

Cystic lymphangioma, incidental finding after pocus in outpatient pediatrics consultation

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Abstract

Cystic lymphangioma is a malformation of the lymphatic system, generally asymptomatic and rare in paediatric age.

We present the case of a 14-year-old girl who underwent a bedside ultrasound (POCUS: Point of Care Ultrasound) from the primary care paediatrics’ consultation. A hypoechoic and septate cystic mass was found in the hypogastrium. She was diagnosed of cystic lymphangioma after completing the study with magnetic resonance imaging (MRI). Treatment consisted of sclerotherapy without recurrence after 2 years of follow-up.

The routine use of POCUS from primary care can contribute to the early detection of asymptomatic abdominal lesions.

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14-year-old adolescent, model at a practical course of ultrasound for paediatric primary care at the Health Center. She had been recruited into the Periodic Child Health Screening Program, being asymptomatic at the time of recruitment and previously having a normal physical examination. As a context, she has no personal

history of interest, other than thalassemia minor, and had no previous pathological antecedents in family history.

During the abdominal systematic ultrasound examination, a cystic mass of approximately 11X6 cm, with hypoechoic content was observed, with septa inside, possibly related to the right ovary, which displaced the bladder, with vascularization at its limits (Figures 1 and 2).

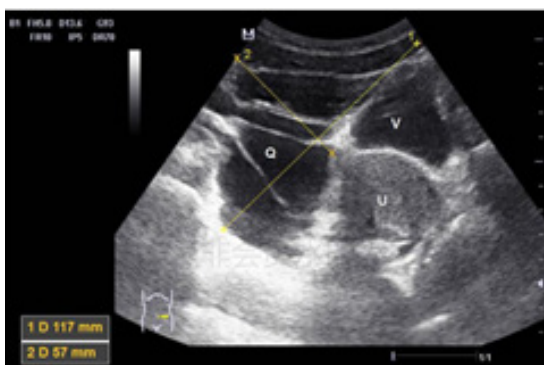


Figure 1: Abdominal US. V: bladder, U: uterus; Q: cystic mass (with complete partition walls inside).

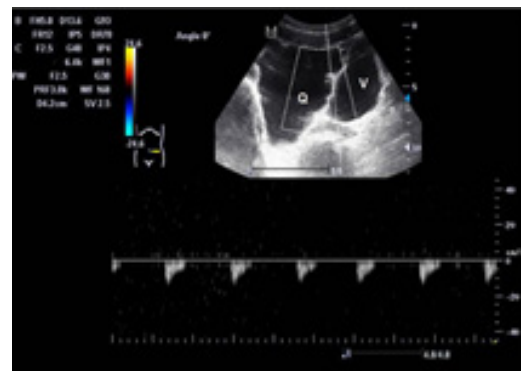


Figure 2: Abdominal US. V: bladder, Q: cystic mass (with vascularization at the base).

Completed the anamnesis, the patient did not report any alterations related to her period, did not take any treatment, she had no abdominal pain, digestive alteration and no changes in the intestinal rhythm. The only antecedent was found in his clinical history is that she had been studied six years earlier by persistent abdominal pain with the analytical studies, celiac disease, microbiological and echographic evaluation normal.

Due to the image found, it was referred to Gynaecology, with the suspicion of a mucinous cystocytoma of the ovary, although the differential diagnosis included other entities such as the cystic teratoma, a cystic lymphangioma or a mesenteric cyst.

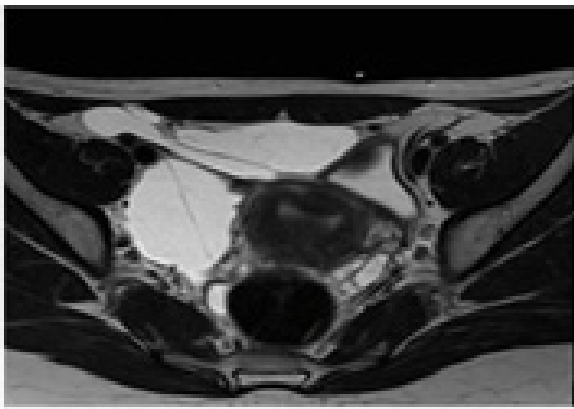


Figure 3: MRI. T2-weighted cross section.

From Gynaecology is recommended assessment by General Surgery for treatment of this lesion. Here, sclerotherapy was performed in two sessions by Plastic Surgery, using 40 cc of detergent (ethoxysclerol 2%) and 50 cc of sterile absolute alcohol, obtaining complete resolution.

Currently, the patient is completely asymptomatic and without signs of recurrence of the lesion in the subsequent ultrasound performed in our center and the hospital.

