CIRUGÍA ESPAÑOLA

www.elsevier.es/cirugia

Letter to the Editor

Lymphatic Malformations: Diagnostic-Therapeutic Management and Current Nomenclature☆

Malformaciones linfáticas: manejo diagnóstico-terapéutico y nomenclatura actual

To the Editor:

We are writing this letter in reference to the article published in November 2020 by Rodríguez Alvarado et al.,[1] ''Bilateral chylothorax and chylous ascites resulting from the spontaneous rupture of a retroperitoneal lymphangioma'', which we have read with great interest. We would like to make some comments about the text.

First of all, we have observed that both in the title of our colleagues' study, as well as on several occasions throughout the text, the term *lymphangioma* is used to refer to the patient's congenital lymphatic malformation. In 1996, the International Society for the Study of Vascular Anomalies (ISSVA) published its first standardized terminology for vascular malformations and tumors. This classification, which was improved and expanded in 2014 and more recently in 2018, is currently used by all professionals specialized in the management of vascular anomalies, both in pediatric and adult patients (Table 1).[2] In this classification, the use of the term 'lymphangioma' has been discontinued. Therefore, the malformation diagnosed in the patient is a common lymphatic malformation (LM), which could be macrocystic, microcystic or mixed (with both components), depending on its morphology.

Secondly, the patient presented initially had an imaging study using computed tomography (CT) and later underwent three surgical procedures (hepatic segmentectomy and bilateral thoracic lymphatic duct ligation) in order to resolve the bilateral chylothorax and chylous ascites presented. Once the previous procedures had been carried out, the authors report that all possible medical etiologies were ruled out and a magnetic resonance (MR)-guided lymphography was performed, which diagnosed retroperitoneal LM. In this context, we agree with Dr. Rodríguez et al. in that chylothorax is the most common cause of neonatal pleural effusion. Its origin is usually postoperative or traumatic, and its spontaneous appearance is exceptional, as among the adult population. Therefore, when confronted with a healthy 37-year-old man with no relevant medical-surgical history, we believe it would have been appropriate to conduct a complete initial study, aimed at ruling out an associated LM and other causes. This approach could have avoided the previously mentioned surgical procedures.

Once the accurate diagnosis of retroperitoneal LM had been made, the authors proposed correct specific management, initiated with sirolimus. The discharge from the lesion then decreased considerably, in a non-resolutive manner. Therefore, low doses of radiotherapy directed at the LM were administered. The authors do not mention the dosage of the sirolimus, or its duration. The efficacy of this drug in LM is not immediate, and we therefore feel that a strict definition of the administration and dose, as well as duration, would have been of great interest to the reader. Also, given its controversial potential for malignancy in vascular malformations, radiotherapy is a therapeutic alternative whose use in recent decades has decreased in this type of lesion.[3]

We agree with the authors that the retroperitoneal and perivascular location limits the surgical excision of the lesion. However, there are other alternatives for its management that are becoming increasingly accepted, such as MRI-guided percutaneous sclerosis.[4,5]

In conclusion, we would like to congratulate the authors for their contribution of a rare clinical case. The interesting world of vascular anomalies is in a moment of emergence and scientific advancement. The exponential progressive increase in studies describing this type of anomaly is a true reflection of

DOI of original article: http://dx.doi.org/10.1016/j.cireng.2020.10.011

☆ Please cite this article as: Casal-Beloy I, García-Novoa MA, Lema Carril A, Gómez Tellado MA. Malformaciones linfáticas: manejo diagnóstico-terapéutico y nomenclatura actual. Cir Esp. 2021;99:324–325.

Table 1 – Classification of Vascular Anomalies by ISSVA (Last Updated 2018).

Vascular Anomalies				
Vascular Tumors	Vascular Malformations			
-Benign -Borderline (locally aggressive) -Malignant	Simple	Combined	Major vessels	Associated with other anomalies
	-Capillary -Lymphatic -Venous -Arteriovenous -Arteriovenous fistula	-Capillary-venous M -Capillary-lymphatic M -Capillary-arteriovenous M -Lymphatic-venous M -Capillary lymphatic-venous M -Capillary-lymphatic arteriovenous M -Capillary-venous-arteriovenous M -Capillary-lymphatic-venous-arteriovenous M	Canal type or trunk vascular malformations.	-Klippel-Trenaunay syndrome -Parkes-Weber syndrome -Servelle-Martorell syndrome -Sturge-Weber syndrome -Limb CM + congenital non-progressive limb overgrowth -Maffucci syndrome -Macrocephaly – CM -Microcephaly – CM -CLOVES syndrome -Proteus syndrome -Bannayan-Riley-Ruvalcaba syndrome -CLAPO syndrome

this current growth. However, one of the main obstacles in understanding and managing these malformations continues to be poor adherence to the standardized nomenclature, which makes it difficult to read and comprehend the existing literature on the matter. Therefore, in order to improve and contribute to current developments in LM, rigorous use of the appropriate nomenclature should be a priority.

REFERENCES

1. Rodríguez Alvarado I, Gómez Hernández MT, Temprado Moreno V, Herráez García J, Jiménez López M. Bilateral chylothorax and chylous ascites resulting from the spontaneous rupture of a retroperitoneal lymphangioma. Cir Esp. 2020;98:563–5.
2. Classification of Vascular Anomalies© 2018. International Society for the Study of Vascular Anomalies. https://www.issva.org/classification.
3. Rudman RA, Clark WJ. A large vascular malformation of the tongue treated with radiation therapy. J Oral Maxillofac Surg. 1997;55:509–14.
4. Dubois J, Thomas-Chausséé F, Soulez G. Common (cystic) lymphatic malformations: current knowledge and management. Tech Vasc Interv Radiol. 2019;22:1–14.
5. De Maria L, De Sanctis P, Balakrishnan K, Tollefson M, Brinjikji W. Sclerotherapy for lymphatic malformations of head and neck: systematic review and meta-analysis. J Vasc Surg Venous Lymphat Disord. 2020;8:154–64.

Isabel Casal-Beloy[a,*], María Alejandra García-Novoa[b], Ana Lema Carril[a], Manuel Adolfo Gómez Tellado[a,c]

[a]*Servicio de Cirugía Pediátrica, Complejo Hospitalario Universitario A Coruña, Spain*
[b]*Servicio de Cirugía General y del Aparato Digestivo, Unidad de Mama, Complejo Hospitalario Universitario de A Coruña, Spain*
[c]*Servicio de Cirugía Pediátrica, Unidad de Anomalías Vasculares, Complejo Q1Hospitalario Universitario A Coruña, Spain*

**Corresponding author.* isabelcasalbe@gmail.com (I. Casal-Beloy).

2173-5077/
© 2021 AEC. Published by Elsevier España, S.L.U. All rights reserved.