

Kaasasündinud Lümfaatilised väärandid

Roman Mažeko
Kolmapäeva seminar
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Etioloogia ja patogenees

- ▶ Lümfaatiline malformatsioon = lümfangioom
- ▶ Kuuluvad vaskulaarsete „low flow“ väärendite hulka, moodustades nendest 5%
- ▶ Võivad esineda teiste vaskulaarsete malformatsioonidega
- ▶ Histoloogiliselt lümfiga täidetud tsüstilised moodustised, mis vooderdatud endoteeliga
- ▶ Harvaesinev patoloogia, arvatav esinemissagedus on 1:4000 elussünni kohta, M=N

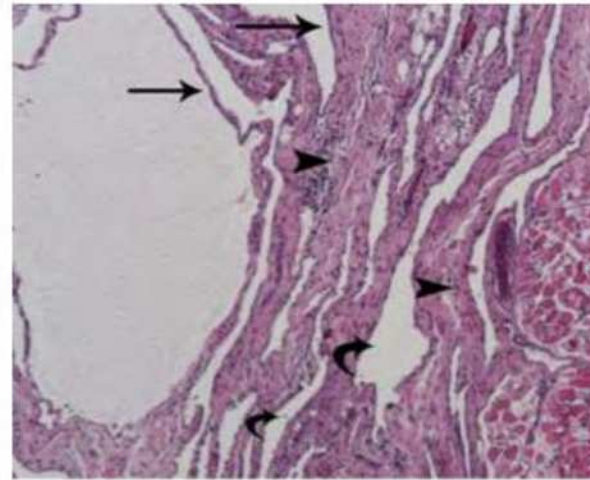


Figure 1. Pathology of lymphatic malformation. High power image of LM demonstrating multiple cysts lined by flat vascular endothelium (arrows), as well as solid elements of fibrous tissue and smooth muscle (arrowheads). Cysts in the solid matrix (curved arrows) may be so small as to be non-

| TUMORS | MALFORMATIONS | |
|-----------------------|-----------------------------|----------------------------------|
| | Low-Flow | High-Flow |
| Infantile Haemangioma | Venous Malformation (VM) | Arterial Malformation (AM) |
| | Capillary Malformation (CM) | Arteriovenous Fistula (AVF) |
| | Lymphatic Malformation (LM) | Arteriovenous Malformation (AVM) |

Table 1: ISSVA classification for vascular anomalies.

Kliiniline leid

- ▶ Esinemissagedus 70-80% kaela piirkonnas, 20% aksillaarsel, mujal alla 10%
- ▶ Kõhu piirkonnas enamasti mesenteriaalsel, harvemini organitega seotud ja retroperitoneaalsel
- ▶ Sageli asümptomaatilised
 - ▶ Sümptomid sõltuvad väärarendi asukohast ja suuruselt
 - ▶ Ilmnevad kõrvalasetsevate struktuuride kompressioonist
- ▶ Sagedasemad ägedad komplikatsioonid on veritsus ja infektsioon
- ▶ Tüüpiliselt olemas 50% juhtudest sünnil, sageli on antenataalselt diagnoositud ja kuni 90% juhtudest ilmnevad kahe esimese eluaasta jooksul

Morfoloogia

- ▶ **Mikrotsüstiline**
 - <1,0 cm tsüstid
 - „Kavernoosne lümfangioom“
- ▶ **Makrotsüstiline**
 - >1,0 cm
 - „Tsüstiline hügroom“
- ▶ **Segavorm**

Diagnostika

► UH

► MRT

| | LYMPHATIC MALFORMATION | HEMANGIOMA | CAPILLARY M | VENOUS M | ARTERIOVENOUS M |
|---------------|--|---|--|----------------------------|--|
| B MODE | Cystic Ducts of greater caliber Collapse | Solid mass Well defined Involute completely | Thin isoecogenic layer of skin and subcutaneous tissue | Spongiform Well defined | Tortuous and small-caliber vessels Poorly delimited |
| COLOR DOPPLER | NO evidence of flow | Multiple vessels. Arterial prevalence Higher echogenicit. | No vessels are appreciated in US | Venous vessels | Multiple vessels (arterial predominance) |
| PRF DOPPLER | | High flow Low resistance | | Low velocities | High flow Low resistance Higher venous flow velocities |

