplastic.

Since the prognosis of lymphomas of the lung is much better than carcinoma, early diagnosis and treatment is essential. So often a second disease will develop so insidiously that it is not appreciated and goes unrecognized, as illustrated in our case. Most malignant lymphomas of the lung, when clinically diagnosed, have already disseminated. Since this type of tumor remains localized for long periods and spreads slowly by direct extension, they therefore can be surgically excised, and there are such case reports in the literature. The case in point will illustrates the localization of a primary lymphoma of the lung over a long period before it spread.

CASE REPORT

This 64 year old Negro female when first seen complained of difficulty in swallowing, indigestion, and weight loss of 15 years' duration. By gulping hard and drinking large amounts of water she was able to force food down. Her weight dropped from 227 pounds to 120 pounds. Her blood pressure was 110/60, pulse 72, respirations 16. The physical examination was essentially negative. The clinical impression was: Stenosis of the esophagus.

A chest x ray was reported negative. An upper GI series revealed a marked dilatation of the esophagus down to the cardia. The obstruction area at the cardia was smooth, cone shaped, and permitted a narrow stream of barium into the stomach. The conclusion was: Car-
diaphragm. She was taken to surgery and a partial gastrectomy was done. The pathological examination of the stomach revealed hypertrophic gastritis. Her post-operative course was uneventful.

Subsequently she continued to experience difficulty in swallowing, and no symptoms referable to her lung. X-rays continued to show dilatation of the esophagus with retained food and negative chest. She was again admitted to the hospital because of shortness of breath, orthopnea, coughing, and chest pains. A diagnosis of aspiration pneumonia was made. Her temperature was 103.6°F.

Review of clinical laboratory findings: Unanalysis negative. Serology was negative. Blood: Hgb. 13.3 grams, WBC 7,200; polys. 65%, lymphs 28%, 2% eosinophils' 5% monocytes. Total protein was 9 g. %; albumin 5.2 g.%; globulin 3.8g%; AG ratio 1.4:1. Sputum was negative for acid fast bacilli.

An x ray of the chest taken five years before she expired revealed increased density of the right midlung field, (See fig.1). This was the first appearance of the pulmonary infiltrate. Two months later the process in the lung appeared bilaterally, (See fig.2).

Two years later a chest x ray showed patchy areas of increased density projected in both lower lung fields-somewhat greater on the right side. The possibility that these areas might represent aspiration changes in the lung was strongly considered, although their position was somewhat unusual for this condition. Three months later, an x ray of the chest revealed considerable infiltration in both lung bases. The findings on the right were essentially the same as previously noted, although there had been considerable progression on the left side since that date with evidence now of pleural effusion. It appeared that there was considerable para-mediastinal widening on the right side with increased prominence of the left hilum. These findings were thought to be related to a mediastinitis. Subsequent x ray films of the chest during the next two years showed changes similar to that noted previously. One year later, chest x ray revealed extensive infiltration throughout the left lung. The last film of the chest one month later or five years after the onset (See fig.3) indicated a marked increase in the density of the right lung base with increased markings in the left lower lung field. The radiologist’s opinion was that carcinoma of the lung could not be excluded.

During her last admission, because of the continued difficulty in her swallowing and continued weight loss a Jejunostomy was performed. Two months later an attempt was made to repair a hiatus hernia. Patient bled quite profusely and expired shortly thereafter. This was approximately eight years after the onset of her acute symptoms.
NECROPSY FINDINGS

The body was that of an emaciated Negro female, weighing approximately 90 pounds. In the left upper quadrant, there was a Jejunostomy opening and a healed surgical scar to the right of it. There was also a recent surgical wound 22.0 cm. in length present in the midline.

The abdominal cavity contained about 500cc. of bright blood. The source of the bleeding was from a tear in the inferior vena cava. The pleural cavities contained numerous abhesions. The heart weighed 160 grams, and no unusual changes were noted.

