Cystic hygroma: characterization by computerized tomography

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Lymphangiomas are benign nonencapsulated lesions composed of sequestered noncommunicating lymphoid tissue lined by lymphatic endothelium and are thought to be caused by congenital obstruction of lymphatic drainage. They are subclassified by vessel size, such as the capillary, which is rare and located in subcutaneous tissue, cavernous (located about the mouth and tongue), and cystic (cystic hygromas). The cystic hygromas show a predilection for the neck (75%) and maxilla (20%), and the remaining 5% arise in rare locations such as the mediastinum, retroperitoneum, bone, kidney, colon, liver, spleen and scrotum. Only 3%-10% of neck lesions extend into the mediastinum. In this paper, we report a rare case of cystic hygroma with a huge dimension discussing the use of computed tomography scanning for diagnosis. **(Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;105: e65-e69)**

Cervical congenital cystic masses constitute an uncommon group of lesions usually diagnosed in infancy and childhood. The most common congenital neck mass that extends to the mediastinum is the cystic hygroma, which is a cystic form of lymphangioma and constitutes about 5% of all benign tumors of infancy and childhood.[1] This lesion usually occurs in the lower neck, commonly in the posterior cervical space. It is usually evident at birth, with 80%-90% presented by the age of 2 years.[1-3] Frequently, cystic hygromas are asymptomatic and manifest as painless masses.

Cystic hygromas can result from environmental factors, genetic factors, or unknown factors. Environmental causes include maternal viral infections and maternal substance abuse, such as alcohol abuse. Parvovirus disease is a common, usually mild, illness. However, when a pregnant woman is infected, the virus can cross the placenta and cause a fetal cystic hygroma.[1-4]

There are several genetic syndromes that involve congenital cystic hygromas. The majority of cystic hygromas found prenatally are associated with Turner syndrome, a chromosome abnormality in which a girl has only 1 X chromosome instead of 2. Chromosome abnormalities, such as trisomies 13, 18, and 21, also have been associated with cystic hygromas. Other genetic syndromes, such as Noonan syndrome, have a cystic hygroma as a common prenatal finding. The pattern of inheritance for these syndromes varies depending upon the specific syndrome. Isolated cystic hygroma can be inherited as an autosomal recessive disorder for which parents are "silent" carriers.[1-5]

Lymphangiomas are thought to arise from a combination of the following: a failure of lymphatic vessels to connect to the venous system, abnormal budding of lymphatic tissue, and sequestered lymphatic rests that retain their embryonic growth potential. These lymphatic rests can penetrate adjacent structures or dissect along fascial planes and eventually become canalized. These spaces retain their secretions and develop cystic components because of the lack of a venous outflow tract.[6]

Lyphamgiomas are classified as microcystic (capillary lymphangiomas), macrocystic (cavernous lymphangiomas), and cystic hygromas according to the size of the lymphatic cavities incorporated. When a lymphangioma is confined to dense tissue it is presented as a cavernous lymphangioma, but when it is located in a relatively loose tissue single or multiple fluid-filled cystic dilatations (usually greater than 1 cm in diameter), i.e., cystic hygromas occur. [7]

CASE REPORT

Patient J.A., 1 year of age, was referred to the Maximagem Medical Diagnosis for a computerized tomography (CT) scan. He presented with a tracheostomy performed just after birth and showed an abnormal mass of huge proportions in the neck and thorax. The lesion was flaccid and extended from the infraorbital suture to the mediastinum area. The mother stated that it was painless, apparently causing

