Occurrence of Hernia of Morgagni with Filial Cervical Lung Hernia: A Hereditary Defect of the Cervical Mesenchyme

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A cervical hernia of the lung of probable congenital etiology is reported in the daughter of a woman with a diaphragmatic hernia of Morgagni. Both of these hernias result from defects in tissues derived from the cervical mesenchyme. The coexistence of these unusual hernias in mother and daughter raises the possibility of a hereditary deficiency of the cervical mesenchyme as an etiologic factor in both of these hernias.

Cervical hernias of the lung and diaphragmatic hernias through the foramen of Morgagni are uncommon entities. Both of these hernias occur as the result of defects in the enclosing structures of the thoracic cavity. Although the respective sites of herniation are anatomically separate in the adult, the tissues involved have a common embryologic origin in the cervical mesenchyme.

Herein described is the first reported occurrence of a congenital cervical lung hernia in the daughter of a woman with a hernia of Morgagni. The possibility of a common hereditary etiology is discussed.

CASE REPORTS

Case 1
The patient is a 37-year-old woman who sought medical attention because of a sensation of retrosternal pressure which had persisted for four weeks. Chest roentgenogram revealed a soft tissue density in the right cardiophrenic angle (Fig 1). No previous chest roentgenogram was available. The patient was otherwise asymptomatic. Her past medical history was unremarkable and there was no known family history of congenital hernia except for her daughter. No abnormal findings were noted on physical examination.

The preoperative evaluation, including barium esophagus and upper gastrointestinal series, was within normal limits. Exploratory thoracotomy was elected for diagnosis and therapy.

At operation, a hernia sac was noted to be protruding through a defect in the right anterior hemidiaphragm. This hernia contained only extrapleural fat and was easily reducible. The sac was excised and the defect was closed with nonabsorbable sutures. The postoperative convalescence was uncomplicated.

Case 2
The patient is a 14-year-old girl who presented for evaluation of an intermittent right supraventricular mass. This mass, noted by her parents since she was eight years old, was present only with crying or severe straining. It caused no symptoms and had remained stable in size over the past five years.

The patient was the product of normal pregnancy and delivery and had no past history of trauma or respiratory disease.

Physical examination revealed a soft discoid right supraventricular mass appearing only upon severe straining against a closed glottis. A barium swallow was normal and the diagnosis of cervical lung hernia was confirmed on the lateral neck roentgenogram (Fig 2). The presence of bilateral cervical ruts was also noted (Fig 3). No treatment was recommended; subsequently, the patient has remained free of symptoms.

DISCUSSION

Morgagni Hernia
A hernia of Morgagni is a herniation of abdominal contents through a defect in the anterior diaphragmatic muscle fibers. This hernia comprises approximately 3 percent of all diaphragmatic hernias and with rare exceptions, a sac is present. 1 2 3 In a series of 50 cases, Comer and Clagett 1 reported an occurrence of 90 percent on the right side, 2 percent on the left side and 8 percent bilateral. The incidence in women is more than twice that in men. 1 4 5

Morgagni hernias rarely cause symptoms during childhood and are frequently detected only on routine chest roentgenograms. In later life, they occasionally cause minor respiratory or gastrointestinal symptoms or sensations of retrosternal pressure. These hernias frequently contain transverse colon; however, herniation of omentum, extrapleural fat, stomach or liver also occurs. 1

The radiographic appearance is usually that of a soft...
Approximately 20 percent of all lung hernias are congenital, and most of these are in the cervical region.  Most lung hernias encountered in the pediatric age group are congenital in origin.  Hochinger has suggested that only those hernias occurring in the first few weeks of life should be considered congenital; however, many early acquired hernias probably involve congenital weaknesses of the thoracic enclosure.  The coexistence of bilateral cervical ribs in the patient presented also favors a congenital etiology.

The cervical excursion of the dome of the pleura is normally limited by Sibson’s fascia.  An inherent defect or weakness in this fascia permits the apex of the lung to protrude into the cervical region during times of increased intrathoracic pressure.  This herniation usually occurs between the anterior scalene and sternocleidomastoid muscles.

