Imaging findings of fetal and perinatal lymphangiomas

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Learning objectives

The purpose of the poster is:

- to present lymphangiomas of different localizations
- to describe prenatal and perinatal ultrasound (US), magnetic resonance (MR) and computed tomography (CT) findings
- to discuss the advantages of those supplementary to ultrasound (US) methods in regard to patient counseling, management delivery decisions and postpartum follow-up.
Background

Lymphangiomas are uncommon benign congenital cystic malformations of lymphatics due to developmental failure with the potential to infiltrate surrounding structures. These uncommon, benign, cystic soft-tissue tumors in neonates and children can affect different body regions. They consist of solid and cystic components, with variable extent and may grow very large even in utero. Spontaneous regression of lymphangiomas is controversial. Sudden increase in size could be observed in intralesional hemorrhage. The local growth and extension occurs along the layers of connective tissue between organs and muscles. Fetal and perinatal imaging is performed by ultrasound (US), magnetic resonance imaging (MRI) and rarely computed tomography (CT) [1]. Lymphangiomas are discovered mainly by US, as first method of choice in fetal examination. However, fetal MR imaging is suitable to confirm or to reject ultrasonographic findings, provides detailed anatomical orientation for evaluation of the extent and character of lymphangiomas and facilitates further surgical planning. This poster describes the common MR and CT findings and the different localizations of lymphangiomas in fetuses and newborns. All cases showed normal karyotype.

A short review of the literature shows that the ability to diagnose fetal tumors has improved over recent years. Avni et al. find ultrasound findings and the location of the tumors suitable for establishing the differential diagnosis, however in selected cases MR imaging can provide additional information [2]. According Rodriguez et al. MR imaging offers various technical advantages over US, including a larger fields of view, limitations due to maternal habitus etc. and can be used to distinguish different types of cystic malformations [3]. Tamizkan et al. suggest to avoid CT in fetal age because of issues relating to radiation exposure to mother and foetus. The method could be used postpartum in limited occasions. According to those authors, MR imaging may play a role in prenatal imaging to provide early comprehensive information about both the anatomy and the extension of cystic hygromas [4]. Some authors like Ljubic et al. describe an extensive comparison between MR and US in the diagnosis of fetal and maternal complications [5].

Four main histological types of lymphangiomas are known and are classified as: a) capillary lymphangioma (a small, irregular mass of thin-walled dilated lymphatics in a rich cellular connective tissue stroma); b) cavernous lymphangioma (a larger spongy compressible mass of dilated lymphatics with fibrous adventitial coat); c) cystic hygroma (a large macroscopic lymphatic space result from sequestration of the central lymphatics); and d) vascolymphatic malformations [7]. The spectrum of manifestations in all types is identical and is part of the same pathologic process [8].
In prenatal and perinatal diagnosis, US still remains a primary screening method. However, recently there is an expansion of the use of MRI due to the limited sonographic anatomical orientation in some pathology. Although US is first method of choice in fetal and perinatal age, MRI MR is more sensitive for detecting fluid levels, could be beneficial in cases of complex extension due to the large field of view enblobing different anatomic regions - ex.from neck to mediastinum - and is less operator dependant [3,5]. MRI could be used preoperatively to evaluate engagement of vital structures (aorta, trachea, nerve bundles, etc.). Therefore, the role of MRI for exclusion of even partial compression of the airways by a cystic neck mass is essential, as the method could reveal more anatomical detail; this could be helpful in planning airway access procedures [4,9]. Despite the fact that most lymphangiomas occur occurs in the cervical region, they may also be found in the mediastinum, in the abdomen, the retroperitoneum and the extremities [8]. Failure to diagnose a mediastinal cystic lymphangioma could be fatal due to their increasing size and the potential to produce acute respiratory embarrassment. According the literature intrapulmonary cystic lymphangiomas are extremely rare [10].

