

**Enfant de 3ans sans antécédent présentant une
anémie hémolytique avec thrombopénie de
consommation, CIVD ...**

... et splénomégalie

**comment orienter les examens complémentaires
d'imagerie ...**



- Rémi DUPRES IHN

CI 28Hz
RV

2D
64%
C 58
P Bas
HGén

1^{er} temps échographique

Volumineuse splénomégalie hétérogène,
avec multiples nodules hypoéchogènes
en son sein

Absence de signe d'HTP



CI 28Hz
RV

2D
62%
C 58
P Bas
HGén

C2 C4
+18.5
-18.5
cm/s

C3

CI 12Hz
RP

2D
47%
C 55
P Moy
Gén

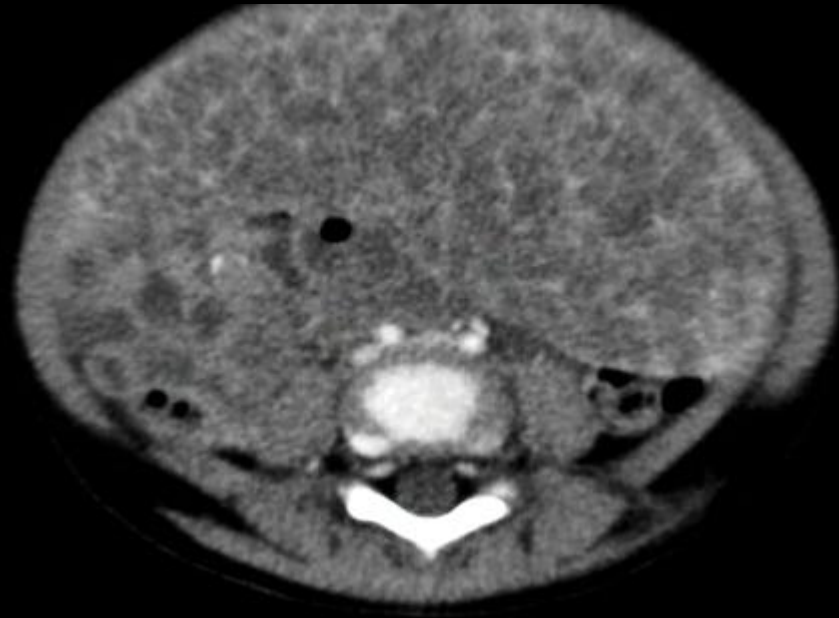
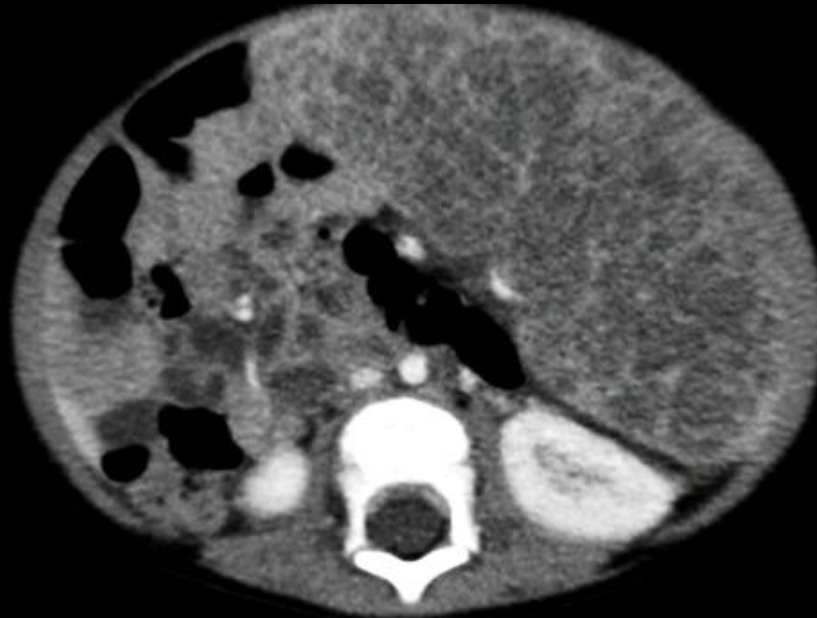
Coul
50%
1320Hz
FP 85Hz
Moy

