

## GIANT MEDIASTINAL CYSTIC HYGROMA IN A CHILD

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Lymphangioma is a benign and common disorder in the pediatric age group. Cervical cystic hygroma was first described clinically in 1828 by Redenbacker and pathologically in 1843 by Wernher.<sup>1</sup> They are frequently seen in the neck (75%) and axilla (20%).<sup>2</sup> Most mediastinal cystic hygromas are extensions of cervical lesions, and cystic hygroma confined solely to the mediastinum is rarely encountered.<sup>3,4</sup> We report a case of a female child with a purely extensive mediastinal cystic hygroma, outlining aspects of its unusual clinical features and management.

### Case Report

A three-year-old female was referred to our hospital because of chest pain and bulging of the right side of the chest of two weeks' duration. The parents gave a history of weight loss, but there was no fever. On examination, she was found to be pale, but not jaundiced or cyanotic. The right side of the chest was found to bulge, in comparison with the left. There was decreased air entry on the right side of the chest, with dullness on percussion. Chest x-ray revealed a right-sided pleural effusion. Thoracentesis yielded serosanguinous fluid, which contained gelatinous material. Biochemical and cytological analysis of the fluid gave the following results: glucose 90 mg/dL, total protein 4.6 g/dL, LDH 115 U/L and 8200 WBC U/L, predominantly lymphocytes and many RBCs. Cytology of the aspirated fluid revealed no malignant cells. Bacterial, fungal and mycobacterial cultures showed no growth. PPD was negative.

A right-sided thoracostomy tube was inserted and postdrainage chest x-ray (Figure 1) revealed a large right-sided mass. HCG was normal and serum BHCG was <5. Ultrasonography of the chest could not be done because of the lack of a proper transducer. CT scan of chest (Figure 2) showed a large right-sided lobulated mass

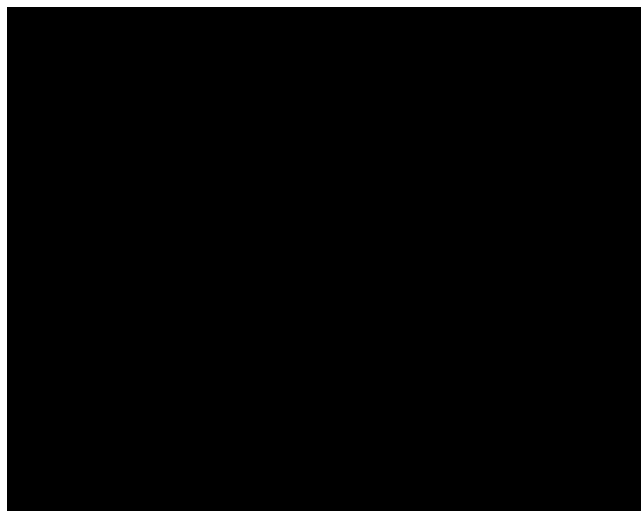


FIGURE 1. Chest x-ray showing a large right-sided mediastinal mass.

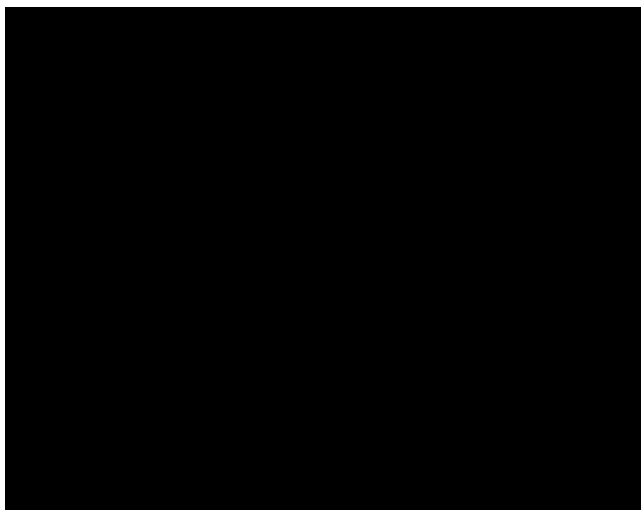


FIGURE 2. CT scan of the chest showing a large diffuse mass in the anterior mediastinum. Note the relation of the mass to the pericardium.

occupying the entire anterior mediastinum. The left margins of the mass were inseparable from the pericardial surface of the heart, which was displaced to the left. The chest was opened via a right anterolateral thoracotomy through the bed of the 5th rib. There was a large tumor which was fixed and infiltrating into the anterior chest

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FIGURE 3. Cystic lymphatic space lined by attenuated endothelium (hematoxylin and eosin, 100x).