Clinically, the hernia presents as a soft, smooth, rounded mass in the supraventricular region, increasing in size during a Valsalva maneuver and diminishing in size during quiet respiration.  The diagnosis is established by lateral or oblique roentgenograms of the neck during a Valsalva maneuver (Fig 2).

Hernias of the lung rarely heal spontaneously; however, strangulation is uncommon because of the compressibility of pulmonary tissue.  Bronsther et al. feel that no treatment is required for most children with lung hernias unless there is a progressive increase in size, a history of incarceration, respiratory distress or a severe cosmetic problem.

The only definitive treatment for lung hernia is surgery.  A variety of methods have been employed including reduction and packing with gauze, utilization of fascia lata strips, Marlex mesh and plastic repair with flaps of peristomeum, ribs and muscle.  These procedures have, in general, been quite successful.

Embryology

Both Sibson’s fascia and the anterior muscle fibers of the diaphragm have their origin in the third, fourth and fifth segments of cervical mesenchyme between the fifth and seventh weeks of embryonic development.  The coexistence of these relatively rare hernias in mother and daughter respectively raises the possibility of a hereditary deficiency in the development of the cervical mesenchyme as a common causative factor in the hernias under discussion.

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Pulmonary Resection in Hemophilia

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Acase of classic hemophilia requiring pulmonary resection is presented as the first known to be recorded in the world literature.

In classic hemophilia, bleeding results from a deficiency of factor VIII, the antihemophilic factor (AHF). This bleeding tendency is initiated by the hemostatic mechanism by a combination of vascular contraction, platelet aggregation, and fibrin formation. In the presence of factor VIII deficiency, insufficient prothrombinase (intrinacic thromboplastin) production results, leading to decreased thrombin generation and inadequate fibrin formation. With the development of superior techniques of plasma protein fractionation, we now have available both human and animal fractions which are rich in factor VIII activity and effective in controlling hemophilic bleeding.

A wide variety of surgical procedures have been carried out in the hemophiliacs. Croom and Hutchinson in 1953 have reported successful thoracotomy with dectoration for intrathoracic hematomas. A review of the literature, however, has failed to reveal a previous case which was subjected to pulmonary resection.

Case Report

A 42-year-old white man was admitted to Sinai Hospital of Detroit on November 7, 1968, complaining of hemoptysis and a low grade fever. In 1945, he had experienced protracted bleeding following a hemorhoidectomy at another institution. He was transferred to Sinai Hospital of Detroit where coagulation studies revealed a normal clotting time and mild prolongation of the partial thromboplastin time; however, a prothrombin consumption test and a thromboplastin generation test revealed factor VIII deficiency with only 10 percent of the normal level of AHF. A detailed history at that time revealed numerous hemorrhagic episodes which had either been minimized or forgotten and had not been appreciated by the operating surgeon. There was no familial history of bleeding. His past history was otherwise insignificant with the exception of an episode of pneumonia one year before admission.

Pertinent physical findings on admission consisted of a temperature of 101° F, numerous fine rales over the left lower lobe posteriorly, and knee joint deformities consistent with chronic hemarthrosis. Laboratory studies revealed hemoglobin 12.5 gms, hematocrit 35 percent, and white blood cell count 7,700 with a shift to the left. Chest x-ray film (Fig. 1) demonstrated multiple fluid levels within a bullous left upper lobe and compression of the adjacent pulmonary parenchyma.

On the fourth hospital day hemoptysis was noted to increase. Hemoglobin and hematocrit had dropped to 7.0 gms and 22 percent respectively. Repeat chest film (Fig. 3) now demonstrated near complete filling of the bullous cyst occupying the left upper lobe with fluid, presumed to be blood and apparent aspiration changes in the left lower lobe.

Because of the continued bleeding and the potential hazards of infection and aspiration with obvious communications between the bullous and the tracheobronchial tree, it was elected to prepare the patient for surgery. He received three units of whole blood to restore his hemoglobin to 12 gms. The factor VIII deficiency was corrected with glycine precipitated AHF utilizing the dosage calculations suggested by Shulman et al. Based on an estimated plasma volume, to restore AHF levels to 80 percent of normal. The dosages were initially calculated for a presumed half-life of four hours to compensate for continued bleeding and consumption of coagulation factors. When evidence of active bleeding had ceased, a half-