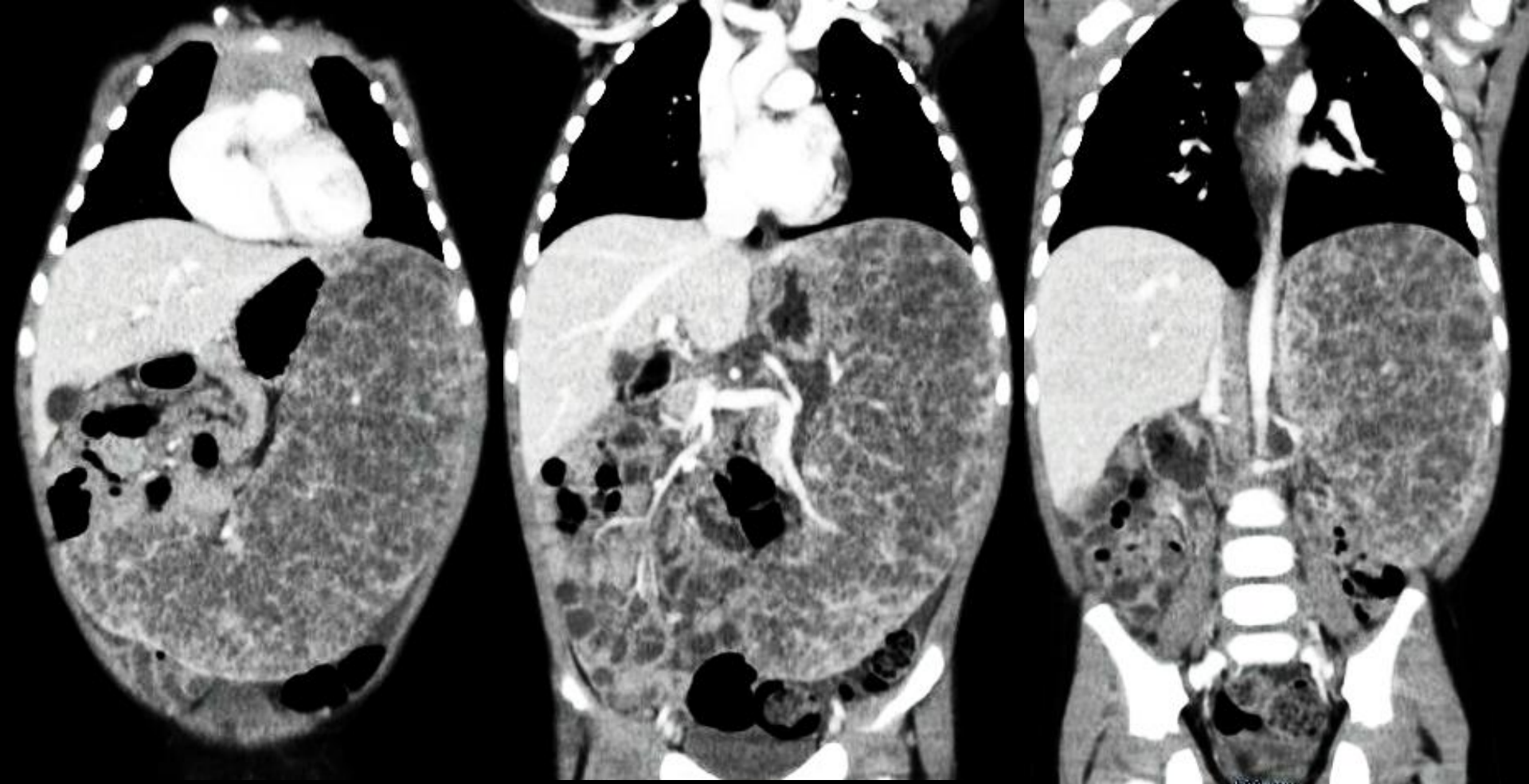


2nd temps scanographique



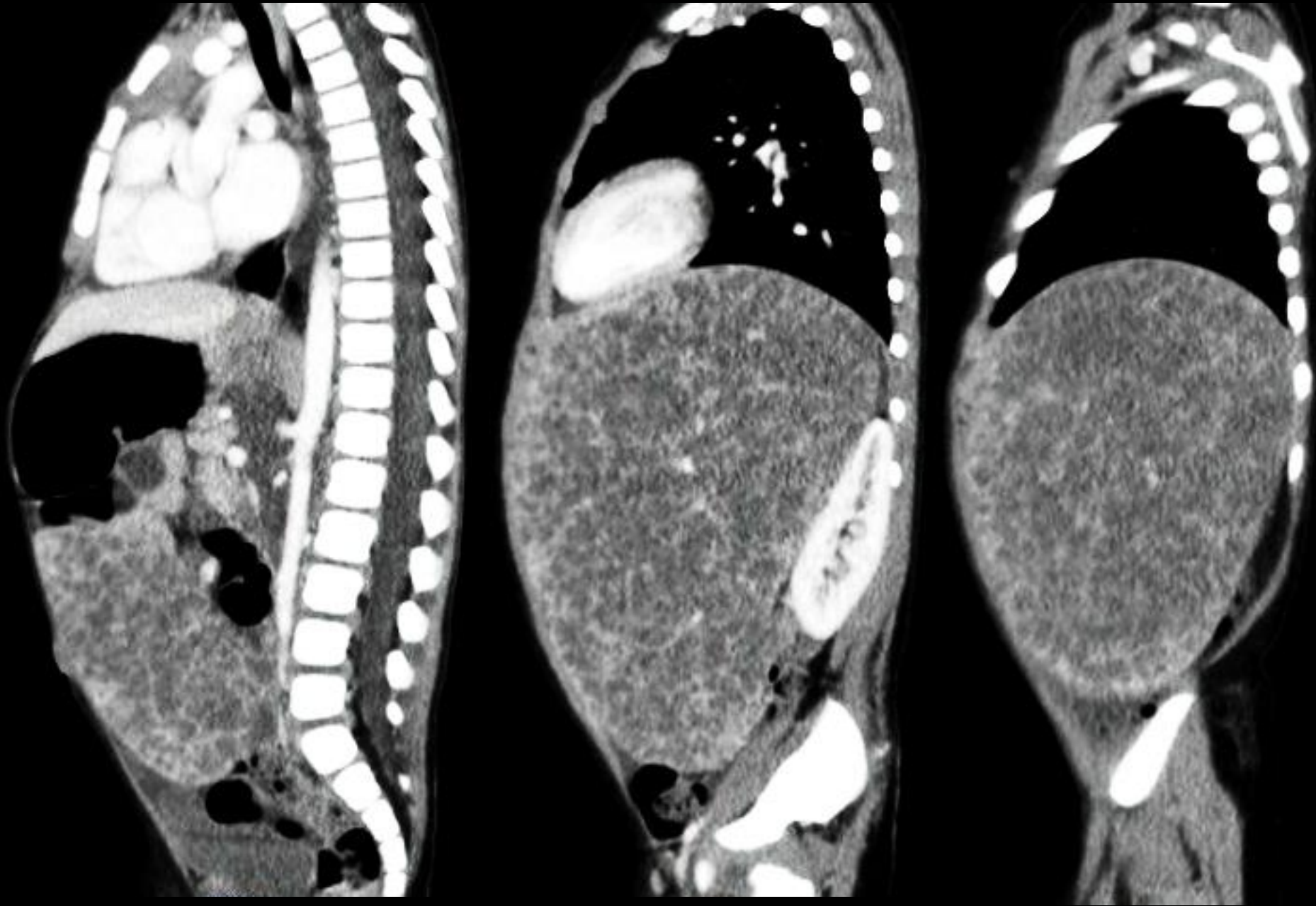
**Parenchyme splénique rendu hétérogène dans son ensemble par une multitude de nodules hypodenses . Refoulement des organes adjacents sans envahissement des structures
Absence d'adénomégalie**





Reformations frontales

Flèche splénique = 16cm . Le pôle inférieur de la rate va jusqu'à la paroi de la fosse iliaque droite

