

Isolated atrial fibrillation is an infrequent arrhythmia in the pediatric population, especially in the absence of organic heart disease of a probably multifactorial etiology. Both of the cases we present here corresponded to a first episode of AF that was haemodynamically insignificant in patients with no known history of heart disease.

Persistent left superior vena cava, diagnosed in case 1, has been associated with the development of supraventricular arrhythmias, including AF. Radiofrequency catheter ablation of these connections can prevent their recurrence. Hsu et al. described 5 cases of AF where electrophysiology testing identified persistent left superior vena cava as the source arrhythmia.^{1,2}

To our knowledge, an association between AF and bicuspid aortic valve, a defect detected in case 2, has not been previously reported, and therefore the coexistence of these 2 disorders could be coincidental and unrelated to the disease process.

Intense physical activity may cause electrical and structural changes in the atria that would promote the development of an episode of AF. In both our patients, AF was associated with physical activity. There is also evidence of an association between chest trauma with commotio cordis and AF.³

Cases of isolated familial AF have been reported in the literature in association with genetic mutations affecting multiple ion channels and circulating hormones such as atrial natriuretic peptide. However, the diagnostic yield of genetic testing for this type of arrhythmia is currently not known.^{4,5} In this regard, the presence of idiopathic AF in the father of case 1 may have increased the risk of AF in our patient. In addition, consumption of some recreational substances, such as cannabis, can trigger AF. In the cases presented here, we ruled out the consumption of arrhythmogenic substance use.

In a series of 1750 cases of isolated AF in children, the only risk factors identified were obesity with a BMI above the 95th percentile, male sex and age greater than 14 years.⁶

At present there is no consensus regarding the first-line antiarrhythmic drug for treatment of AF in the pediatric age group. However, drugs used to control heart rate (such as beta blockers) are the most widely used agents. In the 2 cases presented here, treatment with flecainide was chosen

with the aim of achieving cardioversion. In cases refractory to pharmacotherapy, electrical cardioversion and radiofrequency ablation of the sources of atrial arrhythmia are effective alternatives.⁶

Lastly, the use of anticoagulant therapy was ruled out in both cases due to the low risk of thrombosis and the early recovery of the sinus rhythm. In any case, there are no guidelines providing the indication of prophylactic treatment for prevention of stroke in this population.

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Mesenteric lymphatic malformation: A rare cause of an acute abdomen[☆]



Malformación linfática mesentérica: una causa poco frecuente de abdomen agudo

Dear Editor:

Mesenteric lymphatic malformations, also known as lymphangiomas or cystic hygromas, are rare lesions originating from the proliferation of lymphatic vessels in the mesentery.¹ They mainly affect children aged less than

5 years and the most frequent location are the head, neck and axillary region, with intraabdominal locations accounting for less than 1–5% of cases.^{1–4} The clinical presentation varies depending on the size and location of the lesion from the absence of symptoms to acute abdomen (abdominal pain, abdominal distension, signs of peritoneal irritation).^{4,5} Although recent articles have described management with percutaneous sclerotherapy,¹ the first-line treatment continues to be complete surgical resection.^{3,4} In this article, we describe the approach to the diagnosis and treatment of 2 cases of mesenteric lymphatic malformation managed in our unit.

Case A

Boy aged 3 years with an unremarkable previous history that sought care in the emergency department for diffuse

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abdominal pain of 3 days' duration that exhibited guarding on palpation of the left side of the abdomen. He had been assessed 1 week prior due to a similar episode associated with fever and vomiting that had been managed with conservative treatment. The abdominal X-ray revealed a mass effect in the left side of the abdomen, and the abdominal ultrasound the presence of a large intraabdominal cystic lesion (Fig. 1A and B). As his condition was deteriorating, the patient underwent an exploratory laparotomy that revealed extensive cystic tumor dependent on the omentum and anchored to the greater curvature of the stomach, which was completely resected (Fig. 1C). The findings of the gross and histological examination led to diagnosis of lymphangioma with haematic content.

