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Mediastinal Cystic Hygroma in Children

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Abstract. A 14-month-old child presented with severe respiratory distress from an anterior mediastinal mass. Since its gross appearance suggested malignancy, radical surgical excision was performed. In retrospect, secondary hemorrhage, fibrosis, and chronic inflammation were responsible for the atypical appearance of a benign cystic hygroma. Etiology, clinical presentations, diagnosis and recommended treatment of this relatively rare mediastinal lesion are discussed.

Key words: Cystic hygroma – Lymphangioma – Ultrasonography – Childhood mediastinal tumors

Isolated mediastinal cystic hygroma is a rare condition with only 24 cases reported in children. It represents a distinct clinical entity, often difficult to diagnose preoperatively, which should be distinquished from the more common and easily diagnosed cervicomediastinal cystic hygroma. Failure to diagnose mediastinal cystic hygromas may be fatal due to their increasing size producing acute respiratory embarrassment.

Case Report

A 14-month-old boy was admitted with fever and grunting respirations. Auscultation revealed decreased breath sounds over the left chest with left lung dullness to percussion; respiratory rate was 60 per minute. Chest roentgenograms (Fig. 1) showed a large anterior mediastinal mass obscuring the left heart border and displacing cardiac-mediastinal structures from left to right. Conventional static sonic scanning (Fig. 2) revealed the mass to be heterogeneous with varying size cystic and solid areas; real-time sonography demonstrated its proximity to the pericardium and great vessels. Preoperative impression was mediastinal teratoma.

Extended left thoracotomy disclosed a large lobulated, cystic and solid anterior mediastinal mass occupying the left chest from the apex to the diaphragm. It was intimately attached to the pericardium and aortic arch with extension between the left subclavian and left carotid arteries; the tumor surrounded the left recurrent laryngeal, vagus, and phrenic nerves. Due to its gross appearance resembling a malignant teratoma or neuroblastoma, a pericardial window and sacrifice of the left phrenic and recurrent laryngeal nerves were deemed necessary. Subsequent microscopic examination of the excised specimen was consistent with a benign cystic hygroma (lymphangioma) which contained solid elements and adhesions secondary to "considerable fibrosis, chronic inflammation and hemorrhage." Initial post-operative course was uneventful. Approximately one month later, the patient's mother noted a soft tissue swelling over the left scapula. Ultrasonography of the suprascapular mass was identical to the previous anterior mediastinal cystic hygroma scan appearance. Surgical excision revealed a thin-walled cystic posterior mediastinal mass under the trapezius muscle which was not contiguous with the original anterior mediastinal mass. Pathological diagnosis was benign cystic hygroma containing hemorrhage, presumably representing a separate tumor. Subsequent post-operative course has been unremarkable except for left diaphragmatic elevation and a weak slightly hoarse voice secondary to left phrenic and recurrent laryngeal nerve sacrifice respectively.

Discussion

Cystic hygroma confined to the mediastinum is rare. Cystic hygromas are usually located in the neck and have a mediastinal extension in about 10% of reported patients. Less than 1% are located only in the mediastinum. 75% of purely mediastinal cystic hygromas have been reported in adults with less than 5% in infants under age 1 year [1]. Bratu et al. in 1970 [1] surveyed the literature and discovered only 14 pediatric cases; our review discloses 10 additional cases with our case bringing the total to 24 cases.

Cervicomediastinal hygromas are usually easily recognized by physical examination supplemented by chest roentgenograms. Diagnosing mediastinal cystic hygromas is difficult until they become large and pro-

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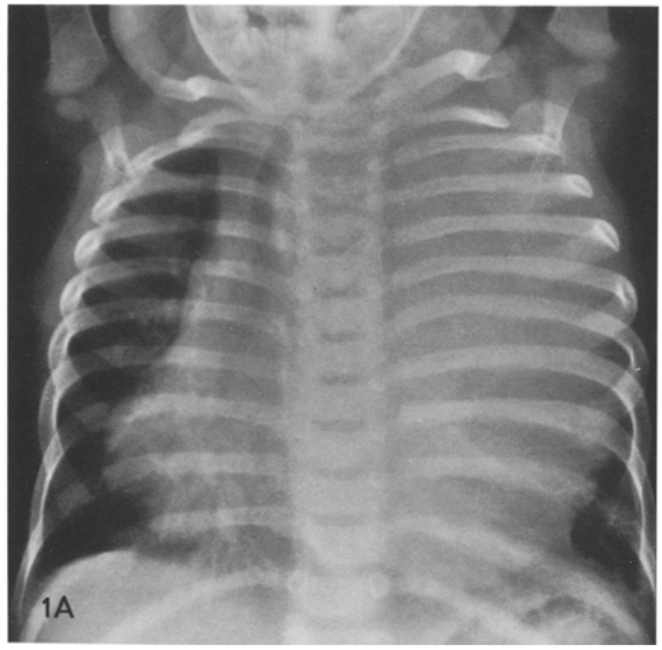

