

Mantle Cell Lymphoma with Atypical Radiologic Presentation: Case Report

Linfoma de células del manto con presentación radiológica atípica: Presentación de caso

Juan Esteban López Amaya¹
 Beatriz Molineros Arévalo¹
 Carlos González Vásquez¹
 Ana María Alvarado Benavides²
 Rafael Llamas Otero²
 Ricardo Uribe González²
 Carolina Echeverri Jaramillo³



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Palabras clave (DeCS)

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Summary

Mantle cell lymphoma is a Non-Hodgkin Lymphoma (NHL). In cases of disseminated disease, lymphadenopathy, splenomegaly, B-symptoms and skin disease are present. Lymphoma affecting the spleen has several radiologic presentations, ranging from normal appearance, to splenomegaly or multiples focal solid lesions. Cystic presentation of lymphoma is rare and few cases have been reported, none of them involving the spleen. We report a case of a 59-year old female patient with cystic spleen lesions that after splenectomy were diagnosed as Mantle cell lymphoma.

Resumen

El linfoma de células del manto hace parte del subgrupo de linfomas no Hodgkin (LNH). Este se manifiesta con adenopatías, esplenomegalia, síntomas B y compromiso cutáneo asociado a enfermedad diseminada. El compromiso esplénico se presenta como: Esplenomegalia sin lesión focal; lesiones sólidas únicas o múltiples e infiltración del bazo sin cambios morfológicos ni lesiones focales. La aparición de linfoma con lesiones quísticas es extremadamente rara, se encuentran solo unos cuantos casos en la literatura, ninguno de ellos en el bazo. Se expone el caso de una paciente de 59 años de edad, quien consultó por dolor abdominal intermitente. En los estudios diagnósticos se observó esplenomegalia con lesiones sólidas y quísticas. Se realizó esplenectomía con estudio histopatológico que confirmó compromiso por linfoma de células del manto.

Introduction

Mantle cell lymphoma (MCL) is a B-cell neoplasm originating in the mantle area of the lymphoid follicles (1). It is part of non-Hodgkin's lymphomas, and is characterized by being aggressive (2). In the spleen, this pathology manifests mainly with splenomegaly (3). The cystic form of the lymphoma is extremely rare, in the literature we present case presentations, but none, to our knowledge, with splenic involvement.

Case presentation

It is a female patient of 59 years of age, with

intermittent abdominal pain of long duration. In extra-institutional studies, intra-abdominal cystic lesions were found with no conclusive cytological findings. In order to expand the studies, a tomography Computed tomography (CT) (Figure 1) of the abdomen revealed splenomegaly with solid, low density and confluent nodular lesions, and multiple cystic lesions, the largest exophytic without a clear origin (pancreas vs. spleen). Magnetic resonance imaging (MRI) with contrast medium (Figure 2) confirmed its splenic origin. A splenectomy was performed (Figure 3) to clarify the etiology of the lesions and the pathological study demonstrated mantle cell lymphoma (Figure 4).

¹Radiologist, Hospital Pablo Tobon Uribe, Medellín, Colombia.

²Radiology resident, Universidad CES, Medellín, Colombia.

³Pathologist, Hospital Pablo Tobon Uribe, Medellín, Colombia.