Diagnostika

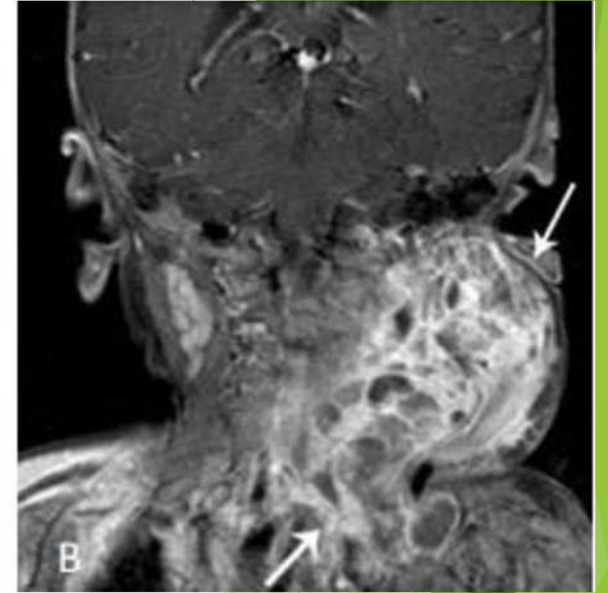
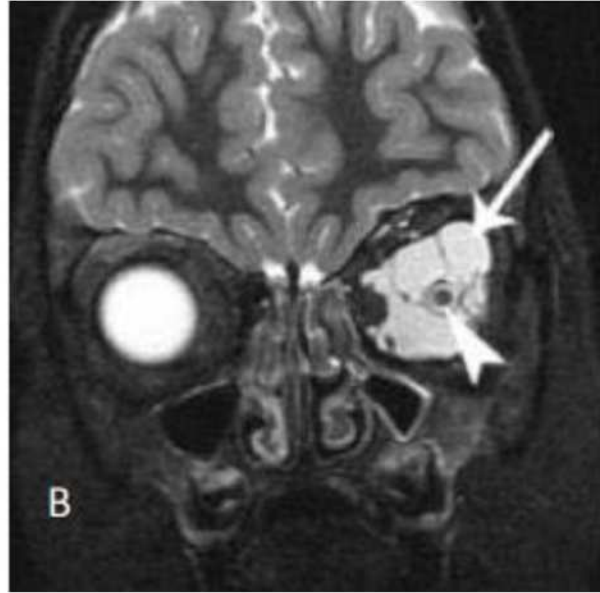
- ▶ UH
- ▶ MRT

Mikrotsüstilised lümfangiomid tavaliselt ei kontrasteeru, makrotsüstilistes kontrasteeruvad seinad ja vaheseinad

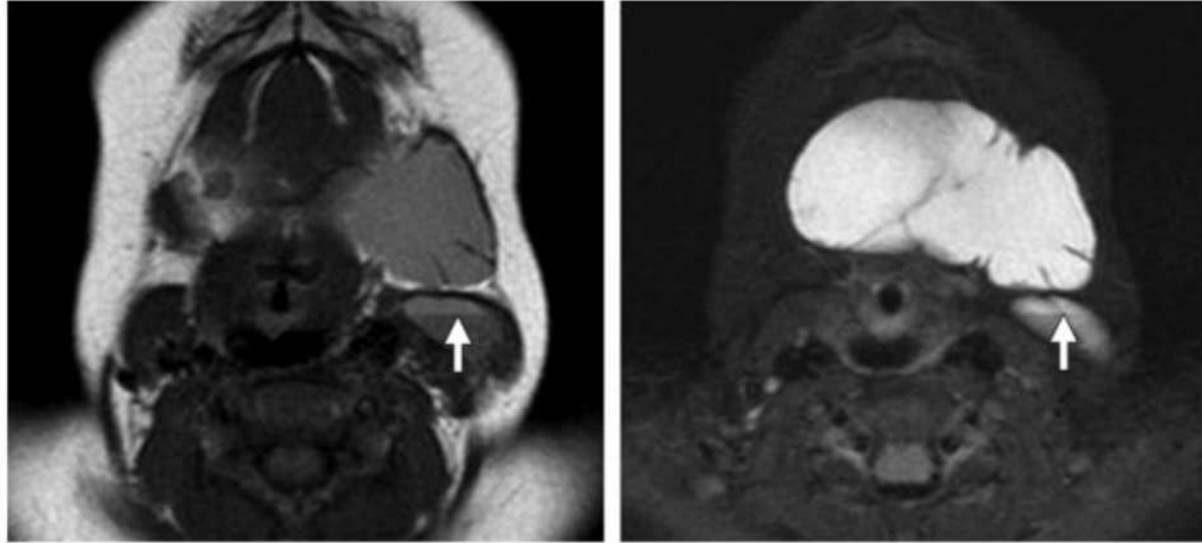
Vahepeal mikrotsüstilised annavad difuuset kontrasteerumist (halvasti eristatavate vaheseinte arvelt), kontrasteeruvad ka segatüüpi venoossed-lümfaatilised malformatsioonid