Case B

Girl aged 4 years assessed in the emergency department for colicky abdominal pain of 5 day's duration associated with vomiting. The patient had had similar episodes before that had resolved satisfactorily with enemas. She underwent an ultrasound examination that revealed a very larger multicystic lesion in the left hypochondrium. In this case, the patient was clinically stable, which allowed performance of a magnetic nuclear imaging (MRI) scan to complete the investigation and schedule surgery. The scan revealed a cystic lesion at the level of the mesentery in close contact with the tail of the pancreas (Fig. 1D and E). The patient underwent laparoscopic-assisted resection of the lesion with

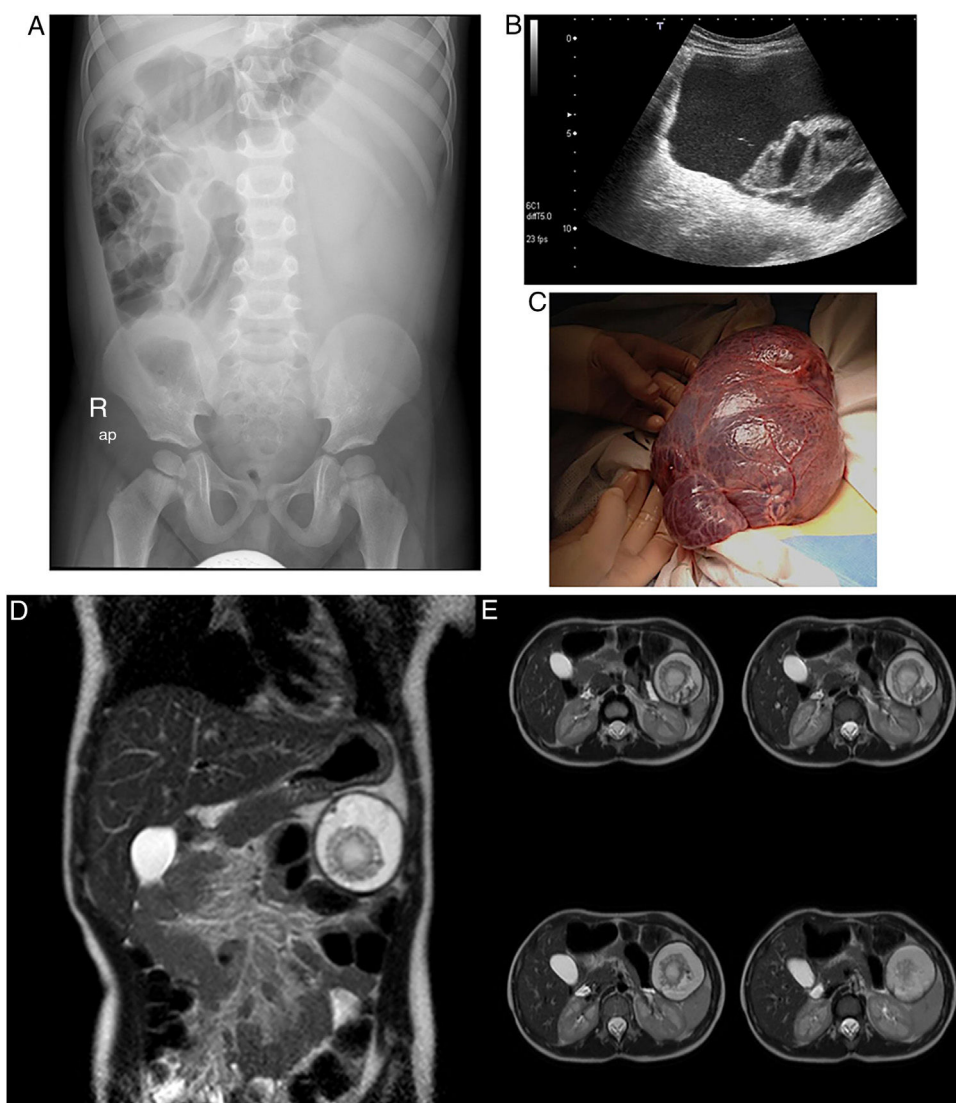


Figure 1 (A–C) Radiological and surgical findings in case A. (A) Abdominal X-ray with the patient in the supine position revealing a mass effect in the left abdomen with displacement of intestinal loops toward the right abdomen. No signs of bowel obstruction. (B) High-resolution grayscale ultrasound image revealing a cystic lesion with echogenic contents, internal septa and another pseudosolid lesion within. (C) Intraoperative image revealing a very large ($15 \times 9 \times 3$ cm) multicystic mass pink in color. (D and E) Radiological findings in case B. Multiphasic contrast-enhanced MRI images in the coronal (D) and axial (E) planes following intravenous administration of gadolinium. Round lesion measuring approximately 5.3 cm along all 3 axes located in the left hypochondrium with contents of probable hemorrhagic origin demarcated by a hypointense shape of a capsule or pseudocapsule. No evidence of organ dependence, edema or infiltration of adjacent structures.

Table 1 Basic characteristics of described patients.

Case	Age	Sex	Symptom	Location	Intervention	Approach
A	3 years	Male	Abdominal pain	Large curvature of stomach	Urgent complete resection	Laparotomy
B	4 years	Female	Abdominal pain	Ilium	Elective complete resection	Laparoscopy

bowel anastomosis. The pathology report described a benign cystic vascular lesion with immunohistochemical features compatible with lymphangioma (Table 1).

The lesions formerly known as lymphangiomas are currently classified under the term *lymphatic malformations* and described as cystic vascular anomalies rather than tumors of the lymphatic vessels. They may present in isolation or in the context of systemic disease.⁶