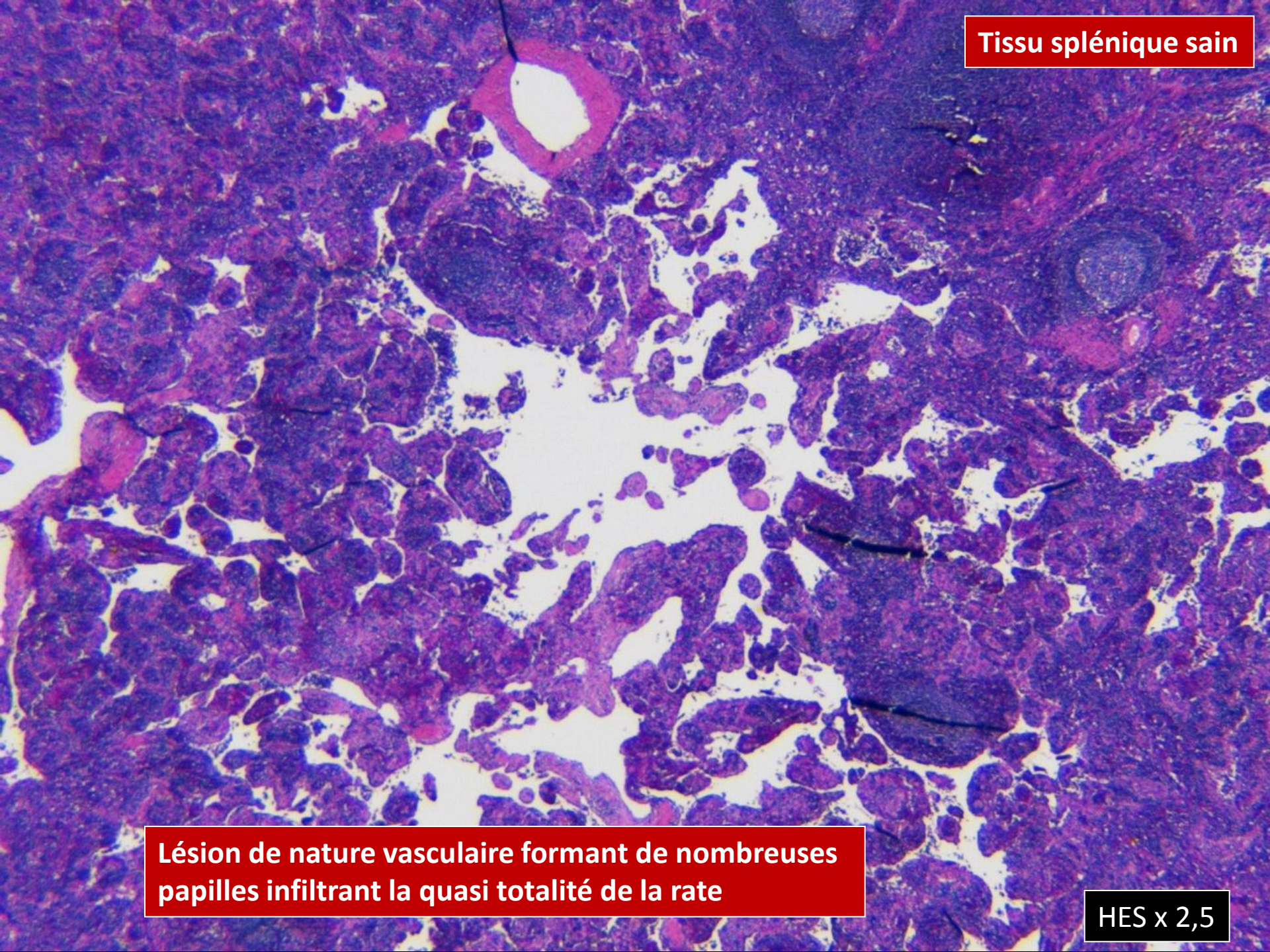


Reformations sagittales

L'aspect peu typique en imagerie, comme c'est souvent le cas des pathologies spléniques, ne permet pas d'apporter un diagnostic de certitude.; il permet toutefois de limiter les hypothèses à une atteinte tumorale ou infectieuse ou à une maladie de surcharge

La splénectomie est nécessaire dans le contexte clinique du patient et l'analyse anatomopathologique s'avère indispensable.

