Lymphangioma of the Mediastinum

Report of Two Cases

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Lymphangiomas, or hygromas, confined to the mediastinum are rare. Blades\(^1\) in 1946 reviewed 109 cases of mediastinal tumors, collected from various Army Thoracic Centers. In this group of patients there were no instances of lymphangioma. In 1947 Watson and Diamond\(^14\) found only one in Navy personnel. Gross and Hurwitt\(^5\) made a thorough review of the literature in 1948 and were able to find but seven reported cases. To this they added one patient. Since then there have been scattered reports of individual cases.\(^15\) Recently Childress and associates\(^3\) found but seventeen cases in which there had been operative intervention, theirs bringing the number to eighteen. We consider it pertinent, therefore, to present two patients with mediastinal lymphangioma treated by surgical excision at the George Washington University Hospital without recurrence.

Case Presentations

Case 1: R. B., a 58 year old white man, was admitted to George Washington University Hospital on April 25, 1953. Symptoms began about four months previously and consisted of hemoptysis, severe cough, and interscapular pain. For many years the patient had had a chronic cough productive of mucus.

On admission to the hospital he appeared to be well nourished and in good general health. There were respiratory wheezes audible over the right upper lobe but no other significant abnormal findings were present.

Laboratory studies revealed a normal hemogram and urinalysis. Serologic test for syphilis was negative.

Roentgenograms of the chest revealed a 5 cm. right mediastinal tumor obscuring the right hilum (Fig. 1). The remainder of the chest was not remarkable.

On April 27, 1953 right thoracotomy was performed under ether anesthesia. The pleural space was free. A cystic tumor was located in the right anterior superior mediastinum closely adherent to the anterior surface of the superior vena cava and extended over the arch of the aorta. Palpation of the right lung revealed no pulmonary tumor. By patient dissection the tumor mass was separated from the structures to which it was adherent.

Since discharge from the hospital the patient has remained well. The latest roentgenogram of the chest did not show any recurrence of the tumor.

Pathologic examination of the specimen revealed a spherical mass measuring 5 cm. in diameter and covered by a thin, adherent, transparent capsule. On cut section the tumor tissue was pinkish-yellow and lobulated. In the center were several cavities, the largest measuring 4 x 2 x 1.5 cm., filled with sanguinous, serous fluid. The smaller cavities contained milky fluid. Microscopic examination of the tumor revealed medium sized, oval cells, having pale, vesicular nuclei. In places, the tumor cells were arranged in solid sheets. Elsewhere there was a honeycomb appearance with the formation of numerous vascular spaces filled with serous fluid. There was a distinct fibrous capsule beneath which was a thin layer of lymphocytes (Figs. 2 and 3). Outside the capsule were several lymph vessels showing hyperplasia of the lining endothelium.

Case 2: N. L. W., a 19 year old white woman was admitted to George Washington University Hospital on June 26, 1955. For several years she had been followed by her physician with periodic chest roentgenograms for a lesion in the right hemithorax. The patient had had no symptoms referable to pulmonary disorder.

On admission to the hospital she appeared to be well nourished and in good, general

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health. There were no significant abnormal findings present.

Laboratory studies revealed a normal hemogram and urinalysis showed a trace of protein. Serologic test for syphilis was negative.

Roentgenograms of the chest revealed a 4 cm. density at the right cardiophrenic angle anteriorly (Fig. 4). The remainder of the chest was considered normal. The impression of roentgenologist at that time was a cyst in pericardium.

On June 27, 1955 thoracotomy was performed under ether anesthesia. At the right cardiophrenic angle there was a 5 cm. grayish-black multi-loculated cystic lesion intimately attached to the pericardium. Two large vessels entered the lesion. By sharp dissection the lesion was removed.

After operation she was given penicillin and streptomycin and she remained afebrile. On the sixth day after operation she developed herpes simplex which cleared with multi-vitamin therapy.

Since discharge from the hospital she has remained well. Interval roentgenograms of the chest do not show recurrence of the lesion.

Pathologic examination of the specimen revealed an encapsulated mass measuring 4.5 cm. in diameter. Cut surface was pinkish-gray and finely trabeculated. Microscopic examination showed the tumor to be well encapsulated, and composed of an intimate admixture of dilated endothelial-lined thin-walled channels, and lesser numbers of small muscular arteries with marked thickening of their media. Small focal collection of lymphocytes and plasma cells lay adjacent to some of the vessels. Some of these channels were filled with blood, while others contained an acellular eosinophilic exudate (lymph). There were a few collagen fibers interspersed between fascicles of smooth muscle fibers in the more solid portions of the tumor (Fig. 5).