FIGURE 4. High-power view showing edematous stroma containing spindle and stellate cells with mild nuclear pleomorphism and no mitosis (hematoxylin and eosin, 400x).

wall. The posterior part of the tumor was firm, but the anterior part was friable and soft. There was a possibility of teratoma or lymphoma, and a large biopsy was taken. Histology revealed a vascular tumor made up of various sizes of vascular channels lined by endothelium (Figure 3). The stroma was edematous and myxoid, containing spindle and stellate cells with mild nuclear pleomorphism and no mitosis (Figure 4). The stroma contained a diffuse infiltrate of lymphonuclear cells and neutrophils. A fibrous capsule was seen with a few thick fibrous septa. The diagnosis of cystic lymphangioma was made. The patient continued to be symptomatic and so the chest was re-explored and near complete excision of the mass was done, leaving only small fragments that were densely adherent to the hilum of the lung. The cysts near the hilum of the lung and adherent to the pulmonary vessels were deroofed and as much of the capsule as possible was excised. Postoperatively, the patient did well and a chest radiograph showed clear lung fields.

### Discussion

Embryologically, the lymphatic system is formed by the eighth week of intrauterine life and arises from six primitive sacs: paired jugular sacs which are located lateral to the jugular veins, an unpaired retroperitoneal sac at the root of the mesentery, an unpaired cisterna chyli dorsal to the aorta and adrenal glands and paired posterior sacs located in relation to the sciatic veins.<sup>5</sup> Cystic hygroma is a benign tumor that results from developmental malformation of the lymphatic system. It most frequently occurs in the cervical location (75%), which probably represents a developmental malformation of the jugular lymphatic sacs or their connecting vessels, followed by the axilla (20%).<sup>2</sup> Most mediastinal cystic hygromas are

usually extensions of cervical hygromas, as 2% to 3% of cervical hygromas have mediastinal extensions.<sup>2</sup> Isolated intrathoracic cystic hygroma is rare, and if encountered, is usually located in the anterior or occasionally in the middle mediastinum.<sup>3,4,6,7</sup> Posterior mediastinal location of cystic hygroma is even rarer.<sup>6</sup> The embryological basis of mediastinal cystic hygroma is not clearly understood. Curley et al.<sup>6</sup> postulated that posterior mediastinal cystic hygroma arise from embryological malformation of the cisterna chyli or its cephalad outbudding.

Lymphangiomas are slow-growing tumors, and in the head and neck usually present as an obvious swelling, which is classically cystic and can be transilluminated. Since 2% to 3% of cervical lymphangiomas have intrathoracic extension, a chest x-ray is recommended in these patients.<sup>7</sup> Mediastinal lymphangiomas, on the other hand, tend to involve neighboring structures such as large blood vessels, the heart, trachea and esophagus. Our patient presented with a bulging right side of the chest, which was due to infiltration of the lymphangioma and pleural effusion, perhaps due to rupture of the lymphangioma, or as a result of super-added infection. Although the radiographic workup of these patients includes a chest x-ray, a real-time ultrasonography and CT scan of the chest, we found CT scan to be of great value in delineating the tumor and its attachments to the neighboring structures.

The treatment of lymphangioma should be complete surgical excision, as these tumors do not disappear on their own. Recently, bleomycin and OK-432 sclerosing therapy have been used successfully to treat lymphangiomas in children.<sup>8,9</sup> In mediastinal lymphangioma, parts of the tumor may be insinuated between or adherent to neighboring vital structures, which makes complete excision hazardous, as in our patient. We agree with other

authorities who recommend deroofing of the cysts with resection of the maximum amount of cyst wall in these patients rather than risking injury to vital structures.

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