

## Experience with Lymphangioma\*

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### Summary

Lymphangioma is a rare and benign tumor, mostly encountered in childhood. Our experience extends to 21 patients, all of which were operated upon. Early radical excision in moderate forms or cervical locations and delayed excision or combined management in complex cervical forms are the more preferable way of treatment.

### History and Ontogenesis Theories

Lymphangioma is a developmental defect of the vascular system. To date, the pathogenesis is still somewhat controversial. In the development of the lymphatic system, cell buds, representing the lymphatic primordium, are pinched off the venous channels and form a capillary network growing into different tissues.

At some stage of the development these primitive lymphatic sacs reunite with the venous system.

According to this centrifugal theory (Dowd, Goetsch, Sabin, Mc. Clure, Silvester, Godart) lymphangiomas develop when the primitive lymphatic sacs fail to re-establish venous communication. We have then a sequestration of lymphatic channels which have a certain growing potentiality.

The *centripetal theory* (Huntington, Kampmeier) claims that at a very early stage of foetal development (9-12 mm), mesenchymal slits appear in the reticulum of venous plexuses. By coalescence of these spaces, larger lymphatic cavities are formed which open secondarily into the venous system or

again, if this mechanism fails, induce cystic formation.

Recent experimental work on the early development of the lymphatic system by *Van der Putte* is of strong support for the centrifugal theory. A more or less regular series of small and blind ending outgrowths of the major embryonic veins form the early lymphatic system. The system is subdivided in four singular and twelve groups of primordia.

Except for some openings at the jugular subclavian junction all connections with veins disappear.

The primordial extensions grow out *centrifugally* and invade the surrounding tissues.

Finally, many regard lymphangiomas as *hamartomas* (Willis) i. e. an embryological tumor formed by an excessive growth of tissues in their normal location.

### Frequency

Lymphangioma is a *rare* tumor and accounts for only 6% of benign tumors in childhood and for 5% of vascular growths. In a series of 152 benign necktumors only 4 lymphangiomas were listed.

### Pathology-Forms

Three main forms of lymphangiomas exist.

1. *Capillary lymphangioma*, an uncommon and insignificant lesion, appearing as an ordinary wart or a group of small vesicles. Histologically it consists of a network of spaces formed by small and medium sized vessels.

\* Read at the VIth International Congress of Lymphology in Prague

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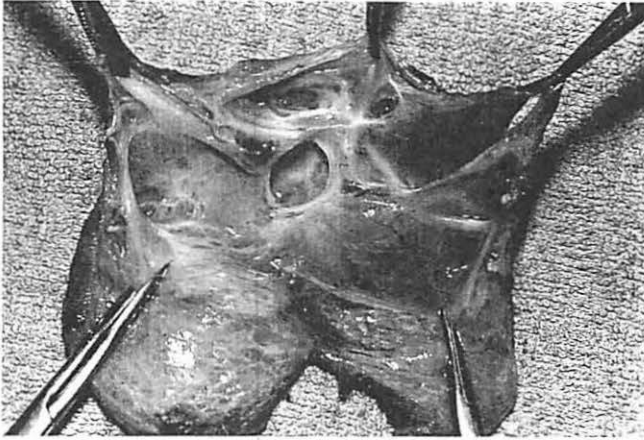


Fig. 1 Resection specimen of cystic hygroma: lobulated endothelial-lined lymphatic spaces

2. The second form is the *cavernous lymphangioma*. They occur usually in the skin and subcutaneous tissue, the salivary gland, tongue and lips, where they are responsible for macroglossia and macrocheilia. They are diffuse, spongy, compressible masses, the margins of which are often difficult to delineate. The microscopic appearance consists chiefly of multiple dilated lymph channels lined with single or multiple layers of endothelial cells. These tumors are noted for their invasive tendencies and they frequently penetrate underlying muscle and contiguous structures.
3. The third form is *cystic hygroma*. It is the most frequent of the three forms and it develops in the neck, the axilla (Fig. 1) and the retroperitoneal region.