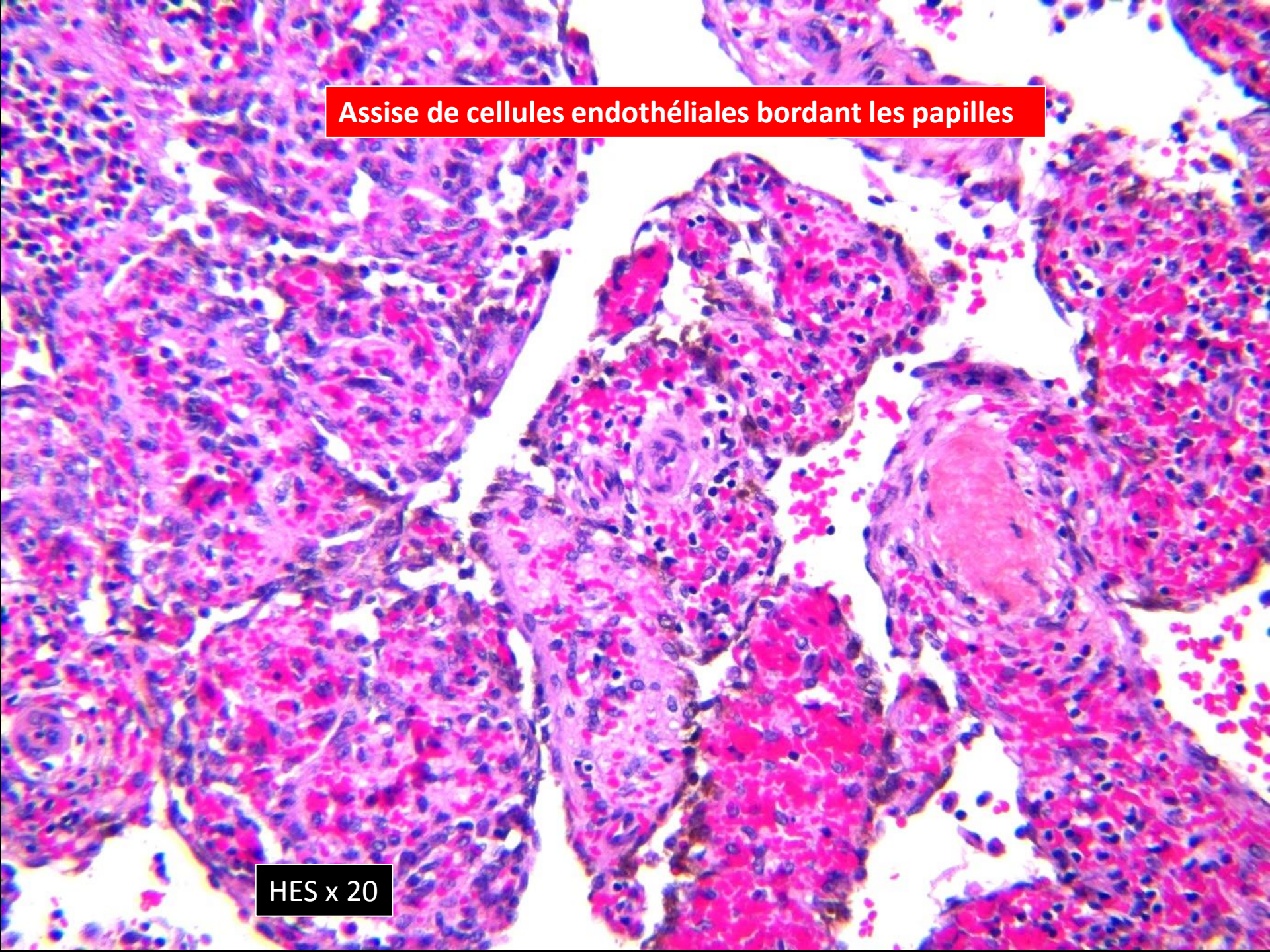


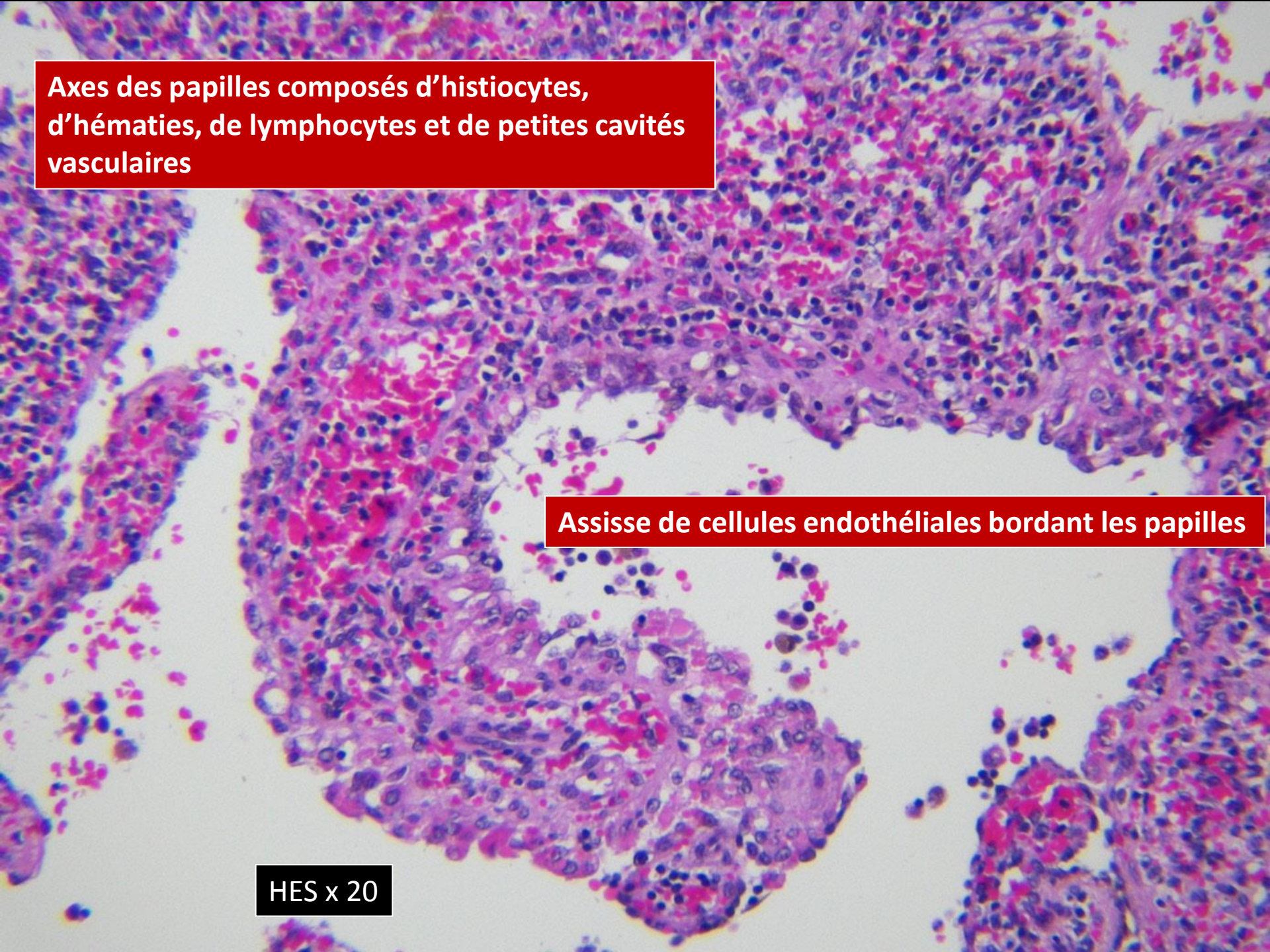


Lésion de nature vasculaire formant de nombreuses papilles infiltrant la quasi totalité de la rate

