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Case Report

Retroperitoneal lymphatic malformations: A case report ☆

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ABSTRACT

Retroperitoneal lymphatic malformation is a rare benign vascular malformation, of various locations, which can reveal itself at any age in its abdominal localizations. The retroperitoneal localization of this malformation is extremely rare. The clinical symptomatology is polymorphic depending on the lesion volume and the presence or absence of complications. The diagnosis is evoked by the liquid character of the retroperitoneal mass on ultrasound, CT scan and abdomino-pelvic MRI, brought during surgery and confirmed by the histological study of the surgical specimen. The treatment of choice is complete surgical removal of the mass.

Introduction

Retroperitoneal lymphatic malformation is a rare benign vascular malformation of the lymphatic system, exceptional in adults, affecting more frequently the craniofacial region, neck, thorax and axillary region [1,2]. Retroperitoneal localization is rare. We report a case of retroperitoneal lymphatic malformations revealed by a large abdominal mass in a 20-year-old woman, with study of different imaging features including MRI findings.

Case report

A 25-year-old single woman with no medical or surgical history consulted for the appearance of an abdominal-pelvic mass that had been progressively increasing in volume for 2 months, associated with diffuse abdominal pain. The clinical examination revealed a mass occupying almost the entire abdomen, painful and nonpulsatile to palpation with a slight abdominal contracture. The biological workup was normal. Abdominal ultrasound revealed a large abdominal cystic

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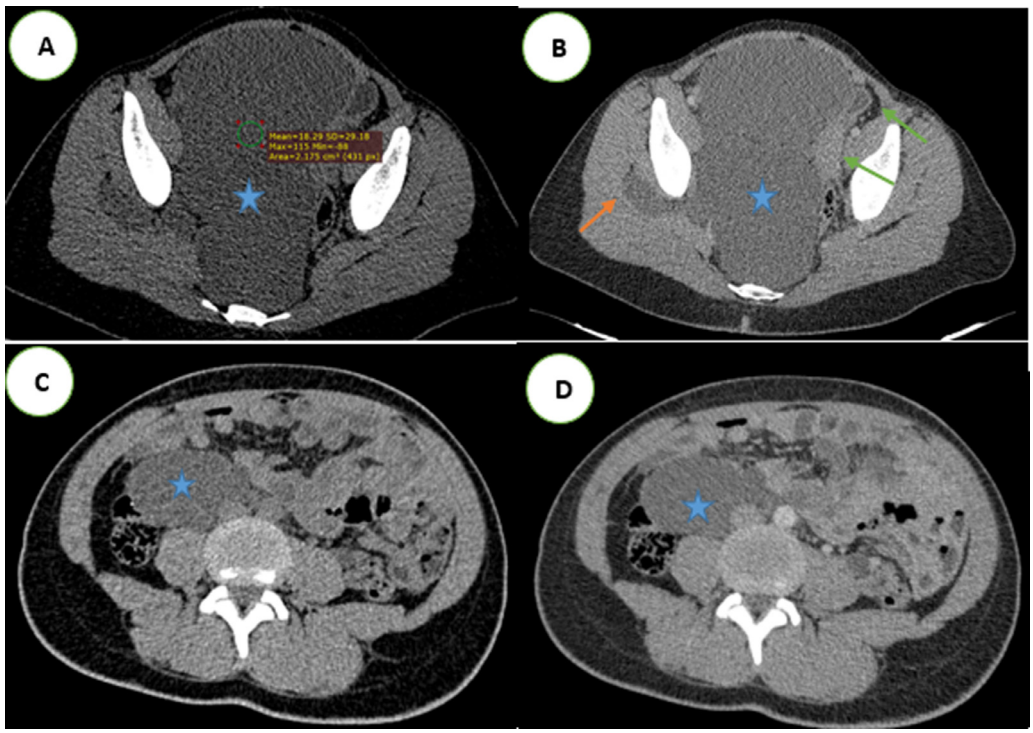

Fig. 1 – Abdominal-pelvic CT in axial sections without (A, C) and after contrast agent administration at portal time (B, D): shows a large cystic mass (blue star) molding the neighboring structures, pushing back the bladder and the uterus (green arrow) and insinuating itself between the right gluteal muscles (orange arrow).

