CYSTIC HYGROMA

Key Concepts
- Definition of cystic hygroma
- Pathology of cystic hygroma
- Causes of cystic hygroma
- Types of cystic hygroma
- Management of cystic hygroma

Abstract
Cystic hygroma is a multiloculated cystic structure which is benign in nature. These are commonly present in head and neck but can be present anywhere. The pathogenesis is developmental failure of lymph channels. This can occur due to environmental, genetic or other causes. Imaging techniques aid in the cysts and related anatomic structures. Treatment options are observation, surgery and sclerotherapy.

Key words: Lymphangioma, Cystic hygroma, Cavernous, Lymphangioma, Sclerotherapy.

Introduction
Lymphangiomas, soft tissue tumors of disputed pathogenesis was originally reported by Reden Backer in 1828 & “cystic hygroma” name was first given by Wernker in 1834.

Cystic hygroma is multiloculated cystic structure that is benign in nature. It may occur anywhere in the body, although it is most commonly encountered in neck. They frequently about the neurovascular structure. Lymphatic channels are formed around the sixth week of gestation; from these channels sacs are formed that establish drainage with venous system. Failure to develop to establish venous drainage result in dilated disorganized lymph channel, which in largest form present as cystic hygroma. In the embryo, the lymphatic systems drain into the jugular lymphatic sac. A communication between primitive structure and jugular vein is formed at 40 days of gestation. Failure of development of this communication results in lymphatic stasis, with formation of cystic structure in cervical region. If a connection between lymphatic and venous system does
not occur at this point, a progressive peripheral lymphodema and hydrops develop, leading to early intrauterine death.

Causes
Cystic hygroma can occur as an isolated finding or in association with other birth defects as a part of syndrome. They result from environmental factor, genetic factor unknown factor.

Environmental causes of cystic hygroma include
- Maternal viral infection such as parvovirus of Fifth’s disease.
- Maternal substance abuse such as abuse of alcohol.

Genetic factor
Chromosome problems are often diagnosed. When a cystic hygroma is seen about 50-60% of babies with their U.S finding have a chromosome problem. The most common chromosome problems are “Turner Syndrome” Down syndrome and trisomy 18.

Birth defects
Baby with cystic hygroma are also likely to have physical birth defects. Heart defects are most common type of birth defect found but many different type of birth defects have been found.

Genetic Syndromes
There are many different genetic syndromes that can lead to cystic hygroma. Most of them are rare and difficult to diagnose fetal loss, (Miscarriage fetal demise/ still birth). If connection between lymph system blood vessel does not form at some point, extreme fluid build up (hydrops) in developing baby & can lead to loss of pregnancy.

Histological Subtypes
Lymphangioma is a benign congenital malformation of lymphatic system. There are three histological subtypes.

1. Capillary lymphangioma (compose of small lymphatic).
2. Cavernous lymphangioma (composed of large lymphatic).
3. Cystic lymphangioma (Cystic hygroma composed of large macroscopic lymphatic spaces with collagen and smooth muscles). Cavernous lymphangioma is the most common subtype, but cystic lymphangioma occurs approximately in 12000 births and 95% occur in second year of life. Although lesion can be anywhere, the most common sites are in post triangle of neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal and pelvis.

INVESTIGATION
Diagnostic testing
Prenatal diagnostic testing (CVS or amniocentesis) can accurately identify chromosomal abnormalities. There is small risk of miscarriage associated with those procedure(<1/300 risk).

Ultrasonography
A high resolution (Level 11) U.S is offered around 18 to 21 weeks in pregnancy to carefully look at baby. If there is concern
about possible heart defect called a fetal echocardiogram.

Follow up ultrasound should be done to watch for changes in amount of fluid build up. Extreme fluid build up in pregnancy may lead to health concern for sock baby & pregnant woman.

**M.R.I Scan**
Lymphangiomas are best visualized by M.R.I scan. High water content allows lymphangiomas to appear hype intense on T2-weighted image. Doppler ultrasound and CT scan are other modalities which may be helpful.

**Will cystic hygroma go away**
Sometimes cystic hygroma goes away (resolves). It is more likely to go away when cystic hygroma is small seen before 14 wks of gestation. When a cystic hygroma goes away, the chance of survival for a developing baby improves. However cystic hygroma that disappears does not provide reassurance about chromosomal problems birth defects or genetic syndromes.

**What about treatment?**
There are 3 options for cystic hygroma. As they are all different there is no substitute for detailed imaging ( Preferably with MRI scanning) and assessment by someone experienced in managing them.

1. **SIMPLE OBSERVATION**
This is generally recommended for small lesions that are causing minor symptoms only.

2. **SURGERY**
If the lesion can be completely removed without damaging other structures then it will not come back and this is the end of it. This is what many patients would like to achieve. An experienced surgeon needs to evaluate general terms large cysts (Macrocysts) are more favorable as our lesions in the neck. If small cysts are left behind after surgery because they are inaccessible, they may never cause problem (See below with sclerotherapy. All the cysts are left in). Problem in the tongue and mouth are more difficult to remove completely. Although surgery to this area to reduce the problem may be appropriate. Cyst on the tongue surface may be treated with laser surgery.

3. **SCLEROTHERAPY**
The principle of sclerotherapy is to inject a substance into the cyst which causes inflammation. The bodies reaction to this foreign substance causes the cyst to scar down, The lymphatic malformation remains present but is scarred so it cannot fill up with fluid again. The end result is that the lump is smaller and not visible. Following the injection the cyst increases in size sometimes dramatically then gradually reduces in size over the next 3 months or so. If the response is incomplete injections can be repeated. The attraction of sclerotherapy is that it avoids surgery and often avoids a scar and risks of nerve damage are less. A general anaesthesia is required for most children.

**REFERENCES:**