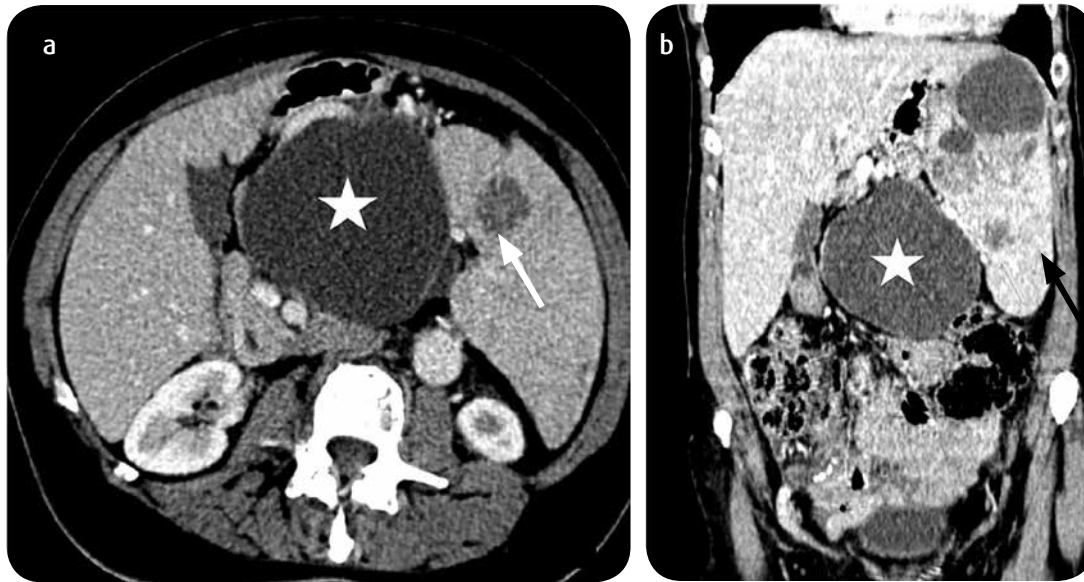


Figure 1. CT scan of the abdomen in the axial plane: a) A large cystic lesion (asterisk) is observed in the midline with enhancement of its walls, displacing the head of the pancreas, mesenteric vessels and intestinal loops. Also, a low-density solid nodular lesion is observed in the spleen (arrows). b) Coronal reconaissance: Splenomegaly, cystic lesions and solid spleen and large intra-abdominal, central cystic lesion. It is not clear whether it originates in the spleen or the pancreas.