Cystic lymphangiomas located inside the abdominal cavity are very rare, occurring most frequently in the mesenteric region and less frequently in the mesocolic, greater omental and retroperitoneal regions, small intestinal and gastric walls, and peritoneum [11]. Fetal MRI is a valuable supplemental tool to enhance prenatal ultrasound examination in the evaluation of abdominal cystic masses, providing better contrast and tissue resolution for clear anatomical details. Ultrafast imaging sequences are helpful to confirm, refine or exclude a sonographic diagnosis, thus providing useful information in prenatal counseling and management. Some authors suggest that cystic lymphangioma, seen as mesenteric masses in neonates and pediatric patients could be misdiagnosed with other mesenteric masses such as cystic teratomas [12].

Other authors describe a very rare condition as cystic lymphangioma of the spleen, detailing correlated US-CT-MRI features of both noninvasive imaging techniques for preoperative diagnosis [13]. CT and multiplanar MRIs are able to present the extent of the disease in case of renal peripelvic lymphangiomas associated with generalized lymphangiomatosis, including retroperitoneal lesions [14]. In the nuchal region, the etiology of lymphangiomas is related to inadequate drainage of the lymphatic vessel into the venous system and those cases are often associated with chromosome abnormalities. However, cystic lymphangiomas in other locations are thought to have a distinct etiology, most likely resulting from insufficient anastomoses with larger lymphatic channels.

The association with chromosomal aberrations in these cases is less distinct [15]. According to some authors describing associated findings, perinatal outcome and prognostic factors in live -born infants with cystic hygroma and lymphangioma the incidence of chromosomal abnormalities was found to be 40.3%. Their observations found as most common abnormality in nonseptated cystic hygroma trisomy 21(25%) and in septated cystic hygroma - Turner syndrome (21.1%) [16].
Findings and procedure details

From all fetal and perinatal MRI exams performed in our Institution between 2013-2015 we evaluated four cases of lymphangiomas. 1,5T MRI findings were compared to the antenatal and postnatal US and pathologic exams. MRI contributed to the accurate diagnosis of lymphatic anomalies and benefits of excellent tissue resolution, large field-of-view and relative operator independency. CT should be avoided when possible in neonatal period due to radiation exposure. In one patient clinically justified low-dose chest CT has been performed.

CASE 1

The majority of lymphangiomas occur in the neck, mainly in the posterior triangle. The first case represents a 6 week old male referred to our department with prenatal suspected vascular lesion of the neck. On physical examination swelling of the left side of the neck of the newborn was observed without any signs of respiratory difficulties or skin defects or coloration. Ultrasound performed prior to MRI depict a thick-walled polycystic formation and has been described as cystic aciniform tumor. The MR examination (single-shot fast spin echo SS-FSE and Fiesta sequences in Coronal, Axial and Sagittal plane FOV 24x18, slice thickness 5mm matrix 51x256) demonstrated a prominent heterogeneous mass on the left side of the neck (Fig.1,2,3). The lesion is lobulated, multicystic, hyperintense on T2 and hypointense on T1 sequences, well defined with mild compression of surrounding structures.

CASE 2

Although lymphangiomas are most common in the neck region, they may also be present in the axillae and medistinum. Primary mediastinal lymphangioma is extremely rare, as it usually affects the upper anterior mediastinum, almost always extending from a cervical hygroma. Acute enlargement is often associated with spontaneous intralesional hemorrhage. In this case, a 1 month old male was referred to low-dose CT with lack of normal breath sound on auscultation of the left hemi-thorax and consolidation of the left lung with displacement of the mediastinum to the right on plain film. Infiltration of the mediastinal soft tissues with compression of the left cardiac chambers and displacement of the trachea, esophagus and aorta due to the presence of the large cystic mass, almost entirely occupying the left hemithorax has been observed (Fig.4). There was no evidence of calcifications secondary to infection or bleeding. CT was conclusive in establishing the diagnosis of intrapulmonary cystic lymphangioma and allowed clinicians allows us to clearly define the extension of the lesion. However, in fetal and neonatal age and in
pediatrics CT should be avoided when possible due to radiation exposure and can be done only when clinically justified. The immediate benefits of CT outweigh the long-term risks in many settings and because of CT’s diagnostic accuracy and speed of scanning, notably removing the need for anaesthesia and sedation in young patients, it will remain in use in clinical practice for the foreseeable future [6].