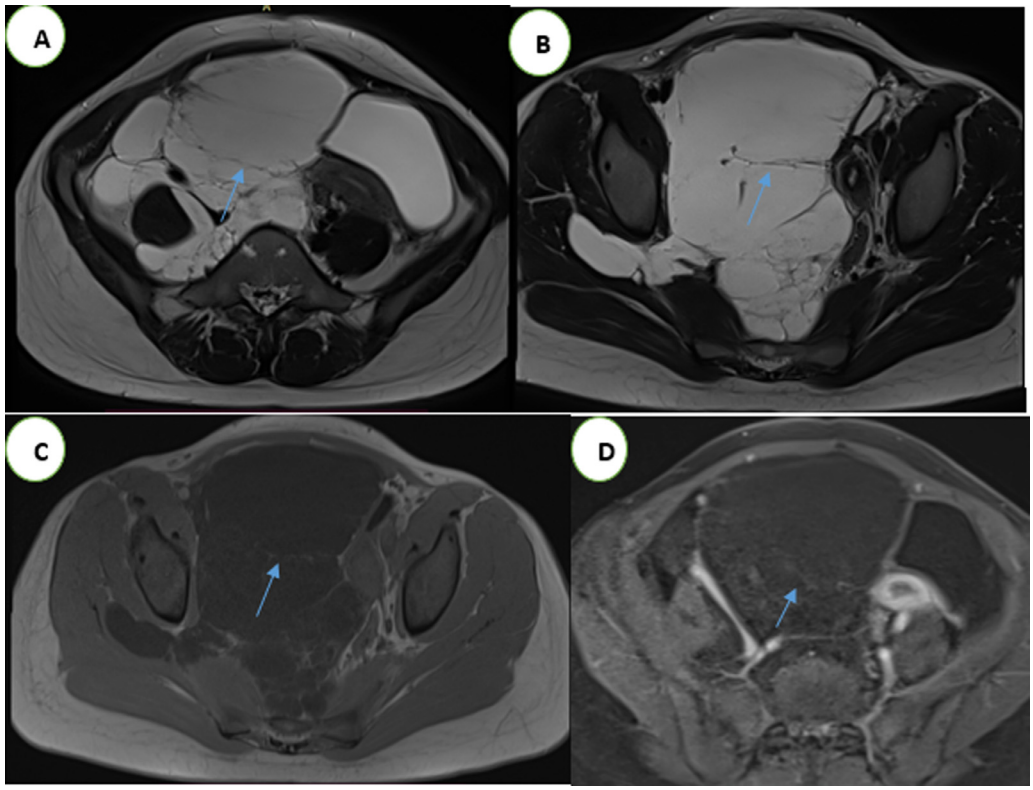

Fig. 2 – Abdominal and pelvic MRI in axial sections, T2 sequence without FATSAT (A, B), T1 without FATSAT (C) and T1 FATSAT with Gadolinium administration (D) shows a voluminous cystic mass with fluid signal and fine septa (blue arrow).

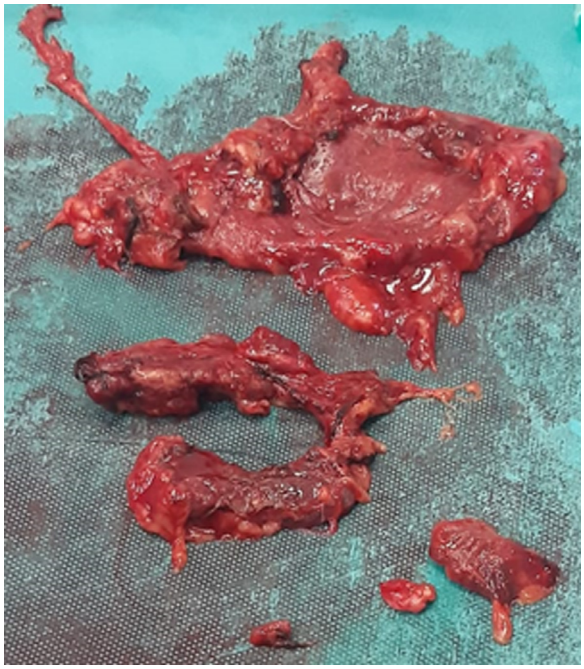

Fig. 3 – Macroscopic cystic appearance of the mass.

mass extending to the pelvis and displacing the bladder and uterus to the left without any suspicious location or associated peritoneal effusion. The abdominal CT scan was in favor of a large non enhancing lobulated fluid attenuating mass (HU +18) measuring 214 × 195 × 130 mm in abdominopelvic cavity with few fine internal septations. No evidence of solid components within. It has abutted adjacent abdominal organs (rectum, sigmoid, bladder, and uterus) with maintained fat plane and has insinuated between the gluteal muscles, it compressed the excretory tracts with bilateral ureteropyleic dilatation and encompassed the right iliac vessels which remained permeable (Fig. 1). MRI showed the same retroperitoneal mass in T1 hypo signal, frank T2 hyper signal, enclosing thin partitions in T2 hyposignal, moderate T1 hyper signal (Fig. 2). The patient underwent a complete surgical removal of the cystic lesion by laparotomy (Fig. 3). The postoperative course was unremarkable. The histological study of the surgical specimen was in favor of a retroperitoneal lymphatic malformation (Fig. 4).