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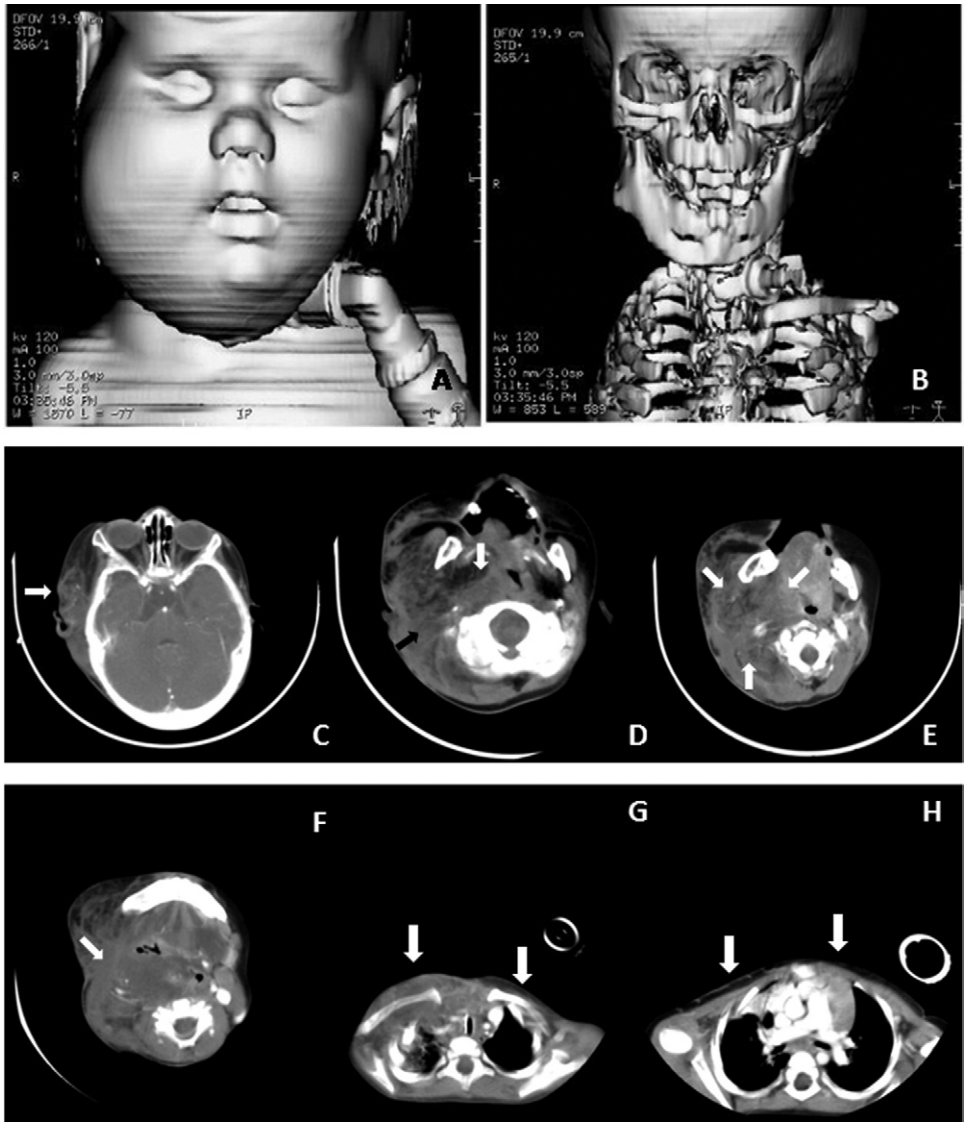

Fig. 1. Extensive and infiltrative lesion extending from the cervicofacial region to the mediastinum and anterior thoracic wall displacing adjacent structures. **A and B,** 3D reconstructions showing the volume and size of the lesion causing facial asymmetry. **C,** Infiltration of the superior portion of the temporal space and the outer ear. **D,** Enlargement the stylomandibular tunnel (*black arrow*) and infiltration of the lateral pharyngeal wall (*white arrow*); **E,** Reduced and displaced pharyngeal space. **F,** Obliteration of pyriform sinus involving the parapharyngeal area and moving the hypopharyngeal air column. **G,** Infiltration of the anterior thoracic wall (*left white arrow*) and the superior portion of the mediastinum (*right white arrow*). **H,** Infiltration of the prevascular space (*left white arrow*) in the mediastinum causing atelectasia of the superior portion of the right lung (*right white arrow*).

no paresthesia, and there was no sign of inflammatory origin, such as fever.

The medical history of the mother revealed that during pregnancy, she presented skin irritation similar to the bites of insects. She also revealed that the prenatal exams showed "an uncommon problem," but she could not give any further information.