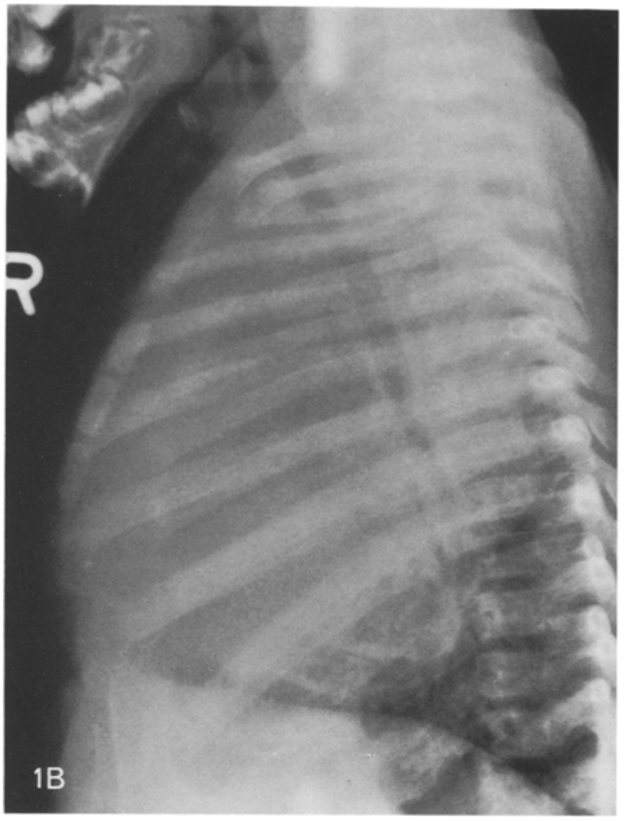

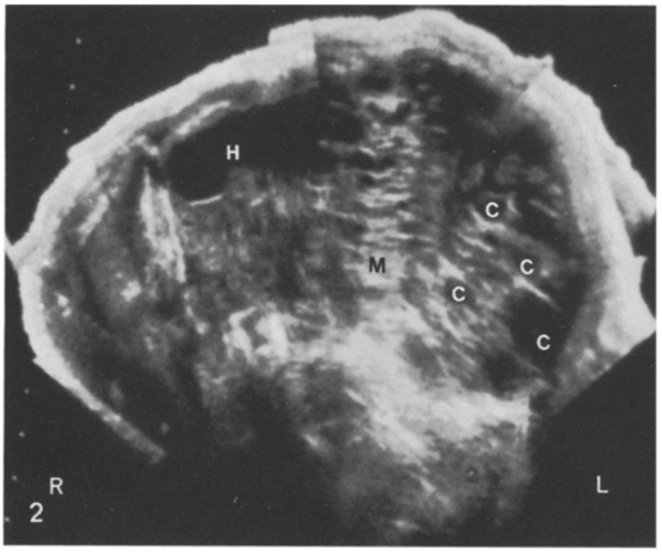

Fig. 1. A Frontal chest roentgenogram showing left chest mass displacing cardiac and mediastinal structures to the right. **B** Lateral chest roentgram demonstrating mass to occupy anterior mediastinum

Fig. 2. Transverse supine thoracic sonogram showing predominantly cystic heterogeneous mass occupying left hemithorax. (H = heart, M = mass, C = cysts, R = right, L = left)

duce pressure manifestations upon surrounding organs or tissues or if hemorrhage or infection develops within the cyst. Symptoms are also age-related in that most children under age 2 years are symptomatic with 50% of those over age 2 years asymptomatic [1]. Respiratory distress characterizes affected children under age 2 years [4], failure to establish the correct diagnosis potentially can result in asphyxia.

The radiographic appearance of mediastinal cystic hygromas is not specific. Usually the mediastinal mass has a smooth sharp border and uniform density [2], but it may be lobulated [3]. Tracheal indentation and/or displacement is common as with other mediastinal masses; calcification within the mass is extremely rare. Employing newer non-invasive imaging modalities facilitates diagnosis in affected children. Contact B-scan sonography is helpful in evaluating mediastinal masses abutting the anterior chest wall, as in our case, in that the predominantly cystic structure is demonstrable. Real-time sonography can delineate the relationship of the mass to the heart and great vessels. Computed tomography theoretically should reveal fluid content. Echocardiography and/or isotopic angiocardiography may also be employed if cardiac or pericardial involve-

ment is suspected. Thoracic aortography and superior venacavography have been employed in adults [2] but these procedures seldom are necessary in children.

Surgical excision is the recommended treatment. Usually a cystic hygroma is cystic, thin-walled, and only rarely contains solid lymphoid elements. Our case differs in that grossly the tumor appeared relatively solid with invasion of pericardium and encasement of adjacent mediastinal vessels and nerves. We wish to emphasize that this atypical gross appearance may occur due to inflammation and hemorrhage within a cystic hygroma; recognition of this possibility can obviate unnecessary sacrifice of entrapped nerves. If diffuse involvement precludes total resection, unroofing with maximum cyst wall removal is advised to deter post-surgical recurrence [3]. Irradiation rarely produces regression and sclerosing agents have not proved effective.

References

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