| | Lymphatic | Hemangioma | Venous | ArteriovenousM |
|---------------|---------------------|---------------------|---------------------|---------------------|
| T1 | Hypointense | Isointense | Hypo/isointense | Isointense |
| T2 | Hyperintense | Hyperintense | Hyperintense | Hyperintense |
| Post-contrast | No/ Rim enhancement | Intense enhancement | Diffuse enhancement | Intense enhancement |

Näited



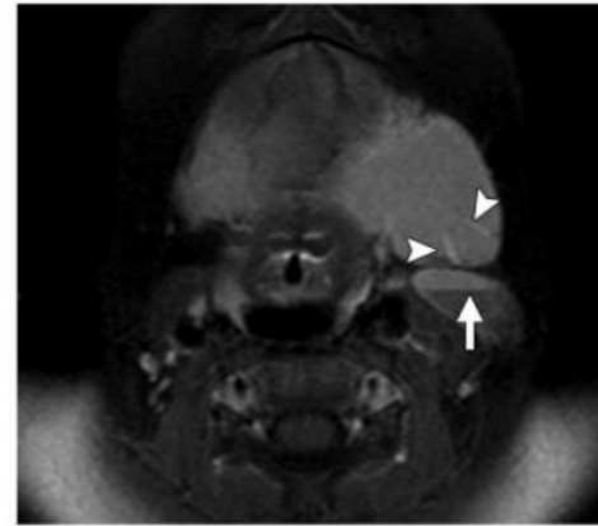
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a.