FIGURE 1: Roentgenogram of chest prior to surgery showing a right hilar density.
Discussion

Cystic lymphangiomas are congenital benign tumors of mesodermal origin. They can occupy the superior or inferior mediastinum; most frequently lying anteriorly and presenting into the right chest. Externally, the mass is grayish-white, shaggy and has ill-defined boundaries. It is adherent to neighboring structures, frequently giving the impression that these structures traverse the tumor mass. Both unilocular and multilocular types have been noted, the spaces of which are lined by flattened endothelial cells. Within the cyst wall can be found smooth muscle, connective tissue, fat, nerves, and blood vessels. This is best explained by Goetsch. He has shown that the lining cells of the cyst give off endothelial sprouts which envelop these elements in the formation of new cysts.

Mode of origin of cystic hygromas is not certain. The most probable theory is that they arise from portions of the primitive jugular lymph sacs which become separated and fail to re-establish communication with the venous channels. If it becomes adherent to structures destined for the mediastinum, such as the pericardium and diaphragm, the mass will then be drawn into the mediastinum with the descent of these structures during fetal life.

Clinical

Mediastinal lymphangioma is a relatively silent disorder which probably accounts for diagnosis late in years in some patients. Symptoms, if they occur, point to some intrathoracic disturbance. This is most frequently caused by pressure on the great vessels, nerves, bronchi, or esoph-

FIGURE 2A

FIGURE 2B

Figure 2: Photograph of microscopic section showing dense aggregates of lymphocytes and endothelial cells.
agus. Chylothorax was reported in one patient and in another there was hoarseness due to paralysis of the left vocal cord. In the two patients presented here, one was symptomatic complaining of pain, cough, hemoptysis. The lesion was discovered by routine roentgenogram in the second patient. With the common practice of roentgenogram survey it is felt that these lesions will be found more frequently.

Treatment

Various methods of treating these lesions have been employed. Chemical sclerosing agents have been utilized when the lesion could not be removed in its entirety. Roentgen therapy was used in one patient with remission leading to an erroneous impression of lymphoma. Skinner and Hobbs performed a two stage excision.

All agree now that thoracotomy is indicated once the lesion is recognized. This allows a tissue diagnosis to be made and the lesion can be

FIGURE 3: Photograph of microscopic section showing dilated lymph channels in tumor.
extirpated. Because of the nature of the lesion, it is not always possible to remove the mass in its entirety.

In our patients the lesions were found by roentgenograms, however, the diagnosis was not established until operation. At that time the lesions were removed, a tissue diagnosis was made, and to this date there has not been a recurrence demonstrable by roentgenogram or suggested by symptoms.

CONCLUSIONS

1. Lymphangiomas confined to the mediastinum are rare.
2. Mediastinal lymphangiomas are probably due to development defects of the primitive jugular sacs.
3. The mass is closely adherent to surrounding structures and may contain smooth muscle, fat, nerves, blood vessels, and connective tissue.
4. Mediastinal lymphangiomas, being a rather symptomless disease, are usually discovered by roentgenograms.

FIGURE 4: Roentgenogram of chest prior to surgery showing a density at the right cardiophrenic angle.
5. Exploratory thoracotomy is indicated once the lesion is found to establish a tissue diagnosis and to extirpate the mass.

CONCLUSIONES

1. Los linfangiomas situados en el mediastino son raros.
2. Los linfangiomas mediastinales son debidos probablemente a defectos de desarrollo en los sacos yugulares primitivos.
3. La neoformación está intimamente adherida a las estructuras circundantes y puede contener músculo liso, grasa, nervios, vasos sanguíneos, y tejido conectivo.
4. Los linfangiomas mediastinales, siendo un padecimiento asintomático, se descubren habitualmente por los roentgenogramas.
5. La toracotomía exploradora está indicada una vez que la lesión se ha descubierto para establecer el diagnóstico y extirpar la masa.
6. Se presentan dos casos de linfangioma mediastinal lo eleva el número de casos relatados a veinte.

FIGURE 5: Photograph of microscopic section showing dilated lymph channels with surrounding aggregates of lymphocytes.
LYMPHANGIOMA OF THE MEDIASTINUM

SCHLUSSFOLGERUNGEN

1. Auf das Mediastinum beschränkte Lymphangiome sind ein seltenes Vorkommnis.


3. Die Tumormasse hängt eng mit den umgebenen Strukturen zusammen und kann glatte Muskulatur, Fett, Nerven, Blutgefäße und Bindegewebe enthalten.

4. Gewöhnlich werden die mediastinalen Lymphangiome, die eine recht symptomlose Krankheit darstellen, durch Röntgenaufnahmen entdeckt.

5. Eine diagnostische Thorakotomie ist angezeigt, sobald die Veränderung festgestellt ist, um eine Gewebsdiagnose zu erlangen und Masse zu extirpieren.

6. Es werden 2 Kranke mit mediastinalen Lymphangiomen vorgestellt, womit sich die Zahl der bekanntgegebenen Fälle auf 20 erhöht.

REFERENCES