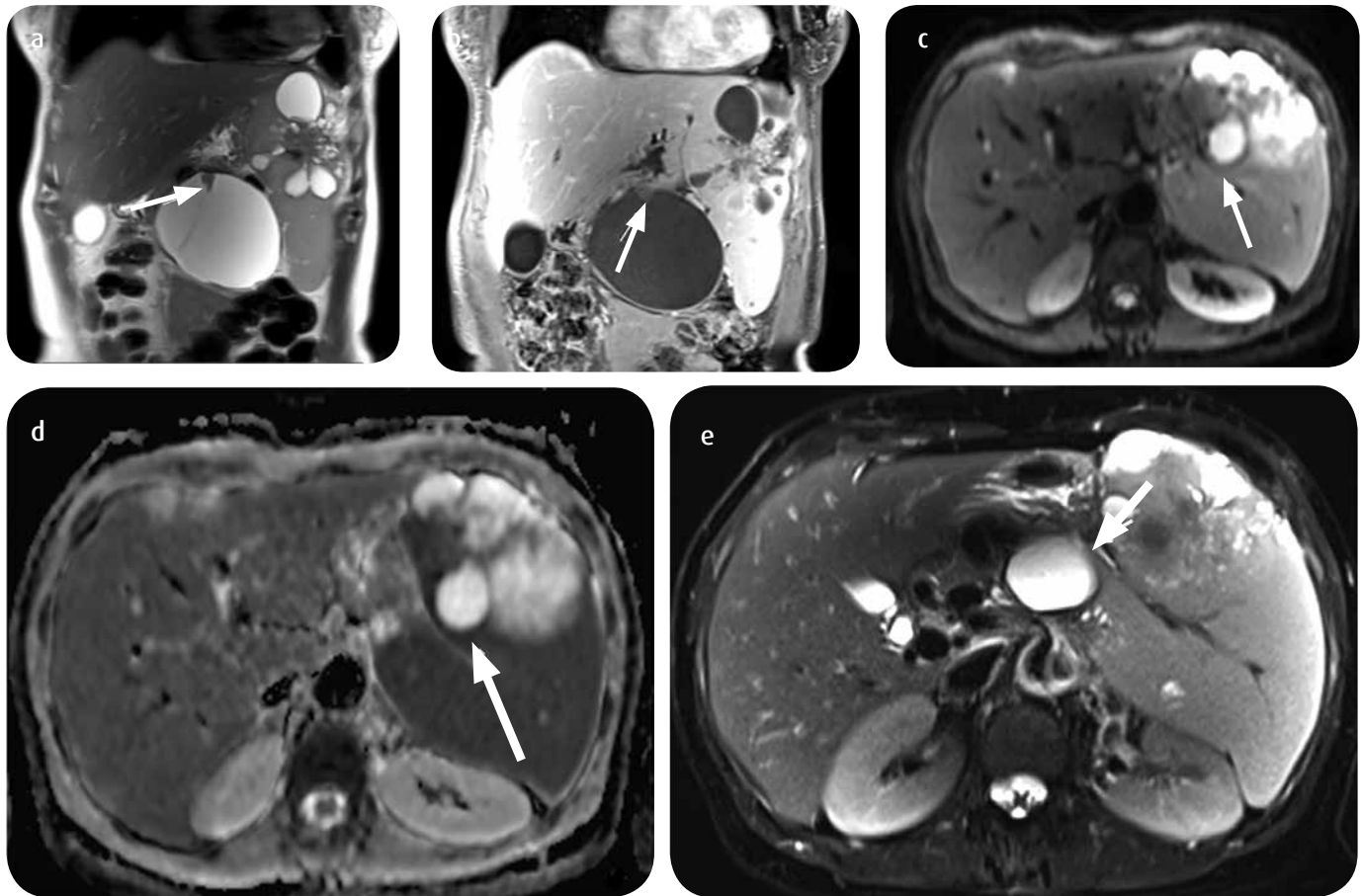


Figure 2. Sequence with coronal T2 information: a) It shows splenic cystic lesions of high signal with some thin septa and small peripheral solid nodular component in the dominant cystic (arrow), exophytic lesion. Solid lesions show slight low signal. b) Coronal with T1 information after contrast medium: A slight enhancement of the exophytic central cyst wall is noted. c) Sequences B1000 and d) ADC map: They show splenic lesions of high signal (T2 shinethrough) (arrows), there is no restriction to the diffusion. e) Axial image with T2 information: Shows the sign of the "peak" (arrow) which confirms the splenic origin of the dominant cystic lesion.



Figure 3. (a) Surgical specimen: Enlarged spleen, lobulated contours by solid and cystic lesions. b) Macroscopic part: Large exophytic cyst originating in the spleen. Courtesy of Dr. Víctor Quintero.

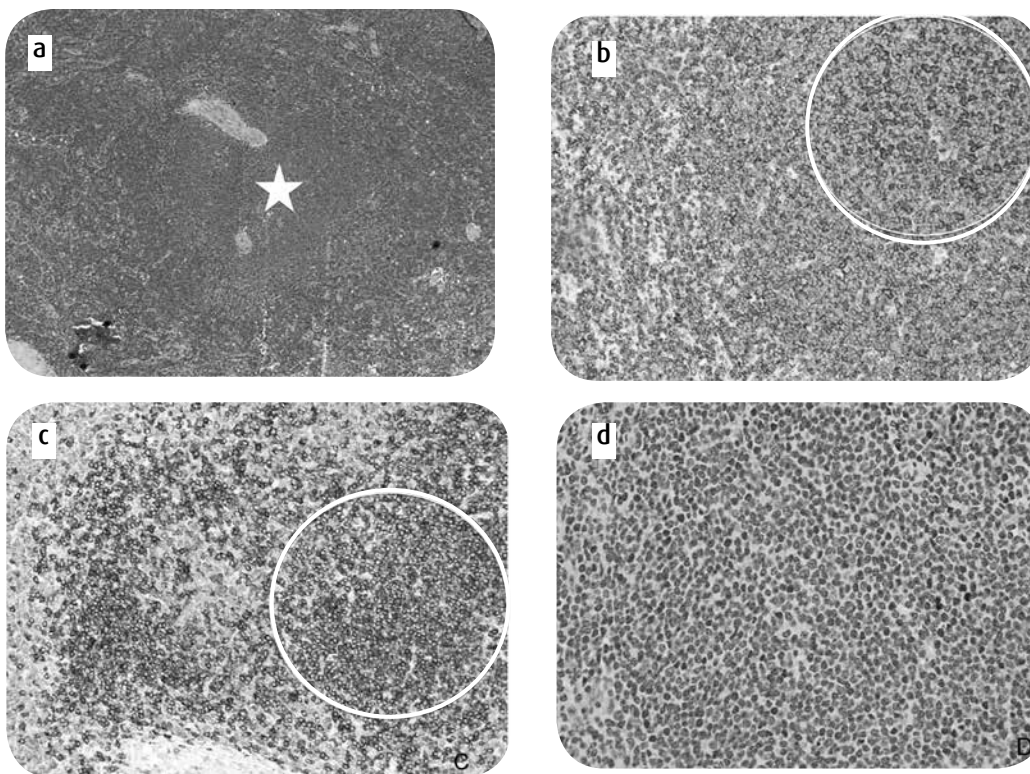


Figure 4. a) Hematoxylin and eosin staining, a central (star) nodule made up of a uniform population of intermediate size lymphocytes with thick chromatin. b) Immunohistochemical staining for CD20 intensely positive by B lymphocytes (cells surrounded by white circle). c) Positive CD5 staining (cells surrounded by a circle) confirming that they are B lymphocytes. d) Positive cyclin D1 staining in mantle cell lymphomas (brown cells).

Overview of Mantle Cell Lymphoma

MCL is a B cell neoplasm originating in the mantle area of the lymphoid follicles. Three patterns are recognized: mantle area, nodular and diffuse. These, before immunohistochemical techniques, show CD20 +, CD5 +, CD43 +, and cyclin D1 + as nuclear marker. MCL has also been related to the translocation of chromosomes 11-14 (4, 7).

It appears at a mean age of 65 years, with predominance in the male population (8) and is usually diagnosed in advanced stages, where extranodal compromise is found. The clinical

manifestations are not very different from those of other types of NHL. It usually appears with lymphadenopathy and splenomegaly (50%), B symptoms (30%), central nervous system involvement (10%) and cutaneous involvement associated with disseminated disease (9).