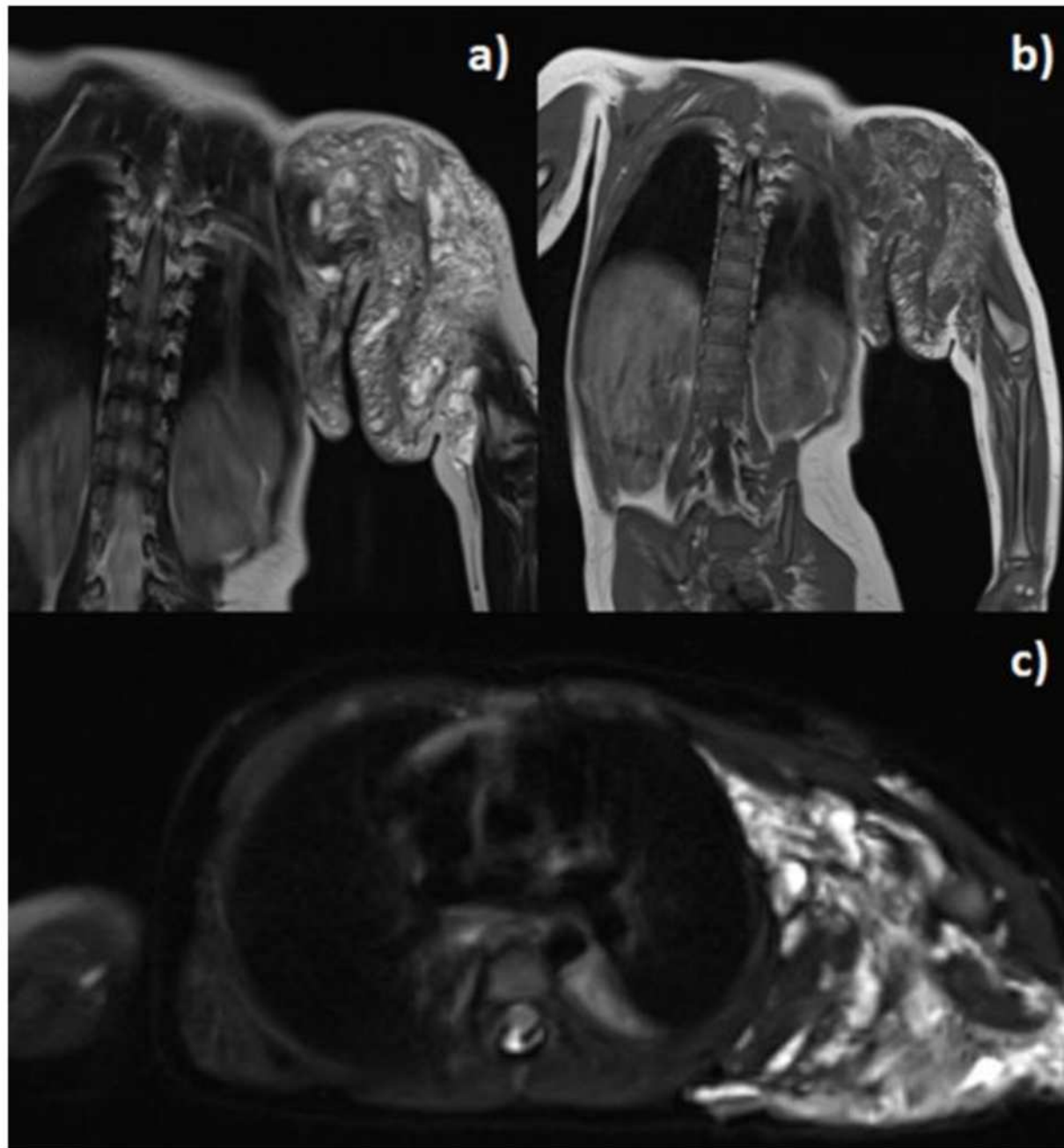
b.

Figure 5. Macrocystic lymphatic malformation in a 6-month-old infant with a swollen mass in the submandibular triangle. **(a)** T1-weighted image shows a well-defined, multilobulated, septated mass that is mildly hyperintense relative to the muscles. The increased signal intensity is most likely related to a high proteinaceous component. Note the fluid-fluid level (arrow) in the posterior component of the mass. **(b)** On a STIR image, the mass is highly hyperintense. Arrow = fluid-fluid level. **(c)** Axial gadolinium-enhanced fat-suppressed T1-weighted image shows rim and septal enhancement (arrowheads) with no enhancement of the lymph-filled spaces. Arrow = fluid-fluid level.