Assise de cellules endothéliales bordant les papilles

HES x 20



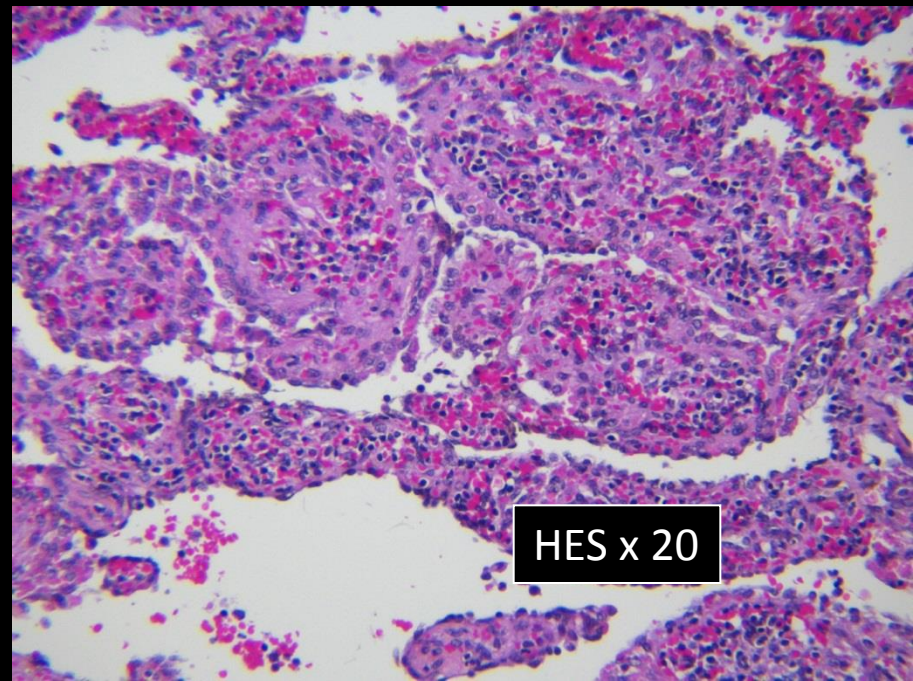
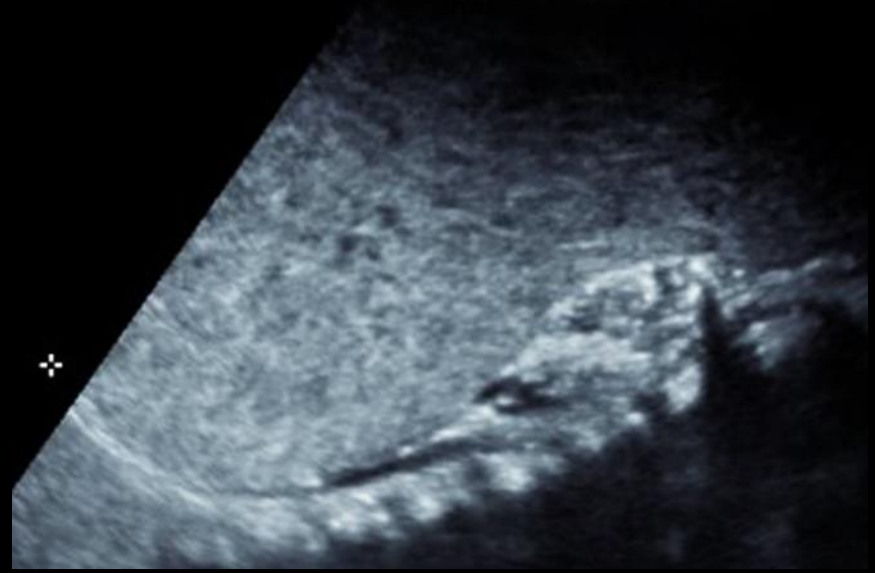


Axes des papilles composés d'histiocytes,
d'hématies, de lymphocytes et de petites cavités
vasculaires