The CT images showed an extensive lesion infiltrating and displacing adjacent tissues. The lesion involved the cervicofacial region infiltrating the superior portion of masticator space, the outer ear, and the lateral pharyngeal wall causing enlargement of the stylomandibular tunnel. This enlargement reduced and displaced the pharynx, the anterior thoracic wall, and the superior portion of the mediastinum. In addition, the infiltration of the prevascular space caused atelectasis of the supe-

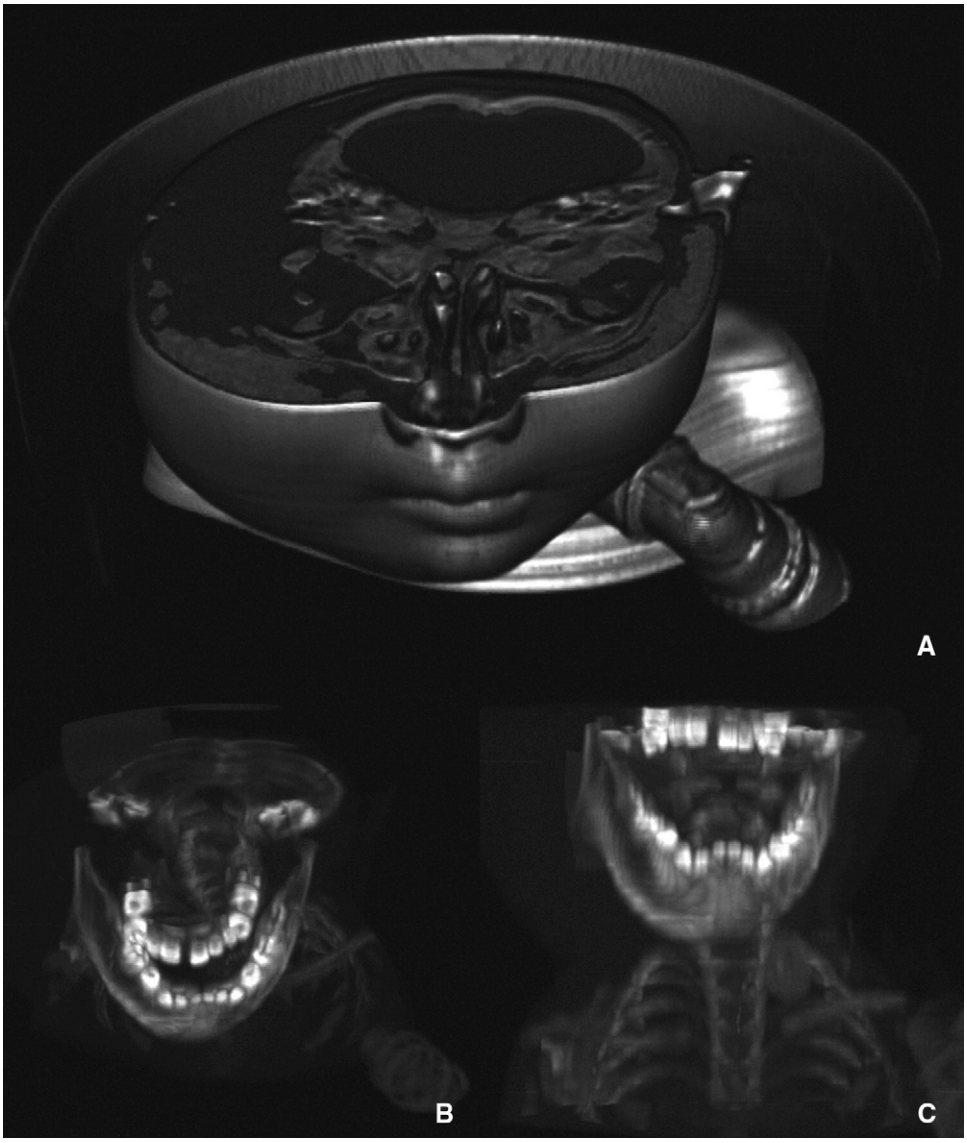

Fig. 2. The 3D and 2D reconstructions showing the volume of the lesion and the compromised tissues (InVesalius 2.0.6-CenPRA). **A,** 3D imaging with axial segmentation: displacement of the mandible condyle and soft tissues. **B and C,** 2D imaging using maximum intensity projection showing bone deformities in the head and neck area.

rior portion of the right lung (Fig. 1). Owing to continuous pressure, the bone structure was also displaced by the lesion (Fig. 2).

