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CYSTIC HYGROMA

Report of Three Cases

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Cystic hygroma is a benign disfiguring tumor of the neck, axilla, or chest wall and is thought to originate in anomalous development of the lymphatic system. Although the term *cavernous lymphangioma* is sometimes used, *hygroma*, derived from the Greek and meaning "moist tumor," is preferred.

EMBRYOLOGY

According to Jordan and Kindred¹ the lymphatic system develops in one of the two following ways:

"There has long been a question as to whether the lymphatic channels of the embryonic body develop as centrifugal endothelial sprouts from existing venous channels or by the fusion of isolated mesenchym-lined spaces, which upon fusion grow centripetally and transitionally

communicate with veins, particularly the subclavian, sciatic, and renal veins."

In a discussion of these two views it is stated:

"The alternate view, namely, that the lymphatic primordia develop from venous sprouts, is based largely on the work of Sabin. According to this view there appears in the embryo of the second month, vascular plexuses in connection with the subclavian, sciatic, and renal veins. These plexuses separate from their veins and become transformed into two paired and one unpaired lymph sacs and the unpaired retroperitoneal sac respectively. From these lymph sacs the sprouting lymphatics invade the body. The lymphatics of the head, neck, and arms grow from the jugular sacs; the lymphatics for the legs and trunk from the inguinal sacs; those for the mesentery from the retroperitoneal sac. Both views recognize the occurrence of these venous plexuses and the associated lymph sacs. According to the earlier view (Sabin) the venous plexuses become transformed into lymph sacs; according to the later view (Huntington and McClure) the lymph sacs arise in the extraintimal (perivenous) spaces of the disappearing venous plexuses. Only the jugular sacs acquire permanent venous connections. They serve to provide permanent openings into the internal jugular veins for the later thoracic duct on the left and the right lymphatic duct."

According to these views endothelial-lined channels or sacs give rise to sprouts, which extend centrifugally into surrounding tissues to form a lymphatic vascular system throughout the body. It is suggested that the cause of cystic hygroma may be accidental failure of some part of the lymphatic plexus derived from the jugular sacs to establish intercommunication with the venous system, with resultant segregation of primordial lymph channels. Endothelial cells of segregated lymphatic vessels form sprouts, which penetrate into surrounding tissues. But because of lack of communication with venous or other lymph channels, hygromatous cysts are formed. Endothelial lining cells of hygromatous cysts also send out sprouts to form new cysts and to secrete lymphoid material.

PATHOLOGY

As might be expected from this development, hygromas have a gross appearance of multilocular and multilobular cystic tumors. Depending upon their age individual cysts vary from less than a millimeter to several centimeters in diameter. Walls of small young cysts are delicate, thin, friable, and translucent, whereas those of old large ones are relatively thick, dense, and fibrotic. The cysts contain clear, straw-

colored, or serous lymphoid fluid. Within the tumor mass are varying amounts of degenerating muscle, fascia, fat, nerve filaments, and vessels, which have been engulfed during the growth and progressive penetration of lymphatic sprouts into adjacent tissues. No definite tumor capsule is formed.

Walls of individual hygromatous cysts are made up of one or more layers of endothelial cells. Supporting stroma in small cysts consists of a thin loose network of fine connective tissue fibers; stroma in large cysts has a large amount of dense fibrous tissue. Within the stroma are accumulations of lymphoid cells and in some areas hyperplastic lymph nodes.

DIAGNOSIS

Hygromas have a peculiar, rather characteristic, rubbery or spongy consistency on palpation. They often are lobulated and vary in size from small tumors to large masses. The skin overlying the tumor is freely movable, but because many tumors penetrate into surrounding tissues, especially muscles, the tumor mass may not be freely movable over underlying structures.

Hygromas usually occur in infancy or early childhood, although cases have been reported in which they did not appear until middle life. Symptoms are due principally to pressure. Protrusion into the floor of the mouth may interfere with deglutition; penetration into the mediastinum or the neck exerting pressure on the trachea may cause dyspnea; pressure on the cervical or brachial nerve plexus may cause pain, weakness, and paresthesia. Usually the tumors progressively enlarge, but regressions and almost complete but temporary disappearance have been observed.

Differential diagnosis. Hygromas should be considered in the differential diagnosis of tumors of the neck and axilla, especially in children. Pathologic conditions with which they may be confused are lymphadenitis, different types of lymphomas including Hodgkin's disease and lymphogenous leukemia, lipomas, simple or branchial cleft cysts, aberrant thyroid tissue or goiter, and metastases to lymph nodes. Hygromas involving the mediastinum resemble thymomas or other substernal tumors on roentgenologic examination, which should be made of all neck tumors.

TREATMENT

A comprehensive pathologic and clinical study of cystic hygromas was made by Goetsch², who, as have many surgeons, stated that radical

resection is the treatment of choice. In some cases, however, operation may not be indicated or successful. Because these tumors are particularly susceptible to infection, the mortality rate is quite high. Infection in addition to hemorrhage is the principal cause of postoperative death. It is not always possible to remove completely hygromatous tissues involving or extending into the mediastinum. Some cases have benefited by roentgen or radium irradiation.