Assise de cellules endothéliales bordant les papilles

HES x 20

diagnostic anatomo-pathologique : **angiome splénique à cellules littorales**

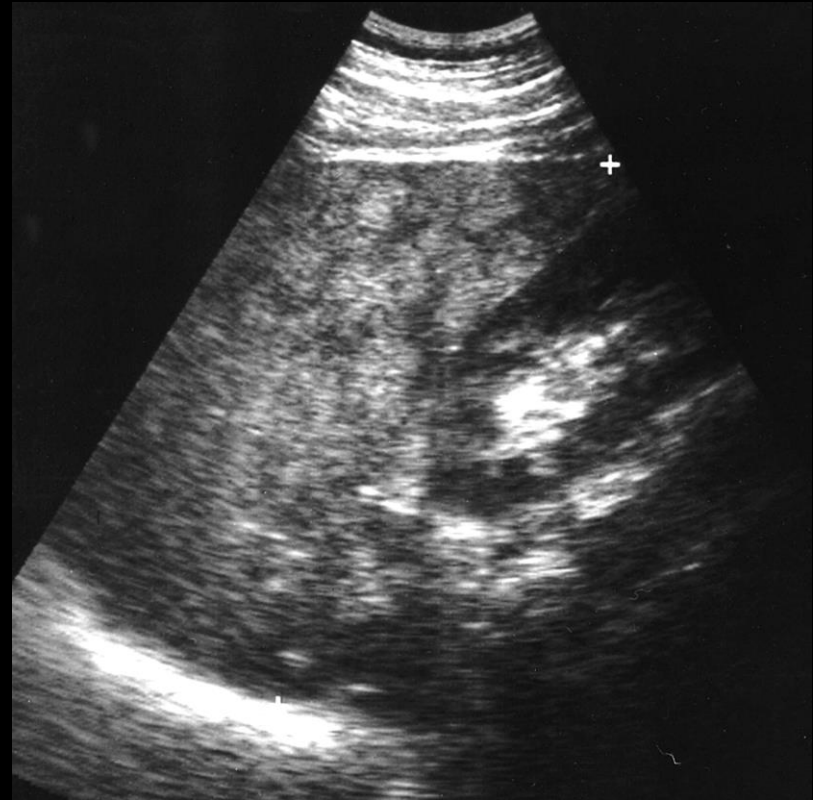


Epidémiologie

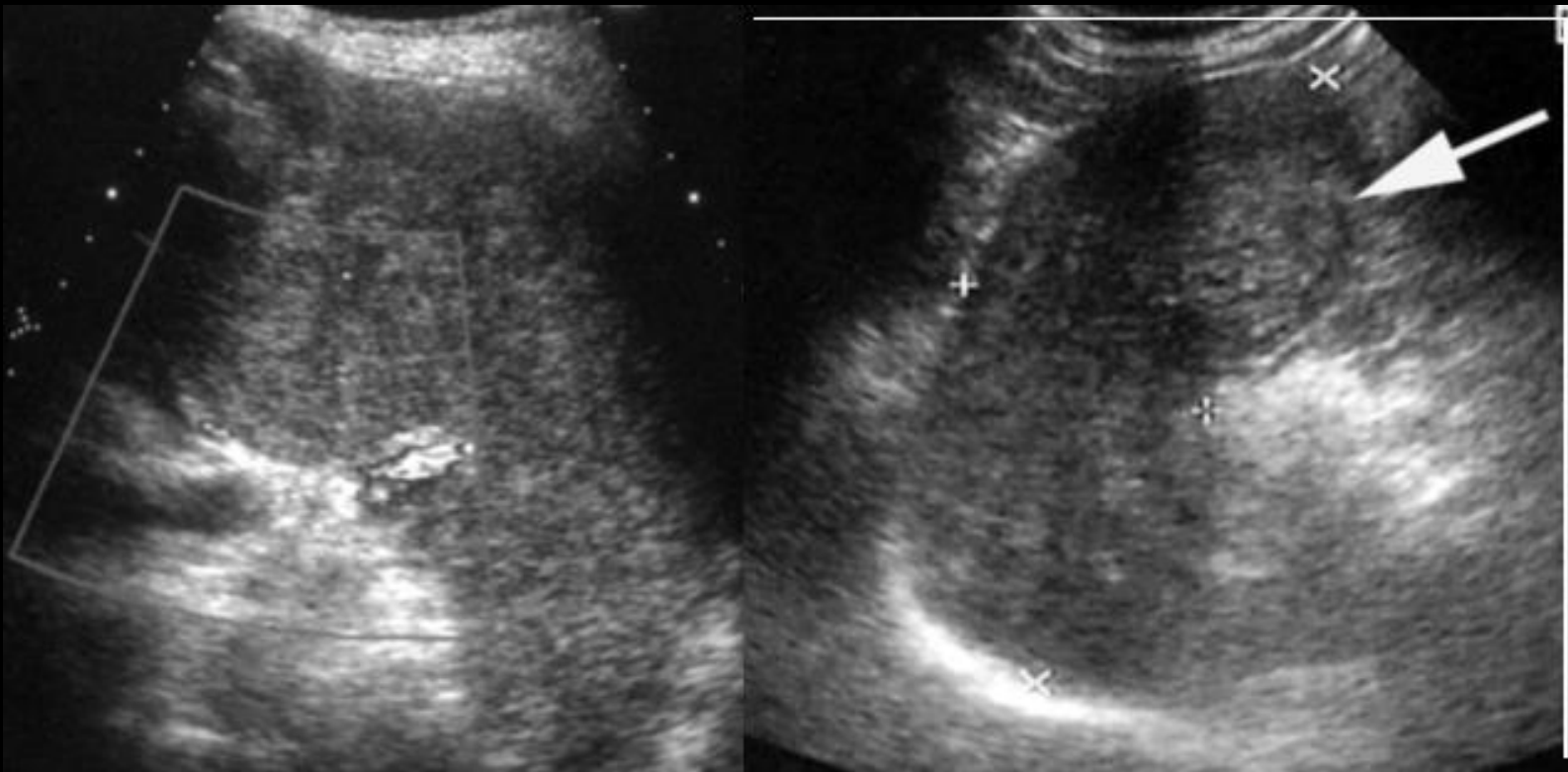
- **Tumeur vasculaire** rare, bénigne (mais rares cas de formes malignes rapportés dans la littérature (Angiosarcomes à cellules littorales))
- 1^{ère} description par Falk et al. En 1991
- Se développe à partir des cellules littorales des sinus de la pulpe rouge de la rate
- SR = 1 ; Pas d'âge de prédilection
- En général **asymptomatique**; possibles manifestations **d'hypersplénisme**

Echographie

- Aspect marbré de la rate avec nodules pouvant être iso-échogènes, hypo-échogènes ou hyper-échogènes
- Vascularisation centrale et périphérique au doppler (diagnostic différentiel avec hémangiome caverneux)



*Littoral Cell Angioma of the Spleen:
Imaging Features. Lisa L. Kinoshita
et al. AJR 2000;174:467–469*



