Splenic and retroperitoneal lymphangioma in an adult: a case report

Published on 03.12.2012

DOI: 10.1594/EURORAD/CASE.10479
ISSN: 1563-4086
Section: Abdominal imaging
Area of Interest: Abdomen
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Ultrasound
Imaging Technique: Ultrasound-Colour Doppler
Imaging Technique: MR
Special Focus: Cysts Neoplasia Case Type: Clinical Cases
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Patient: 21 years, female

Clinical History:

21-year-old woman presented with diarrhoea for 4 weeks, abdominal distension and diffuse supraumbilical pain without fever and eosinophilia. Stools had mucus but no blood or pus. She had loss of appetite for a long time. The episode was self-limiting and the standard tests only showed the incidental imaging findings.

Imaging Findings:

An abdominal ultrasound showed multiple cystic splenic lesions without septa, the largest of 1.4 cm in size (Fig. 1) and mild ascites.

Computed tomography (CT) with intravenous contrast confirmed the cystic splenic lesions (Fig.1) and also showed a retroperitoneal, well-defined, hypodense (27-30 United Hounsfield -UH-) mass surrounding great vessels from origin of celiac trunk to iliac bifurcation, of 7.4 x 1.6 cm of maximum transverse per anteroposterior diameters (Fig. 3). It encases the aorta, its visceral branches, inferior vena cava and renal veins without compressing or infiltrating them. Cystic splenic lesions and retroperitoneal mass had similar cystic appearance and they showed no internal enhancement (Fig. 1 and 3).

No abdominal lymphadenopathies were seen. Ascites of 15 UH (no haemoperitoneum) was confirmed. Magnetic resonance imaging (MRI) showed the same findings as CT (Fig. 2, 4 and 5).

There was no change of the splenic cysts or of the retroperitoneal mass after one year (Fig. 6).

Discussion:

Cystic lymphangioma is a benign neoplasm originating from a malformation of the lymphatic system [1, 2]. It generally occurs under the age of 2 years and is very rare after 20 years of age [2, 3]. There is no difference in the incidence between sexes [1, 2].

The most commonly involved sites are the neck (75%) and axilla (20%) [1, 2, 4, 5, 6, 7]. It is infrequent (5%) in the omentum, gastrointestinal track, retroperitoneum, abdominal viscera, mediastinum, lung and bone [2, 4, 8].

The two hypotheses for the pathogenesis of lymphangioma are abnormal congenital development, bleeding or
inflammation of the lymphatic system which causes an obstruction of lymphatic channels or both [2, 4].
Clinical manifestations of lymphangioma are left upper quadrant pain, abdominal distention, loss of appetite,
nausea, vomiting, palpable mass, rarely ascites, bleeding, torsion or rupture [2, 4, 5] and it can even be
asymptomatic [4, 2].

It is usually found incidentally during ultrasound or CT examinations [4].
Findings in imaging techniques contribute to a more accurate diagnosis of this condition, which can sometimes
avoid the use of invasive techniques or even surgical procedures to reach the diagnosis. Ultrasound typically shows
septated cystic structures and predominantly anechoic content, with CT attenuation coefficients similar to water [7]
usually without internal enhancement [4]. The signal pattern of lymphangiomas on MR resembles that fluid: low
signal intensity on T1-weighed images and high signal intensity on T2-weighed images. The presence of
haemorrhage or infection may alter echogenicity, CT attenuation and MR signal pattern to give a more solid
appearance [8].
Splenic lymphangiomas may be single or multiple. Usually they are small multicystic, thin-walled subcapsular
proliferations that are difficult to distinguish from either haemangioma or mesothelial cysts [8].

Our patient is a young adult woman incidentally diagnosed of a retroperitoneal and splenic lymphangioma during a
gastroenteric event. Retroperitoneal and splenic involvement makes it especially unique. We have found only one
similar case in the literature [2]. The key to diagnosis is the fact that it is a retroperitoneal cystic lesion
homogeneously hypodense (15UH) showing no enhancement after intravenous contrast and with non-invasive and
no aggressive behaviour, besides being associated with similar cystic lesions in the spleen and no associated
lymphadenopathy or other tumours.
Confirmatory invasive or therapeutic tests were dismissed in the absence of symptoms and by patient's refusal.
Subsequent controls demonstrated the stability of the findings.

**Differential Diagnosis List:** Splenic and retroperitoneal lymphangioma, Lymphoma., Retroperitoneal primary
tumour and cystic splenic lesions., Retroperitoneal and splenic hydatid cysts., Retroperitoneal cystic
lymphadenopathy and splenic cystic metastasis.

**Final Diagnosis:** Splenic and retroperitoneal lymphangioma

**References:**
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Chung SH, Park YS, Jo YJ, Kim SH, Jun DW, Son BK, Jung JY, Baek DH, Kim DH, Jung YY, Lee WM (2009)
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**Description:** Axial MDCT images with intravenous contrast show a hypodense retroperitoneal mass (yellow asterisks) that surrounds great vessels (arrows) but without causing stenosis of them. **Origin:** “Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012.”
Description: Axial MR images of the retroperitoneal mass (yellow asterisk) on sequences of chemical shift in phase (a) and on T2 WI (b and c) that show the cystic nature of the lesion. Origin: "Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012."
Description: Axial LAVA MRI sequences without (a) and with contrast on arterial (b), portal (c) and late phases (180 seconds after intravenous contrast administration)(d) that show no contrast enhancement of the retroperitoneal mass (yellow asterisk). Origin: "Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012."
**Description:** Axial image of MDCT with intravenous contrast at initial study (a and b) and a year later (c and d) showing no changes. **Origin:** “Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012.”
**Description:** Ultrasound images on B-mode (a) and Doppler-colour mode (b) show multiple splenic simple cysts without Doppler flow (blue asterisk). Axial image of MDCT with intravenous contrast (c) shows multiple splenic non septated cysts without enhancement. **Origin:** “Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012.”
Description: Axial T2 weighted images (WI) (a), T1 WI without (b) and with intravenous gadolinium on portal phase (c) MR images show multiple splenic non septated cysts without enhancement (blue asterisk). Origin: “Department of Radiology, Hospital General Universitario Jose María Morales Meseguer / Murcia 2012.”