Less frequently it occurs in the mediastinum and the limbs, and rarely in bone, the wall of the gastrointestinal tract, the mesenterium, the omentum, the spleen, the pancreas or the liver and in the urogenital tract. Even in the heart a lymphangioma has been described and recently we came across a case of lymphangioma of the parotid gland. The tumor is multicystic, and the thin walled cysts are lined by lymphatic endothelium.

*Bill and Sumner* support the theory that the three forms of lymphangiomas are basically similar entities and that their morphologic differences are related to their anatomic

location. In lips, cheek and tongue, limited expansion is possible and the cavernous form is induced. In the axilla, neck and chest wall areas with loose fatty tissue permit relatively unlimited dilatation and cystic hygroma occurs.

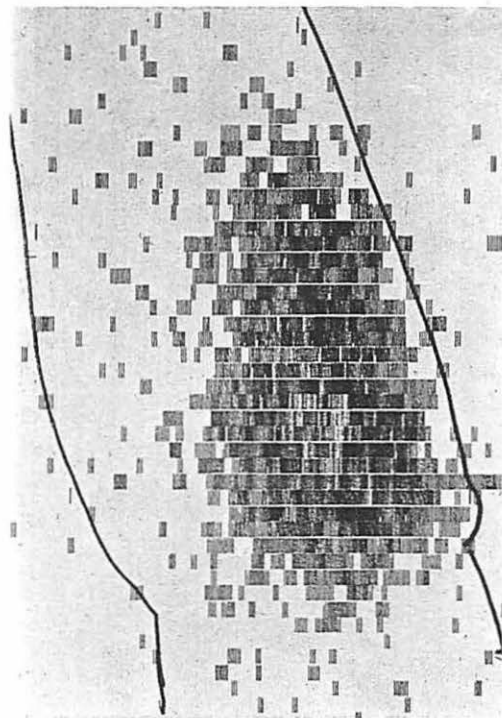
Some authors classify the following other tumors as lymphangioma:

- 1) Lymphangio-haemangioma
- 2) Lymphangio-lipoma
- 3) Lymphangiosarcoma
- 4) Lymphangioma of bone

This disease has other names like "Vanishing bone", "Phantom bone", multiple lymphangiectasis of bone. The X-ray appearance is diagnostic and scintigraphy is very helpful in locating the lesion. Osteolysis is slowly progressive and results in complete disappearance of bone after months or years. (The authors presented three cases in extenso at the IVth International Congress of Lymphology). Fig. 2-3).

It is worthwhile to mention two other related conditions:

1. *Lymphangiomyomatosis of the lung and the mediastinum*. A chronic progressive disease encountered in middle-aged females, often associated with chylothorax, hormone-dependant and carrying a very poor prognosis of a mean survival rate of 18 months.



Lymphangioma of bone: (Fig. 2) Radiographic and (Fig. 3) scintigraphic appearance

2. *Lymphangiomatosis of liver and spleen* with bone involvement has a similar poor prognosis.

We collected 21 cases of lymphangiomas, the distribution of which among the different forms is mentioned on the following table.

#### 21 PATIENTS

Lymphangioma capillare	1
Lymphangioma cavernosum	4
Lymphangioma cysticum	8
Mixed form (cavern. + cyst.)	4
Lipo-lymphangioma	1
Lymphangioma of bone	3

#### Symptoms and Diagnosis

Usually the diagnosis is obvious: an abnormal mass progressively increasing in size, translucent and sometimes rapidly enlarging as a consequence of bleeding or infection.

In a little girl with a large lymphangioma of the axilla, puncture for biopsy

was followed by infection with rapid enlargement of the tumor.

A number of symptoms depend on the location of the tumors and rare clinical pictures belonging to the chylous syndromes can occur.

One of our patients, a young woman, presented with vulvar chylorrhea. Investigation brought about a huge cavernous lymphangioma of the pelvis. (Fig. 4)

Lymphangioma is often a disease afflicting children as can be concluded from our series where 65% of the patients appeared in the pediatric age group.