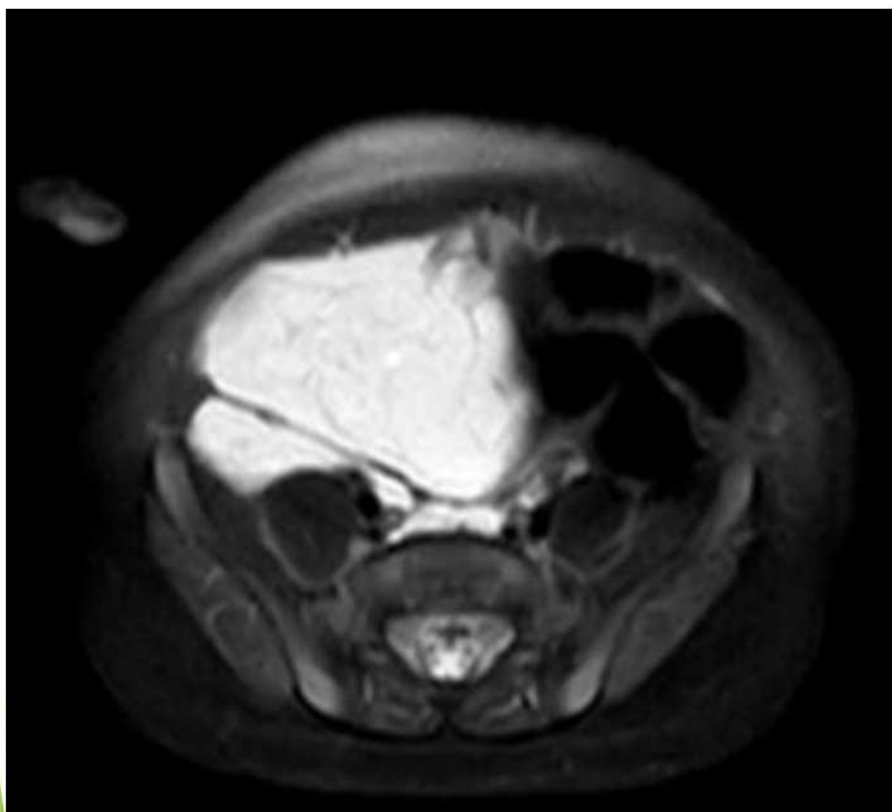


c.

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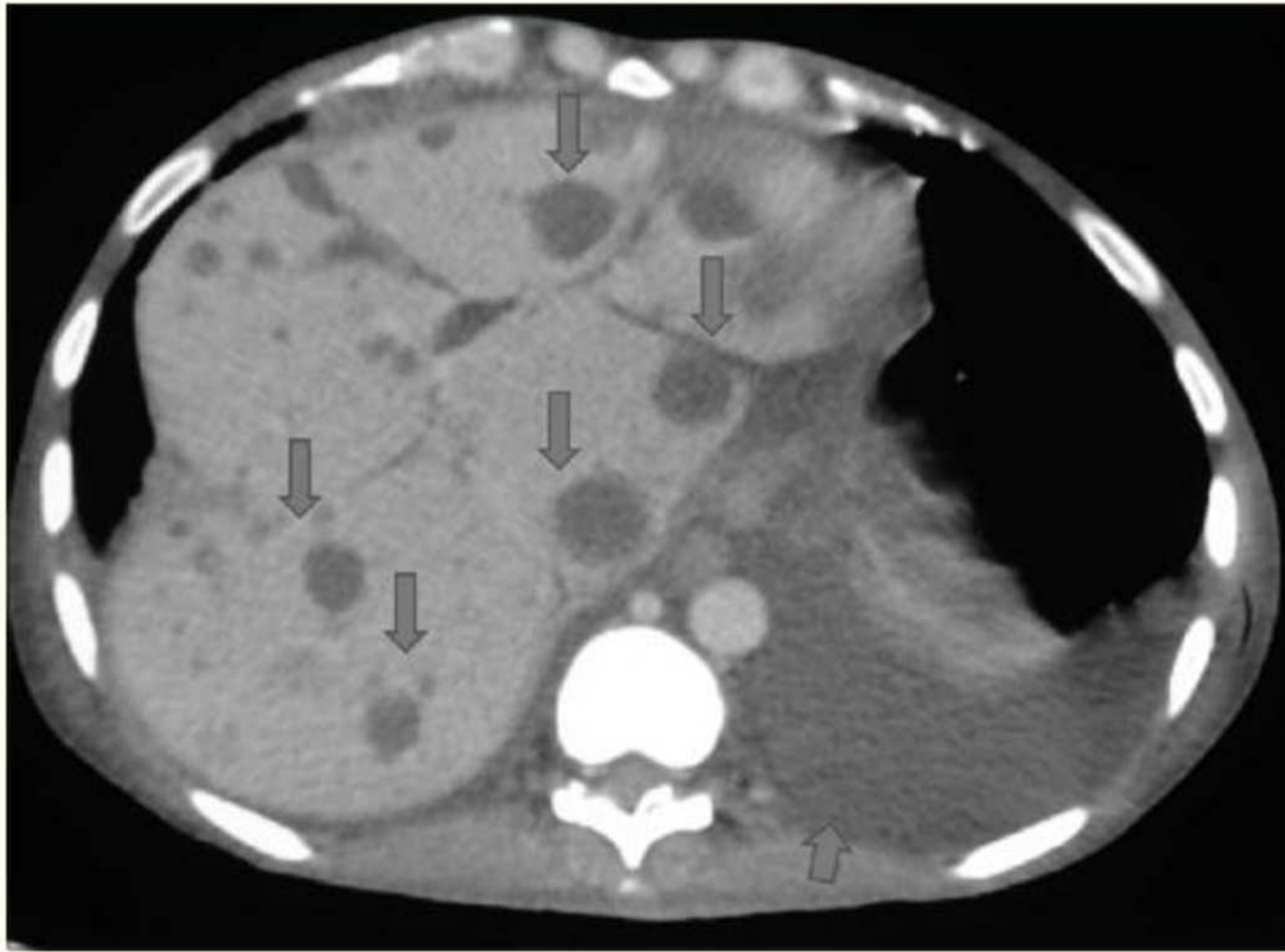