Discussion

Retroperitoneal lymphatic malformation is a rare benign vascular malformation that originates in the lymphatic vessels. The regions frequently affected by this malformation are the craniofacial, cervical, thoracic and axillary regions. Abdominal localizations are rare, dominated by mesenteric and epiploic involvement [3]. The retroperitoneal form is extremely rare and less frequent compared to the mesenteric localization [4]. This malformation is most often found in children and is more exceptional in adults, representing 7% of abdominal cysts [5].

From a pathophysiological point of view, the most probable theory for the development of retroperitoneal lymphatic malformation is a congenital malformative origin due to a defect in the connection between the retroperitoneal lymphatic chains and the venous system, resulting in the formation of an isolated lymphatic bud that will evolve into a cyst [6–8]. The second theory consists rather of an acquired origin by obstruction of the lymphatic vessels secondary to traumatic, inflammatory or degenerative mechanisms [6]. This theory is unlikely and is increasingly abandoned.

Retroperitoneal lymphatic malformation is often asymptomatic, discovered incidentally on radiological imaging or during surgery for another condition. Clinical symptoms such as abdominal pain or heaviness with an abdominal mass may be related to the volume of the tumor, that is, the cyst becomes large enough to cause compression of the surrounding structures, as in the case of our patient. The clinical signs may also be related to complications such as rupture, torsion, superinfection or intracystic hemorrhage. The risk of malignant degeneration is exceptional [4].

Radiologically, the typical appearance of this malformation on ultrasound is a well-limited, uni or multilocalized cystic mass with fluid content, containing septa of variable

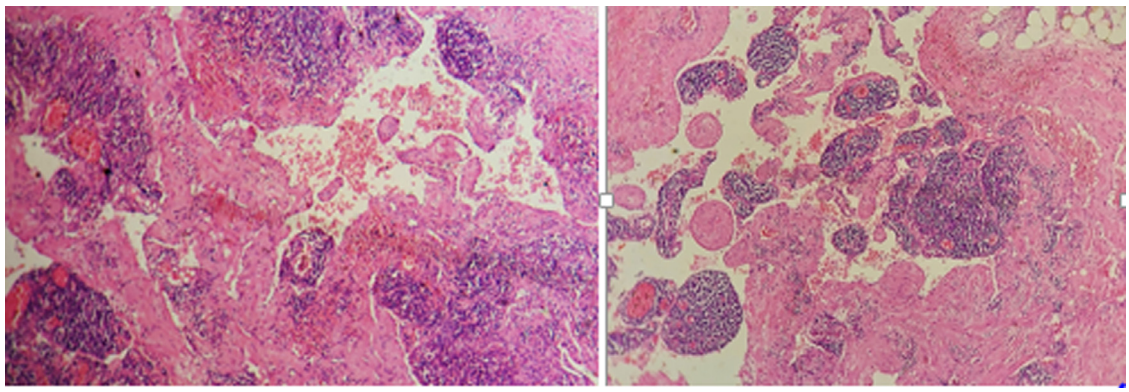

Fig. 4 – Histological appearance of cystic lymphangioma (Magnification x 40): Lymphatic cystic spaces lined with endothelial cells containing red blood cells and lymphocytes, and a fibrous stroma showing lymphoid aggregates.

thickness and often presenting fine hyperechoic echoes [9]. Atypical aspects are not uncommon, and therefore abdominal CT remains important to determine the density of the mass, its size, its extent and its relationship with the surrounding organs. The typical CT appearance is that of a cystic mass, well circumscribed, of liquid density, with a thin wall enhanced after contrast agent administration. Involvement of more than one retroperitoneal compartment in the form of a poorly circumscribed cystic mass, molding and insinuating itself between adjacent structures, with partitions of variable thickness are characteristic of cystic lymphangioma. In our case, the cystic mass was voluminous, molding the neighboring structures without infiltrating them, insinuating itself between the gluteal muscles, and it contained fine partitions. MRI provides a more precise diagnosis. The cystic nature of the mass can be appreciated by the signal characteristics shown by the mass in MRI- that is, lesion shows low signal intensity on T1W images and high signal intensity on T2W images with few fine internal septations within, as shown on our patient's abdominal and pelvic MRI images. It also allows to appreciate the perivascular extension [8].

The differential diagnosis is with peritoneal inclusion cyst and hydatid cyst (in endemic countries), and only the histological study of the surgical specimen after surgical resection allows a definite diagnosis to be made by demonstrating lymphatic cystic spaces lined with endothelial cells with fibrous stromata containing lymphoid aggregates.

Surgical resection should be as complete as possible to avoid recurrence and complications such as superinfection, rupture, bleeding or progression [10].

Conclusion

Retroperitoneal lymphatic malformation is a rare benign malformation of the lymphatic system, with progressive development, noninfiltrating and non-metastatic. The clinical symptomatology is not very specific, the diagnosis is evoked by abdomino-pelvic imaging (ultrasound, CT, and MRI) and confirmed by the histological study of the cystic lesion after its complete surgical excision which constitutes the treatment of choice.

Patient consent

Informed consent for publication was obtained from patient.

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