Discussion

PoCUS (Point of Care Ultrasound) is understood as the ultrasound performed at the bedside by the same doctor who treats the patient and interprets the images, at the same time of clinical care. It has the aim of answering a specific diagnostic question or guiding the diagnostic process. It is not included in the training programmes of the Family and Community Medicine specialty yet. However, in different areas of paediatrics it has already been included, for

In the Gynaecology's consult of the Hospital, a new abdominal ultrasound is performed, reported as: "paraanexial echonegative cystic formation of 117X57 mm with complete septa inside 2.6 mm with vascularization at the base. No free fluid in the lower pelvis". Study is completed with Nuclear Magnetic Resonance Imaging (MRI), which confirms the presence of the cystic lesion observed in the previous ultrasound scans. Its looks like a macrocystic lymphangioma by its characteristics, with very similar images between the ultrasound scan performed in the course of the Health Center and the MRI (Figure 3 and 4).

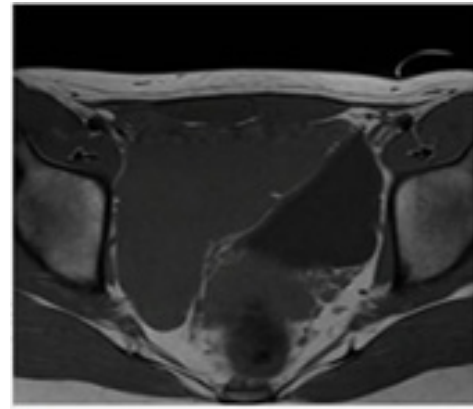


Figure 4: MRI. T1-weighted cross section.

example, lung ultrasound in the critically ill patients or in guided cardiac ultrasound. In the United States, it has an important role in paediatric emergencies [1]. In Spain, for more than 2 decades, it has been included as a routine complementary technique in various specialties such as Cardiology, General Surgery, Vascular Surgery, among others, being added to the classic specialties that previously used it: Radiology and Gynaecology and Obstetrics. In recent years, an important effort has been made to get that this technique will be used in the field of Primary Care. After a good anamnesis and physical examination with an initial clinical suspicion or an initial diagnosis, ultrasound can be very useful as a complementary tool to the first clinical interview. It is helping us to rule out or diagnose, avoiding unnecessary referrals to Hospital Care and, if required by the patient's situation, carrying out these with less dilation. It should be remembered that it can be interesting in the follow-up of certain clinical entities, as well as applied to rule out urgent pathologies, as shown in Table 1 [2].

Table 1: Core emergency ultrasound applications.

Trauma (eFAST: extended Focused Assessment with Sonography in Trauma)
Ectopic or intra-uterine pregnancy
Cardiac/haemodynamic assessment (contractility, pericardial effusion/tamponade...)
Abdominal aorta (detection of aneurysm)
Thoracic (pleural effusion, pneumothorax, hemothorax...)
Biliary (cholecystitis, choledocholithiasis)
Urinary tract (hydronephrosis)
Deep vein thrombosis
Musculoskeletal (infection, foreign bodies, masses/abscess, fractures...)
Ocular (retinal detachment, dislocations, optic nerve)
Bowel (appendicitis, bowel obstruction, pneumoperitoneum, diverticulitis, masses, hernias...)

In addition, the use of ultrasound is also very useful in interventional procedures such as thoracentesis, paracentesis, lumbar puncture, arthrocentesis, joint infiltrations, as well as peripheral and central venous access [3,4].

For all of these reason, specific training in ultrasound skills is required. Previous experience with ultrasound facilitates learning future of other locations. However, it depends on the usual practice that we have in consultations and the different patient profiles, not being the same for a child than obese adult, or a patient with ascites than a patient with a lot of intra-abdominal gas [2]. Between 2 and 320 hours are needed to learn this tool. Some areas, such as pulmonary ultrasound, will require only a few hours to acquire its management. Others, such as the cardiac ultrasound, will require a great deal of practice and theoretical understanding [5].

It should be noted that if PoCUS is not used responsibly, it can lead to overdiagnosis and with it, the emotional repercussions and consequences that this has on the patient's life. For this reason, the Primary Care physician is encouraged to apply this PoCUS technique specifically on the organ or apparatus to be studied, following the diagnostic suspicion. They cannot skip the second level of care and the radiology confirmation, because there is a risk of delaying adequate treatment. It should be used to complement the usual clinical assessment and answer specific questions [1,6,7].

In this specific clinical case, a chance finding detected through routine use of Ultrasound in Primary Care, has facilitated early diagnosis of a serious pathology (such as cyst rupture or infection), an early therapeutic approach and, therefore, a good prognosis.

Cystic lymphangioma is a congenital or acquired malformation of the lymphatic system that appears as large masses at the level of the soft tissues. Pathophysiologically it occurs due to the absence of communication between the retroperitoneal or mesenteric lymphatic tissue and the lymphatic vessels, during the embryological development of the lymphatic system or due to obstruction of the lymphatic circulation by inflammatory processes. It produces a cystic, soft and usually slow growing mass

[3,4]. It is an unusual pathology, of which 40% are diagnosed in the first year of life and 80% before the age of 6. It usually presents in the head and neck area. Being infrequent, only 2-8% of cases, are intra-abdominal, retroperitoneal or mesenteric [8,9].