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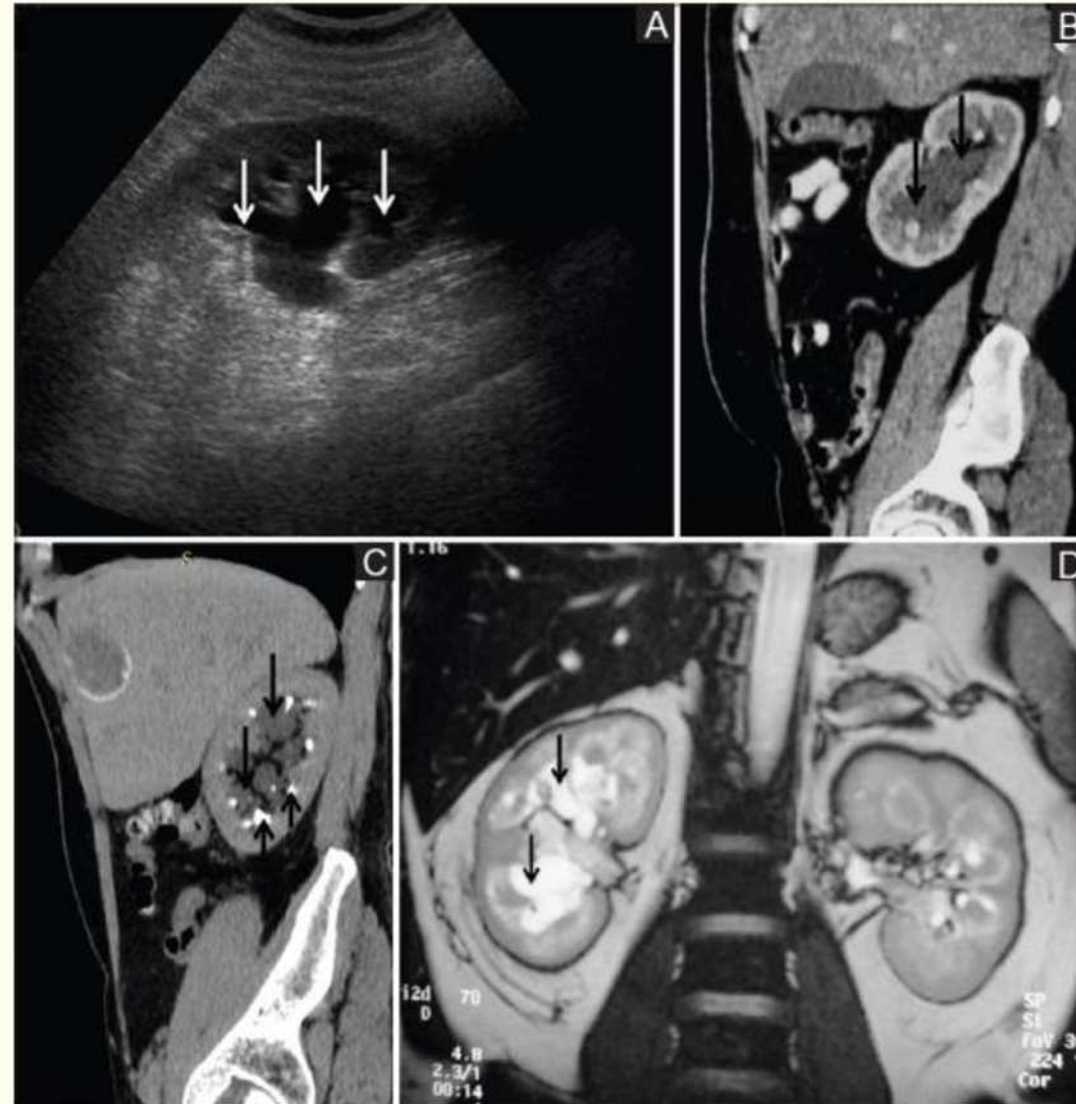
Abdominal lymphatic malformation: Spectrum of imaging findings. Anupam Lal, Pankaj Gupta, Manphool Singh, Saroj K Sinha, Sadhana Lal, Surinder Rana, and Niranjana Khandelwal. Indian J Radiol Imaging. 2016 Oct-Dec; 26(4): 423-428