We have seen 7 patients with hygroma at the clinic, of which 3 are of particular interest and will be reported briefly to illustrate results of operation and irradiation.

CASE REPORTS

Case 1—A baby girl, aged 10 months, had progressively enlarging masses on either side of the neck below the mandible. The masses were pressing into the floor of the mouth (fig. 1a). The inside of the mouth had been incised at the age of six weeks without benefit. She had dyspnea and difficulty in taking nourishment.

The masses in the neck were lobulated and elastic but were not freely movable. The overlying skin was normal and movable. Roentgenograms of the chest showed a sub-sternal mass (fig. 1b). Except for the tumor masses and undernourishment, physical and laboratory findings were normal.

The clinical diagnosis was cystic hygroma of the neck and mediastinum.

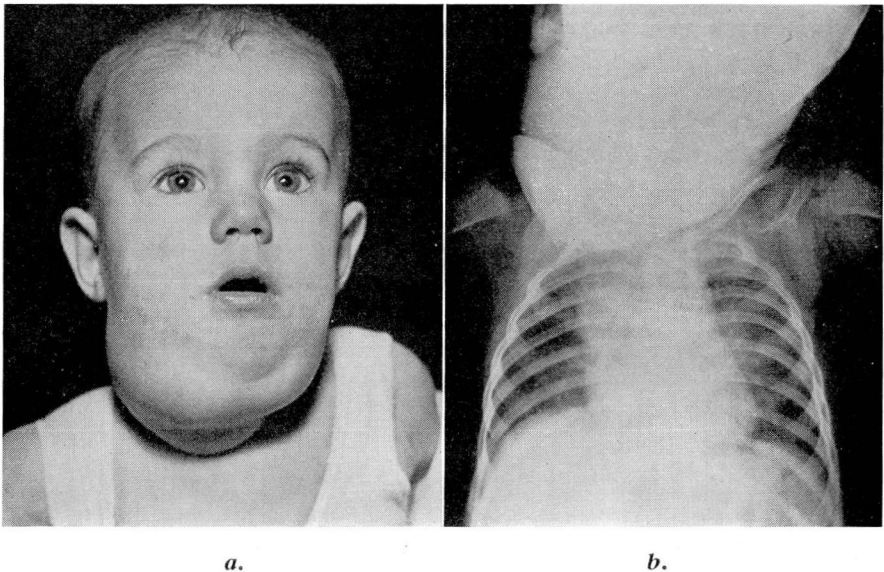


FIG. 1. a. Photograph showing bilateral involvement of neck. b. Roentgenogram of chest showing involvement of mediastinum.

A trial of roentgen therapy was advised. The technical factors employed were 200 kv., filter equivalent to 0.9 mm. hvl. copper, 50 cm. focus, skin-distance. Each side of the neck and the upper anterior mediastinum were given 100 r., measured on the skin through portals 10 by 10 cm. square, weekly for six weeks. The cervical masses began to diminish in size. When she returned for further treatment, the child had a fever, which followed vaccination a week previously, and it was decided to defer treatment until the fever subsided.

A few weeks later the child was taken elsewhere to a surgeon who attempted to remove the tumors in spite of extensive, surgically inaccessible, mediastinal involvement. The child recovered from the initial operation on one side of the neck. Later a second and more radical operation was attempted on the other side, and uncontrollable hemorrhage resulted in immediate death. The pathologist's diagnosis was "cystic lymphangioma."

Case 2—A baby boy, aged 2, was born with a cyst the size of a walnut on the right side of the neck. During the first year the tumor did not increase in size or cause symptoms. Then after a blow on the neck the cyst enlarged rapidly for about two weeks, and although it remained stationary, increasing dyspnea occurred. A roentgenogram of the chest revealed a substernal mass thought to be persistent thymus. A small amount of roentgen therapy was given to the neck and mediastinum. The tumor in the neck disappeared temporarily but soon returned to its original size, and a second treatment was given without benefit.

Physical examination of the child on admission revealed a large, lobulated, cystic mass in the right side of the neck extending from left of the midline anteriorly to the border of the trapezius muscle posteriorly (fig. 2a). The cyst was infected and was draining. Roentgenograms of the chest showed a substernal mass connected to the tumor in the neck (fig. 2b).

