International Foundation for Pediatric Imaging Aid
Lymphangiomas in infancy and childhood: An overview and imaging approaches

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Introduction

Lymphangioma:

- is a congenital malformation of the lymphatic system.
- arises from sequestration of lymphatic tissue failing to communicate with the lymphatic pathway.
- tends to surround and invade normal anatomic structures.
Introduction

- The incidence is 1:4000 - 6500 life births.
- Lymphangiomas are commonly benign and asymptomatic, but incidentally cause pressure and a life threatening complication.
- Hemorrhage or infection are both the cause of sudden enlargement.
Features of lymphangiomas

Based on size and anatomic structure, lymphangiomas are classified in:

a. Cystic hygroma.
b. Cavernous lymphangioma.
c. Capillary sized simple lymphangioma.
d. Vasculolympathic lymphangioma (with hemangiomatous compartments).
Imaging modalities

- Conventional X-ray examination depending on location.
- Sonography.
- MRI.
- CT.
Sonographic appearance of lymphangiomas

- An- or hypoechoic, mostly multilocular cystic mass.
- Thinwalled with septa of variable thickness.
- Cluster of microcystic lymphatic channels may be hyperreflective.
- Exceeding to other anatomic region and neighbouring organs.
- Hypo- or avascular by color Doppler evaluation.
- Hypervascularity in cases of mixed type vascular elements (hemangiolymphangiomas).
Different echographic pattern of lymphangiommas from different location
MRI characteristics of lymphangiomas

- High signal intensity on T2 and low signal intensity on T1.
- The signal characteristics can be variable in some cases depending on degree of protein.
- Multiple cysts with well demarcated margins on T2.
- Delineation of full extent of the lesion by imaging of neighbouring organs.
Giant hygroma colli by two infants.

MRI of a giant hygroma with hyperintensity on T2 and septations.
Locations of lymphangiomas

- Lymphangiomas may occur at any anatomic location in the body with exception of the central nervous system.
- Approximately 75% is located in the posterior triangle of neck and axilla.
- Extension of hygroma colli in mediastinum is reported in about 10%.
- Mesenteric location is around 8%.
- Other incidentally reported sites of lymphangiomas are retroperitoneal space, kidneys, scrotum, upper and lower extremities and trunk.
Prenatal MRI from a foetus with giant hygroma colli.
Hygroma colli growing in the retrolaryngeal space.
Lymphangioma of the neck with intrathoracic extension
A boy with mesenterial cystic lymphangioma in the age of 8 and 9 years.
Cystic intraperitoneal lymphangioma in a 5-year-old girl
2-months-old girl with a giant retroperitoneal cystic lymphangioma with displacement of kidney and right-sided hydronephrosis.

Note: the displacement of bowels.
4-year-old girl with retroperitoneal lymphangioma.
Lymphangioma in a newborn boy with cluster of microcystic lymphatic tissue.

Suprarenal located.
Newborn girl with a cystic lymphangiomma of the right flank.
1,5-year old boy with a cystic lymphangioma on the right thoracic wall.
5-year-old boy with a cavernous lymphangioma located on right medial malleolus.
Lymphangioma with vascularty malformation in a 5-year-old boy in the right cheek.
6-year-old boy with a disseminated lymphangioma with vascular malformation.
Cystic hygroma with an acute bleeding.  
Note the fluid level of the blood.
Lymphangiomatosis

- Is a disease characterized by maldevelopment of lymphatic drainage.
- Occurs mainly in children and adolescents (very rare).
- Single organ involvement known as pulmonary lymphangiectasia.
- Multi-organ involvement known as generalized lymphangiomatosis.
- In Gorham’s disease lung and bone are both affected.
4-year-old boy with persistent hydrothorax and clavicle destruction.

(Gorham syndrome)
Boy born with an excessive lymphedema and respiratory distress.
Later on he suffered from recurrent pleural effusion.
CT and chestfilm both show the sign of lymphangiomatosis of the lung.
Conclusions

- A cystic lesion in infancy and childhood can be classified as either neoplastic or non-neoplastic.

- Cystic lymphangiomas of the neck should be differentiated from other congenital cystic neck masses.

- An intra-abdominal cystic lesion of unknown origin should be considered to be a lymphangioma.
Conclusions

- Recurrent chylothorax of unknown etiology can be the first manifestation of lung lymphangiomatosis.

- Bone destruction together with chylothorax is the classical sign of Gorham’s disease.

- Most lymphangiomas exceed to other anatomic layers.
Conclusions

- Sonography is the modality of choice in screening and follow-up of cystic lymphangiomas.

- Thinwalled, an- or hypoechoic cystic lesion, mostly multilocular, is characteristic for lymphangioma.

- Color Doppler or duplex sonography should be routinely used to demonstrate the vascularity of the masses.
Conclusions

- MRI is absolutely indicated to assess the extension of lymphangiomas and its relation to neighbouring organs, especially in neck, thoracic cage and abdomen.

- The indication to use CT in lymphangioma is to look for angiomatous components and the extension of hemangiolympanghiomas.

- In pulmonary lymphangiomatosis CT is superior to MRI.