They are typically present at birth and grow with the child, becoming increasingly noticeable. This leads to diagnosis by age 5 years in 60–90% of cases, and diagnosis in adulthood is rare.^{3,4}

Most cases (95%) involve the lymphatic vessels in the head, neck or axillary region, while abdominal involvement (in the mesenteric or retroperitoneal regions) is rare.^{1–6} Nevertheless, given the heterogeneity in the terminology used historically to describe them (lymphatic malformation, lymphangioma, cystic hygroma, etc.) and the fact that it may affect both the pediatric and the adult populations, it is very likely that its incidence is somewhat underestimated.⁶

The most widely accepted theory on the etiology of these malformations is the “blind sac” hypothesis, according to which these are congenital lesions resulting from sequestrations of lymphatic tissue that do not communicate with the lymphatic system during embryonic development, leading to dilatation of the vessels and formation of a cystic mass as the lymph accumulates.⁴ However, different authors of articles in the literature have proposed an association with specific factors such as radiation exposure, abdominal trauma, surgery or inflammation, among others.^{3,4}

Most identified cases are painless masses growing in proportion to the child, but in some instances these malformations can cause complications such as infection, rupture, bowel obstruction, volvulus or spontaneous bleeding,^{2,6} making it a surgical emergency, as occurred in case A presented here. The latter cases typically present as acute abdomen, and the most frequent symptom is abdominal pain.

Ultrasonography is the gold standard of imaging for the initial evaluation and can be supplemented with computed tomography or MRI for a more thorough definition of the lesion if the condition of the patient allows it. In many cases, the definitive diagnosis is based on the histological findings.^{4,6}

Surgical treatment involves the complete resection of the lesion and is associated with favorable long-term

outcomes and a low risk of malignant transformation or recurrence.^{1,3}

Although lymphatic malformations manifest infrequently as acute abdomen, they should not be excluded from the differential diagnosis of this clinical picture, especially in case of detection of a palpable abdominal mass.

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