In a study carried out between 1990 and 2004, it was seen that acute onset is more frequent in children and predominantly in the male sex, than in adults, which predominates in women and on chronic manner [10]. This can be explained because the children have a smaller peritoneal cavity than adults, which contributes to an earlier and acute presentation of symptoms [11].

It has no specific clinical features. It may present asymptotically and be discovered as an incidental finding, like our patient. Sometimes it is identified by palpation of a mass, which appears suddenly and grows rapidly, then stops growing or regresses spontaneously. At other times, the symptoms are due to the presence of the mass effect itself. As it grows in size, it compresses adjacent structures and produces constant abdominal pain, defence, nausea and/or vomiting. It can even be complicated by intestinal obstruction (volvulus, extrinsic compression or entrapment in the pelvis), there may be elevated leucocytosis, displacement of kidneys or ureters or compression of the abdominal vascularization. Other times the symptoms are due to other complications such as torsion, haemorrhage and/or infection of the cyst or cyst rupture, with possible secondary infection or associated peritonitis, which will require emergency surgery [11].

An image study is essential for an adequate diagnosis as well as for its management and posterior follow-up. By means of this, we will be able to have information on the size, location, extension to adjacent structures and possible complications. The ultrasound appearance of these lesions, and the typical characteristics of the fluid, will be sufficient to make a correct diagnosis in most cases. About this characteristic we find a mass consisting of multiple cysts with different sizes, everything separated by thin walls. The content can be variable according to the presence of blood and/or pus, however in uncomplicated cases it is anechogenic. Color Doppler shows little flow, appearing only in septa. In Computed

Axial Tomography (CT), we can identify unilocular masses in 25% and multilocular in 75% of the cases. The content is hypodense, compatible with water, and with septa of variable thickness, usually thin, which are reinforced with contrast and, generally, free of compression of adjacent organs. There are atypical findings, which are not uncommon, such as fatty or haemorrhagic content, calcified septa or unilocular presentations [12]. The major contribution of CT is to define the size, extent and presence of adjacent structures and complications [9,11,13]. However, both ultrasound and CT are very sensitive and quite specific studies.

MRI can also be useful, providing information similar to CT, but with less ionizing radiation, to be taken into account in the case of the study of lymphangioma in children; however, it is more expensive [11].

The histological study is fundamental in the differential diagnosis of an abdominal cystic mass. We must keep in mind normal anatomical structures (cisterna chylosa or tubes), as well as other pathologic structures such as cystic teratoma, mucinous cystadenoma, ovarian cyst, extrapulmonary bronchogenic cyst, common bile duct cyst, pancreatic and non-pancreatic pseudocysts, neoplastic peritoneal lesions with mucinous or myxoid content. Also, cavernous haemangioma when the lesion presents secondary haemorrhage, and complicated ascites, which we would identify it by the displacement of loops, the presence of focal septa or the absence of fluid in a typical site such as the cul-de-sac.

The definitive diagnosis is provided by the histopathological study of the surgical piece. Regarding the histologic characteristics of lymphangioma, it appears as a conglomerate of dilated lymphatic spaces, coexisting with lymph node formations and an important haemangiomas development [12,14].

Complete surgical resection and sclerotherapy are valid treatment options. The most experienced sclerosing agents are OK-432 (Picibanil®) and tetracyclines, which have demonstrated to be effective and safe. However, these therapies could be recurrences, especially if the resection is incomplete, which occurs more frequently in lymphatic malformations involving the root of the mesentery [15].

Total excision was performed in 77.4% and partial excision in 20.7% with recurrence rates of 11.8% and 52.9%, respectively. The incidence of postoperative complications was 31.3%. Despite being benign lesions, lymphatic malformations have a high morbidity rate due to complications of the disease and its management. However, complete excision offers the greatest chance of complete cure [16].

Conclusion

Lymphangioma is a rare pathology, although more common in children. It is clinically asymptomatic or it manifests as a non-specific acute abdominal pain, which can lead to complications such as compression of adjacent structures, obstruction, torsion,

haemorrhage and/or peritonitis. It can be diagnosed by imaging tests such as ultrasound, CT and/or MRI. The latter two being used to confirm the extension and situation with respect to adjacent structures. Regarding treatment, the best option is complete resection of the cystic mass. Ultrasound controls can be performed a posteriori to check the growth of the mass.

The use of PoCUS more routinely by Primary Care (family doctors and paediatricians) after appropriate training could provide a greater number of early diagnoses of pathologies such as we showed in our clinical case, Also could make referrals to Hospital Care more efficiently. In addition, it serves as an interesting point for follow-up and assessment of the absence of recurrence after definitive treatment.

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