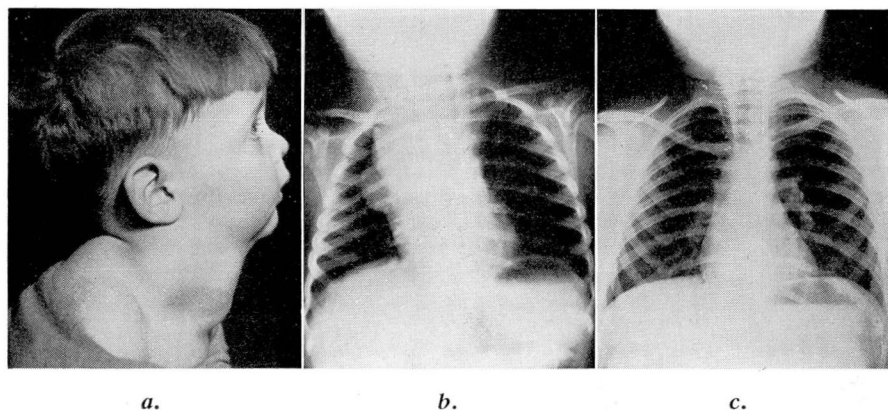


FIG. 2. a. Photograph showing right side of neck. b. Roentgenogram of chest showing involvement of mediastinum. c. Roentgenogram of chest five years after implantation of radon seeds seen in right neck and upper mediastinum.

A diagnosis of cystic hygroma was made and excision advised.

The infected area was incised and drained. At operation several weeks later the cyst was found to involve the carotid sheath and to extend high up, posterior to the trachea, and downward into the mediastinum in close proximity to the large vessels.

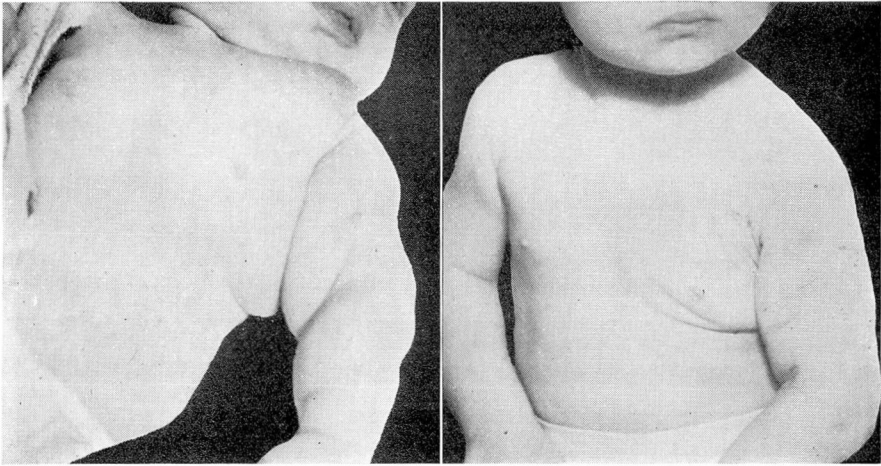
*a.**b.*

FIG. 3. *a.* Photograph showing involvement of left neck, chest, axilla, and upper arm. *b.* Photograph eighteen months after beginning of roentgen therapy shows considerable improvement.

Because it was impossible to resect all the mass, 14 radon seeds, each 1.4 mc. (filter 0.3 mm. gold), were implanted in the tumor remnants in the neck and mediastinum (fig. 2c). The pathologist reported the tumor to be cystic hygroma. Infection occurred after operation, but the child recovered and had no recurrence in nine years.

Case 3—A baby girl, aged 5 weeks, was born with “puffiness” of the left side of the neck, arm, and lateral chest wall. A diagnosis of sarcoma was made before her admission to the clinic. She was referred for roentgen therapy.

Examination revealed lobulated tumor masses filling the left supraclavicular fossa and axilla and extending down to the elbow and over the entire chest wall from sternum to axilla (fig. 3a). The rubbery consistency of the masses was characteristic of cystic hygroma. The skin was normal over the tumors, which were quite movable in some areas but somewhat fixed in others. The child was otherwise normal. Roentgenograms of the chest did not show mediastinal extension.

A clinical diagnosis of cystic hygroma was made.

After consultation with two surgeons who considered operation inadvisable because of extent of involvement, roentgen therapy was instituted. The technical factors used were the same as for the other cases. Through varying sized portals, different areas were given 100 r as measured on the skin at weekly and later monthly intervals for approximately two years. As nearly as it can be estimated, each part of the tumor mass received approximately 1200 r during this time.

Progressive reduction in size of all tumor masses occurred (fig. 3b). Only remnants remained above the elbow. At $2\frac{1}{2}$ years of age the child was normally developed without apparent interference with bone growth or muscle function.

SUMMARY

Three cases of cystic hygroma, a benign tumor thought to originate in anomalous development of the lymphatic system, are reported. One operative death was caused by uncontrollable hemorrhage. The second patient, treated by operation and implantation of radon seeds in surgically inaccessible remnants of the tumor, had no recurrence in nine years. The third patient, treated by roentgen irradiation, improved symptomatically, and the tumor masses were reduced.

Hygroma is usually detected in infancy or early childhood, although occurrence in middle life has been reported. Symptoms are caused by pressure on adjacent structures, since the common sites are the neck, axilla, and chest wall. Differential diagnosis is sometimes difficult, and roentgenologic examination is necessary.

Surgical resection has been considered the treatment of choice, but the mortality rate is high, hemorrhage and infection being the principal causes of postoperative death. When surgical removal of all tumor tissue is impossible, patients have benefited by irradiation.

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