Sonographic features of littoral cell angioma

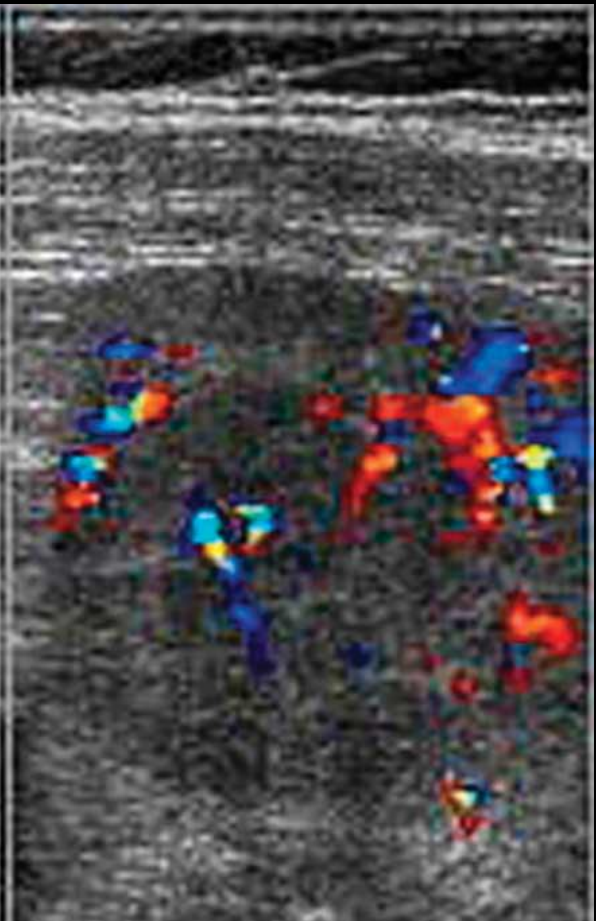
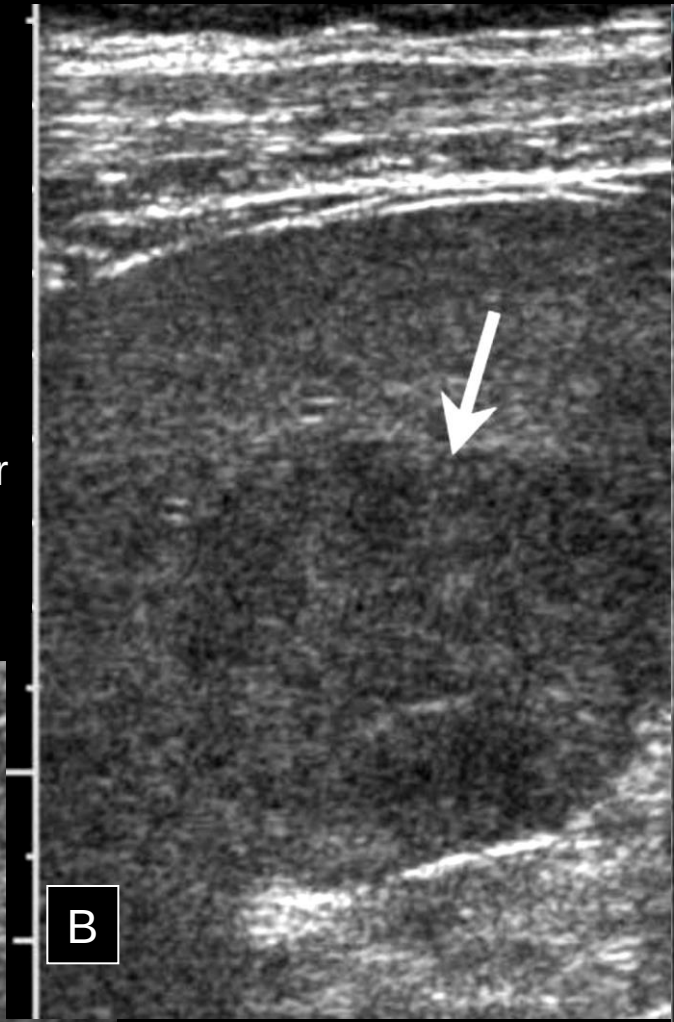
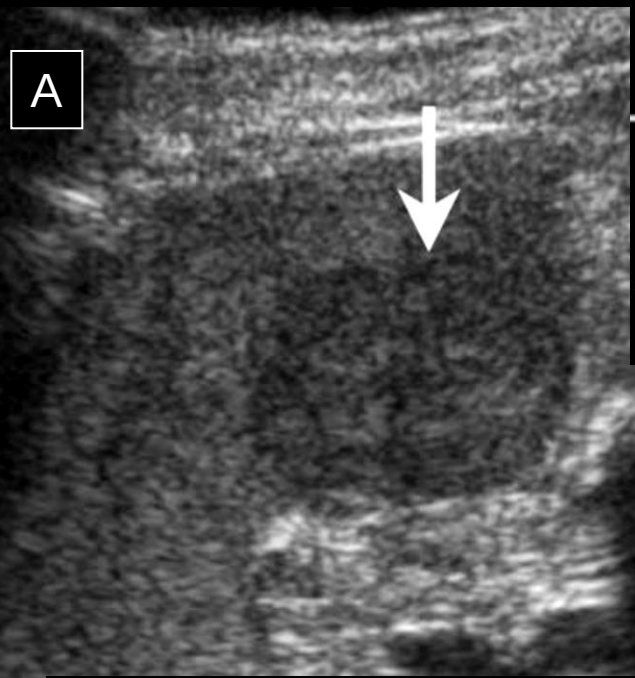
(a) Longitudinal sonogram of the spleen in a 50-year-old woman with asymptomatic splenomegaly shows a **heterogeneous splenic echotexture** with multiple hyperechoic masses.

(b) Transverse sonogram of the spleen in a 75-year-old man with asymptomatic splenomegaly shows a focal hyperechoic mass (arrow).

***Primary Vascular Neoplasms of the Spleen: Radiologic-Pathologic Correlation.
Robert M. Abbott et al. RadioGraphics 2004; 24:1137–1163***

A, Longitudinal gray scale sonogram of the spleen obtained with a 4.5-MHz transducer reveals a well-circumscribed, hypoechoic intrasplenic mass (arrow).

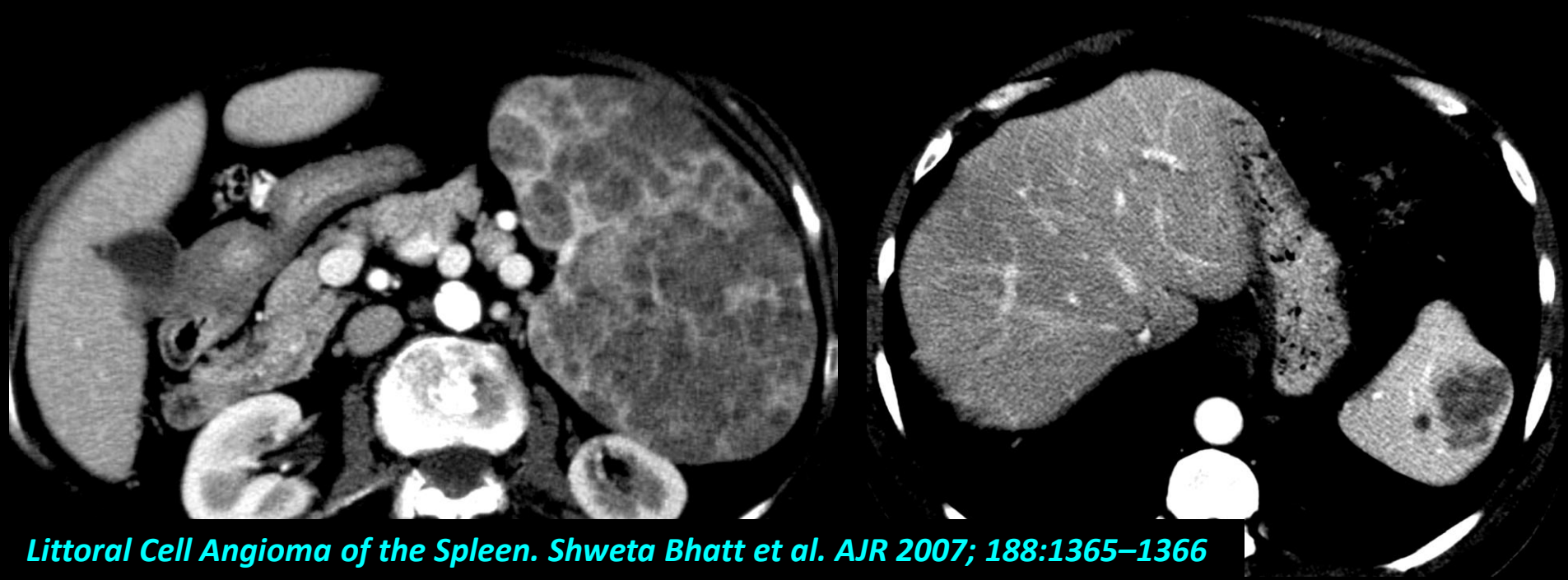
B, Sonogram of the same lesion obtained with a 6-MHz transducer shows better delineation of the lesion (arrow).