The upper two-thirds of the esophagus was dilated up to 40.0mm. in circumference and the wall was moderately thickened. The lower one-third was narrowed up to 20.0mm. in circumference. The mucosa was injected. The stomach revealed a functioning gastrojejunalostomy, and in the wall there were two discreet nodules up to 50. mm., which on microscopic examination were benign leiomyomata.

LUNGS:

The right lung weighed 500 grams. The pleura was thickened and in the region of the apex of the upper lobe there were several emphysematous blebs up to 5.0cm. in diameter. The upper lobe felt crepitant, while the middle and lower lobes were firm. On sectioning, the surfaces of the middle and lower lobes appeared consolidated, greyish-brown, moist and glistening (See fig.4). The middle and lower bronchi were narrowed; the mucosa was pale.

The left lung weighed 380 grams. It showed blebs on the surface and was firm in consistency. The surface, made by sectioning, was uniformly greyish-brown. There were several small peribronchial lymph-onges, which on section appeared anthracotic. The remaining on section appeared anthracotic.

The remaining organs showed no unusual changes.

MICROSCOPIC EXAMINATION:

Multiple sections made from different sites of the tumor masses from the left and right lungs revealed the typical appearance of a lymphoma of the lymphocytic type. The tumor was composed of solid sheets of lymphocytic-like cells, containing oval, hyperchromatic nuclei, occupying most of the cell and little cytoplasm present (See fig.5). The tumor cells occasionally surrounded blood vessels (See fig.6). Mitotic figures were inconspicuous. There were no germinal centers noted nor evidence of necrosis. At the periphery of the tumor the alveoli appeared compressed by a fibrillar connective tissue. The blood vessels were unchanged, and some of the bronchi showed a squamous metaplasia of the lining epithelium.

The peribronchial lymph nodes contained no neoplastic cells. Sections from the vertebral bodies showed no tumor.
DISCUSSION

A primary malignant lymphoma, lymphocytic type, in the lung or co-called lympho-sarcoma, implies that the tumor is restricted to the lung and no other sites are involved. The pulmonary hilar lymph nodes may or may not be involved. This type of tumor must be differentiated from the inflammatory pseudo-lymphomas. In the latter, microscopic sections usually show the presence of lymph follicles and areas of necrosis, which are absent in primary malignant lymphomas. Our case illustrated, as others do, how these tumors merge with the surrounding parenchyma and without interposition of a capsule. The tumor may be large, solitary or produce large bulky masses. Early diagnosis has resulted in surgical excision and five year survival. However, many cases go unrecognized and are disseminated in the lungs by the time they are diagnosed clinically, such was our case. It is interesting to note that primary lymphomas of the lung are no different in their development from those that are primary in the lymph nodes. Dr. Averill Liebow in reviewing the case and the slides stated that "everything about the lesion, including the light brown color observed grossly is certainly compatible with a primary lymphoma of the lung".

SUMMARY

A primary malignant lymphoma of the lung was present clinically and noted on repeated x-ray examination for almost five years in a 64 year old Negro female while treated for Achalasia at the esophagus. Early recognition of malignant lymphoma of the lung has a better prognosis when treated surgically with or without radiation than carcinoma. To have been able to observe the natural history in the development and duration of a malignant lymphoma in the lung for five years is most unique.

REFERENCES

2. Liebow, A. A. : Atlas of Tumor Pathology A. F. I. p., Section V, Fascicle 17: Fig. 18–135, Fig.17–138.
Fig 1. First appearance of tumor appearing as an infiltrate in the right m.d. lung. Note cardiospasm in esophagus.
Fig. 2. Four years after first appearance of neoplasm. There are large patches of consolidation somewhat symmetrically located in both lung fields.
Fig. 3. Five years later—There is further extension of the infiltrative process on the right and extensive honey-combing on the left.
Fig. 4. Photograph show the large diffuse tumor mass in the right middle lobe. Note grayish brown color.
Fig. 5. Low power shows the lymphocytic character of the tumor (H and E x460).
Fig. 6. High power shows tumor cells frequently surrounding blood vessels (H and E × 400)