The course of the disease is variable. It can be painless or asymptomatic and chronic as the presented clinical case or, on the contrary, very aggressive, in the literature an average survival is mentioned between 3 to 5 years (10).

Discussion

The lymphoma comprises a group of tumors derived from cells of the immune system, heterogeneously histologically. It is the fifth most common tumor in the United States and the fifth cause of mortality secondary to cancer (4). It is divided into 2 large groups: Hodgkin's lymphoma (HL) and non-Hodgkin's lymphoma (NHL) (5), the latter being subdivided into B-cell lymphoma and T cells (4) (Table 1).

Lymphoma is the tumor that most commonly affects the spleen (2), either primary or secondary (2). It may be by HL or NHL,

although the second (1) is more common, which has been found in up to 70% of postmortem studies (2). The histologic subtype that most commonly affects the spleen is large cell lymphoma. On the other hand, primary splenic lymphoma is rare, representing only 1-2% and has the histological characteristics of NHL in most cases.

Mantle cell lymphoma is an aggressive B-cell NHL (6). It has an evolution with relatively frequent relapses, short remissions and, unlike other subtypes of NHL, therapy has not been shown to be curative (7).

Table 1. Subtypes of lymphomas

Tipo de linfoma		
Hodgkin	Nodular	
	Classic	
	Nodular sclerosus	
	Rich in lymphocytes	
	Mixed Cellularity	
	Lymphocyte Depletion	
No Hodgkin	B cells	Lymphoblastic leukemia
		Chronic lymphocytic leukemia
		Plasmocytoma
		MALT
		Mantle
		Follicular
		B-cell diffuse
	Burkitt	
	T cells	Lymphoblastic lymphoma
		Mycosis fungoides
		Peripheral T-cell lymphoma
		Angioimmunoblastic
		Anaplastic

Spleen and lymphoma in images

According to studies showing high correlation of macroscopic images and pathology, possible patterns of lymphoma in the spleen are described (5, 2):

Normal spleen, only with microscopic infiltration.

- » Splenomegaly without focal lesion. This is the most common pattern. The MCLs that compromise the spleen are manifested mainly by splenomegaly, a characteristic finding of this lymphoma (11).
- » Single focal mass.
- » Multiple nodular lesions that may have miliar presentation or multiple masses larger than one centimeter.

In TC, the masses are usually low signal without significant enhancement with the contrast medium (2). In some cases its density may be low, almost like water, because of its high cellularity (3) or necrosis of liquefaction (2). Other reported features are dystrophic calcifications secondary to necrosis, but this is exceptionally uncommon in untreated lymphomas (2).

The sequences with T1 and T2 MRI information for the evaluation of spleen lymphomas have low sensitivity because lymphomatous infiltration has a behavior very similar to that of healthy splenic parenchyma (2). The solid lesions are of low signal in images with information T1 and of high signal with information T2 (2). Alterations in the sequences with T1 information following the administration of contrast media allow a more accurate determination of the extent of compromise in the spleen (2).

Differential diagnoses of low signal lesions in the spleen correspond

to macrocystic lymphatic malformation, the most common of all. Also, we must think of infectious diseases, hemangioma and, as a last option, the angioma of the littoral cells (2).

Cystic lymphomas reported in the literature are few, one of which corresponds to large, diffuse, hepatic B-cell lymphoma, manifested as a single cystic lesion (12). There are two cases in the literature studied as pseudocysts: one in the adrenal gland and another in the paratesticular region, with a diagnosis of diffuse large B-cell lymphomas, an extremely rare case (13). Another case of cystic lesion of the larynx that corresponds to mantle cell lymphoma is found (6). Finally, diffuse large B-cell lymphoma was described in the cystic jaw (14).

The case is interesting because it is a primary lymphoma of the spleen that, in addition to splenomegaly, shows solid lesions of low signal and cystic lesions, one of them exophytic of great size with mass effect on the neighboring structures.

The role of the images is limited in these cases since the main finding is the splenomegaly, which is caused by multiple pathologies. In addition, it does not present any specific signs for its diagnosis; however, this diagnosis should be taken into account between splenomegaly differentials.

Conclusion

MCL is an aggressive B-cell NHL, which at present does not have therapies with curative potential. It mainly affects extranodal organs, isolated splenomegaly is its most frequent form of appearance. Our case is interesting because it is a lymphoma with atypical presentation due to its cystic and solid presentation and, to our knowledge, is the first case found in the primary lymphoma literature of the spleen with cystic lesions.

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Correspondence

Ricardo Uribe González
Residente de Radiología Universidad CES
Diagonal 29D # 9A Sur-150
Medellín, Colombia
ricardouribegonzalez@gmail.com

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