Näited



Abdominal lymphatic malformation: Spectrum of imaging findings. Anupam Lal, Pankaj Gupta, Manphool Singh, Saroj K Sinha, Sadhana Lal, Surinder Rana, and Niranjana Khandelwal. Indian J Radiol Imaging. 2016 Oct-Dec; 26(4): 423-428

Näited



Dif diagnostika

- ▶ Tsüstilised hea- ja pahaloomulised tsüstilised kasvajakud
- ▶ Teised vaskulaarsed väärendid
- ▶ Vedeliku kogumikud
- ▶ Abdominoskrotaalne hüdrotsüst
- ▶ Kõhukoopa tsüstilised muutused:
 - ▶ Renaalsed: hüdronefroos, polütsüstilised neerud, põie divertikkel
 - ▶ Gastrointestinaalsed: mesenteriaalne tsüst, gastrointestinaalne duplikatsioon

Kirjandus

- ▶ Lymphatic Malformation: Radiologic-Pathologic-Therapeutic Correlation and Management Implications. William E. Shiels II, DO, MS, FAOCR. Department of Radiology, Nationwide Children's Hospital, Columbus, Ohio. J Am Osteopath Coll Radiol 2012; Vol. 1, Issue 3
- ▶ MR Imaging of Soft-Tissue Vascular Malformations: Diagnosis, Classification, and Therapy Follow-up. Lucía Flors, MD • Carlos Leiva-Salinas, MD • Ismaeel M. Maged, MD, MSc Patrick T. Norton, MD • Alan H. Matsumoto, MD • John F. Angle, MD Hugo Bonatti, MD • Auh Whan Park, MD • Ehab Ali Ahmad, MD • Ugur Bozlar, MD • Ahmed M. Housseini, MD • Thomas E. Huerta, RRT (MR) Klaus D. Hagspiel, MD. RadioGraphics 2011; 31:1321-1340
- ▶ Lymphatic Malformations in the pediatric patient. Image findings in US and MRI. J. Sanz Díaz, L. García Suárez, S. González Sánchez, L. Terán Álvarez, K. del Castillo Arango, D. Vizcaíno Domínguez, F. Arias, G. Anes; Oviedo/ES. ECR 2018
- ▶ Abdominal lymphatic malformation: Spectrum of imaging findings. Anupam Lal, Pankaj Gupta, Manphool Singhal, Saroj K Sinha, Sadhana Lal, Surinder Rana, and Niranjan Khandelwal. Indian J Radiol Imaging. 2016 Oct-Dec; 26(4): 423–428
- ▶ Kaasasündinud lümfaatilised kõhukoopa väärendid. Kati Korjus. ELS noorte lastearstide konverents Pühajärve 2020.