The clinical characteristics and the CT images led to the diagnosis of cystic hygroma. After diagnosis, no other examinations were done and the patient was followed-up for 6 months without any alteration of the clinical aspects. Owing to respiratory difficulties, the tracheostomy was maintained.

The treatment proposed includes the careful surgical removal of the lesion without compromising vital neurovascular tissues. After the surgery a new evaluation will be performed in an attempt to estimate the necessity of any other complementary treatment, such as the administration of sclerosing agents.

DISCUSSION

Cystic hygroma presents as a soft tissue mass in the posterior triangle of the neck and only rarely extends into the mediastinum. Some cystic hygromas are diagnosed during childhood only if the lesions are very large.[8] The present case showed early signs of growth before birth and at 1 year of age was already assuming a huge proportion and invading the mediastinum and

adjacent tissues. Although there was no apparent cause, the patient's mother presented "skin irritation similar to insect bites," and the infection for human parvovirus B19 may show a similar clinical appearance.

Airway obstruction is the most critical complication of cystic hygromas occurring in the neck.[6] In this case, a tracheostomy was needed owing to the pressure on the airways by the lesion. Other possible complications include hemorrhage, infection, and deformation of surrounding bony structures.[6,9,10]

Congenital cervical cystic lesions are usually slow-growing masses and typically cause symptoms only because of enlargement or infection. A painless soft or fluctuant cervical mass is the first clinical manifestation in most cases. After physical examination, ultrasonography (US) is usually performed to define the size and extent of the mass; however, US has limited ability in assessing mediastinal and retropharyngeal structures. Computerized tomography also provides important information and is ideally suited for evaluation of soft-tissue planes adjacent to larger masses that cannot be entirely visualized with US.[7] Computerized tomography was preferred in the present case because it was previously known that the lesion growth was compromising and infiltrating adjacent tissues, including the mediastinum. The biopsy was considered to be unnecessary for diagnosis and was avoided owing to the risk of secondary infection.

Computerized tomography scanning is faster and may be more readily available than magnetic Resonance imaging. Computerized tomography scanning carries the risk of radiation exposure and the disadvantage of loss of details if the cystic hygroma is surrounded by tissue of similar attenuation. Contrast helps to enhance cyst wall visualization and the relationship to surrounding blood vessels. On the CT scan, cystic hygromas appear isodense with cerebrospinal fluid.[6]

The differential diagnosis for congenital neck lesions includes thyroglossal duct cysts and branchial cleft cysts, encephalocele, meningomyelocele, and teratoma.[11,12] The diagnosis of thyroglossal duct cysts is easily established by the presence of a cystic lesion in the anterior midline portion of the neck. The first cleft cysts, or parotid lymphoepithelial cysts, manifest as recurrent abscesses or other inflammation (sinus tract) either around the ear or at the angle of the mandible. Second branchial cleft cysts can occur anywhere along a line from the oropharyngeal tonsillar fossa to the supraclavicular region of the neck, usually in the submandibular space. These anomalies typically occur between 10 and 40 years of age, in contrast to fistulas or sinuses, which manifest most commonly during the first years of life.[11] Teratomas are uncommon and are generally solid. Encephaloceles have associated calvarial abnormalities and contain brain tissue. Meningomyeloceles are uncommon in the cervical region and have associated spine abnormalities.[12] Imaging studies are not usually necessary for the differential diagnosis of these congenital neck lesions, whereas in this case of cystic hygroma the CT scan was crucial.

There are several possible approaches to treat the cystic hygroma. Sclerosing agents and/or steroids can be used[6] and combined with antibiotics when the case is infected. On average, surgical excision of the abnormal mass tissue and management of airway is indicated.[3,6,9,10,13] In the present case, during the first 6 months of follow-up no treatment was performed, because the child was too young and a possible regression was expected.

Recurrence after complete resection occurs in approximately 10% of the cases; however, cystic hygromas can be expected to recur if residual disease remains.[6] Spontaneous regression occurs in 6% of cases.[14,15] Follow-up is always necessary to guarantee the success of any treatment.

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