#### 21 PATIENTS

present at birth	7	33%
0-2 y	11	55%
2-14 y	2	10%
14-30 y	3	14%
30-65 y	5	24%

Rate of occurrence related to age

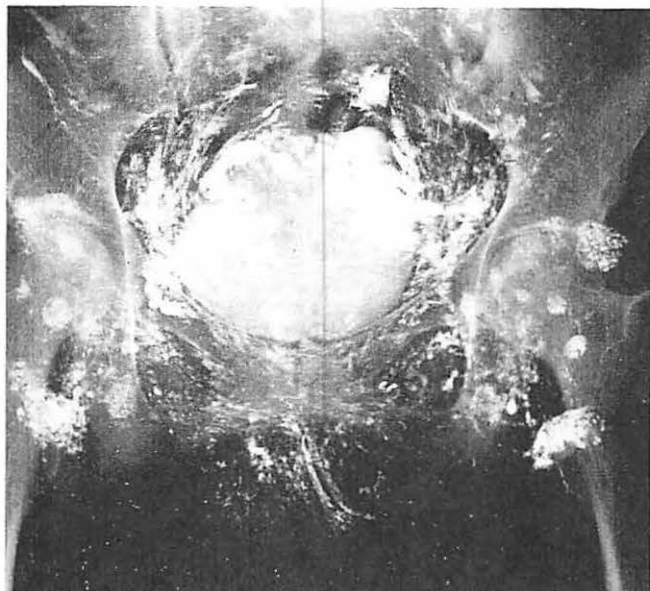


Fig. 4 Lymphangiography in a case of vulvar chylothorax due to cavernous lymphangioma of the retroperitoneum and the pelvis.

The different locations in 453 patients which is a review of the literature, compared with our own 21 patients are listed in the following table.

Location	Literature	Own pt.
Neck	39%	30%
Head	20%	
Axilla	13%	25%
Trunk	8%	10%
Limb	11%	15%

#### Anatomical distribution

#### Associated Developmental Pathology

Six of our patients showed other associated pathology as: dysmaturity, microcephaly, low implantation of the ears, mental retardation, large hiatus hernia, pubertas tarda, eosinophilic granuloma-lesions, angiofibroma, haemangioma, liposarcoma, fibro-lipo-haemangioma and a gastric haemangioma.

#### Treatment

The goals of the treatment are (1) eradication of the lesion and complete healing; (2) absence of morbidity and mortality; (3) good esthetic result.

1. *Spontaneous remission* did not show in our patients.
2. *Sclerosing injections* were used by us for a very mild recurrence after resection in the patient with vulvar lymphorrhea.
3. *Radiation* can be dangerous and in our series we have a possible example of neoplastic potentialization. In a man of 30 years who initially had an angiofibroma of the right thigh which was treated by resection and radioactive gold seeds implantation recurrences occurred and the man finally ended up with a presacral liposarcoma and metastases.
4. *Incision and drainage* may be used as a temporary measure to overcome life-threatening situations. In one case of our series with extensive lymphangioma of the neck, cheek and mouthfloor, this way of treatment had to be applied after partial resection of the tumor.
5. *Surgical excision* seems to be the most reliable form of therapy and should be used at the earliest opportunity. Many patients have to be operated upon several times as is listed in the next table. (Fig. 5-6).



Fig. 5-6 Huge cervical cystic lymphangioma and morphological result, several years after excision.

1 operation	16 pts.
2 operations	3 pts.
3 operations	1 pt.
4 operations	2 pts.

Number of operations per patient

For large lymphangiomas a multistage procedure can be preferable.

Complications of surgery essentially are damage to nerves, lymphedema of the surrounding tissues, lymphocele formation or lymphorrhoe are other possible complications.

In our series we encountered complications in 4 patients: partial facial nerve paralysis in two, right phrenic nerve paralysis, shoulder paresis, *Claude Bernard Horner* syndrome and hoarseness in a third and a neurinoma in the scar of a fourth patient.

### Conclusion

Although a benign condition, lymphangioma can be puzzling in regard to the histology and the radical treatment. Especially large, infiltrating forms in the head and neck region of infants, represent a challenge for radical cure without morbidity. We are convinced that an eclectic attitude comprising early radical excision in moderate forms or non cervical locations and delayed multistage excision or combined management in complex cervical forms, is mandatory.

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