CASE 3

Abdominal lymphangioma is very a uncommon congenital anomaly. Therefore, this is a an unusual case, which on prenatal sonography at 30 weeks of gestation of 26 years old woman, first pregnancy a suspicion of neuroblastoma or hepatoblastoma was suggested (Fig.5). Subsequent fetal MRI at 31 weeks gestation was performed. Findings demonstrated a large hyperintense multicystic mass adjacent to the inner liver contour with mesenteric origin. The liver was displaced towards the abdominal wall. On MRI, no solid mass was evident in addition to the cystic component of the lesion, measuring 37/24mm in the axial plane and 33/30mm in coronal view (Fig.6,7,8). No liquid levels were observed within the cysts.

CASE 4

In the routine ultrasound examination at 30 g.w. a septated cystic lesion in fetal pelvis was identified (Fig.9). No significant clinical signs and changes were detected during delivery and infancy. The next examination of the child, at the age 1,5 years (Fig.9), revealed insignificant growth of the cystic mass up to 60 mm, the largest cyst was 30 mm. No thickness of the wall or vegetations inside were present on ultrasound and MRI (Fig.10). The lesion changed its location according to prone or supine position of the patient.
Fig. 1: MR appearance of lymphangioma colli at the age of 6 weeks. Coronal SS-FSE images. Multiples internal locules, compressing adjacent structures from a left side cystic mass of the neck are visualized.

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Fig. 2: MR appearance of lymphangioma colli at the age of 6 weeks. Axial images. Multiloculated cystic mass of the neck is clearly demonstrated.
**Fig. 3:** MR appearance of lymphangioma colli at the age of 6 weeks. Sagittal image demonstrates the same cystic mass on the left side of the neck. Axial T1 fat-saturated image shows some hyperintensities within the cystic component, indicating intralesional hemorrhage (arrow).
Fig. 4: Contrast-enhanced low-dose CT scan on different levels demonstrates peripheral enhancement of a loculated, septated cystic mass in the left hemi-thorax with marked compression and displacement of major mediastinal structures. Displaced thoracic aorta by the cystic lesion is demonstrated on coronary MPR (arrow).

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Fig. 5: Prenatal ultrasonographic findings of fetal cystic abdominal lymphangioma.

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Fig. 6: Fetal MRI of abdominal lymphangioma. Sagittal FIESTA image and Gradient Echo image.

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**Fig. 7:** Fetal MRI of abdominal lymphangioma. Subsequent sagittal SS-FSE images showing a large hyperintense multicystic mass adjacent to the inner liver contour with mesenteric origin.

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**Fig. 8:** Fetal MRI of abdominal lymphangioma. Subsequent sagittal SS-FSE image and sagittal FIESTA image demonstrating the umbilical cord wrapped around the fetal neck (arrow).

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Fig. 9: Prenatal in the 32g.w. and postnatal at age 1,5 years ultrasonographic findings of cystic abdominal lymphangioma. A multiloculated cystic mass between the liver and the small bowel is visible. No abnormalities of the parenchymal abdominal organs are evident.

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Fig. 10: Abdominal MRI of lymphangioma at the age 1.5 years. Coronal T2 image clearly visualizes an intraperitoneal multicystic mass without parenchymal component and without peripheral vegetations.

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Conclusion

Fetal MRI represents a safe and useful imaging method which may confirm, complete and clarify or change the diagnosis of antenatally detected cystic masses. Prenatal MRI is a valuable adjunct to sonography for the diagnosis and characterisation of lymphangiomas and gives a precise anatomical delineation of the extent of the lesion, detects associated and concomitant pathologies and allows a better postnatal surgical planning. MRI examination could alter patient management regarding the type and time of treatment and the decision of continuation or termination of pregnancy. CT, even low-dose, should be avoided in fetal and perinatal age and in the postnatal period is rarely applicable only in cases where other imaging methods, such as MRI, are impossible to be performed.
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