Color flow Doppler evaluation of the splenic lesion shows peripheral and central vascularity.

Tomodensitométrie

- Splénomégalie
- Multiples nodules de taille variable, hypodenses spontanément et au temps portal avec homogénéisation à la phase tardive
- < 100cas décrits dans la littérature anglosaxonne; <10 cas présentant une forme nodulaire unique
- Absence de calcification



CT Features of Littoral Cell Angioma with Gross Pathologic Correlation

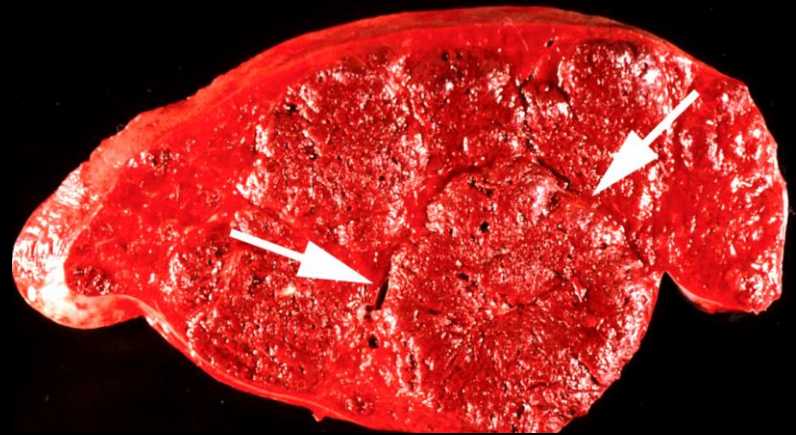
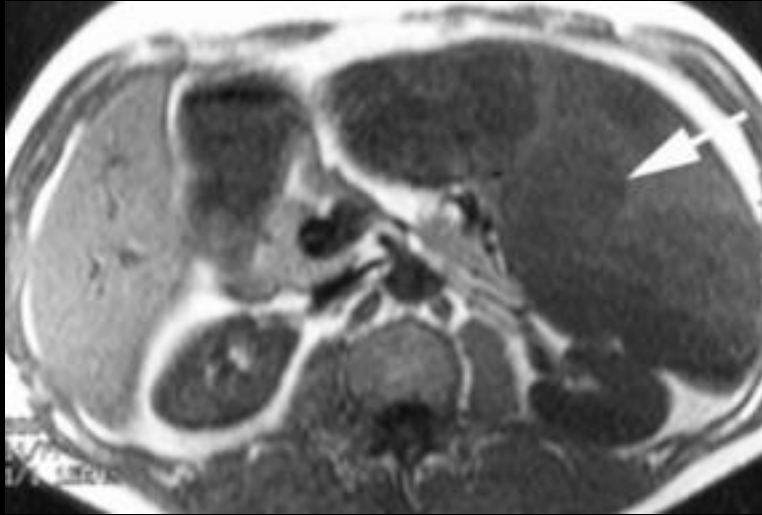
Patient No./ Age (y)/ Sex	Spleen Length (cm)	Spleen Volumetric Index (cm ²)	No. of Masses	Mass Diameter (cm)	Mass Margins	Contrast Enhancement Phase	CT Attenuation and Enhancement	Gross Pathologic Features*
1/55/F	22	97	Too numerous to count	0.5–2.0	Well defined	Early and late portal venous	Homogeneously hypoattenuating and isoattenuating in late portal venous phase	Brown nodules
2/61/M	19	63	Too numerous to count	0.2–4.0	Well defined	Late portal venous	Homogeneously hypoattenuating	Brown nodules
3/32/M	21	81	Too numerous to count	0.5–2.0	Well defined	Late portal venous	Heterogeneously hypoattenuating	Black nodules
4/41/M	23	202	Too numerous to count	1.0–6.0	Well defined	Early portal venous	Heterogeneously hypoattenuating	Dark red nodules with cystic spaces ...
5/36/F	16	36	Too numerous to count	0.2–1.5	Ill defined	Unenhanced and late portal venous	Not visible on unenhance image, homogeneously hypoattenuating in late portal venous phase	
6/84/F	23	77	Too numerous to count	0.5–2.0	Well defined	Early portal venous	Homogeneously hypoattenuating	Brown nodules with cystic spaces
7/74/F	12	24	4	1.5–2.0	Ill defined, some subtle	Unenhanced and late portal venous	Not visible on unenhance image, homogeneously hypoattenuating in late portal venous phase	Dark red nodules with cystic spaces
8/39/F	21	65	Too numerous to count	0.2–2.5	Well defined	Early portal venous	Homogeneously hypoattenuating	Black nodules

* Photographs of the gross pathologic spleen specimen resected from patient 5 were not available.

Littoral Cell Angioma of the Spleen: CT Features with Clinicopathologic Comparison. Angela D. Levy et al. Radiology 2004; 230:485–490

IRM

- **Nodules spléniques hypo-intenses en pondérations T1 et T2**, reflétant la **présence d'hemosidérine** (mais de façon non systématique)



Littoral cell angioma in a 41-year-old man who was found to have thrombocytopenia before blood donation.

- (a) T1-weighted MR image shows splenomegaly and multiple subtle hypointense splenic masses (arrow).
- (b) On a T2-weighted MR image, the masses remain hypointense (arrow), adjacent to the normally bright splenic parenchyma.
- (c) Photograph of the cut surface of the resected spleen shows multiple well-demarcated masses (arrows).

Primary Vascular Neoplasms of the Spleen: Radiologic-Pathologic Correlation. Robert M. Abbott et al. RadioGraphics 2004; 24:1137–1163

Quelques cas de la littérature pour se faire l'œil et garder ces images dans un coin de la tête ...



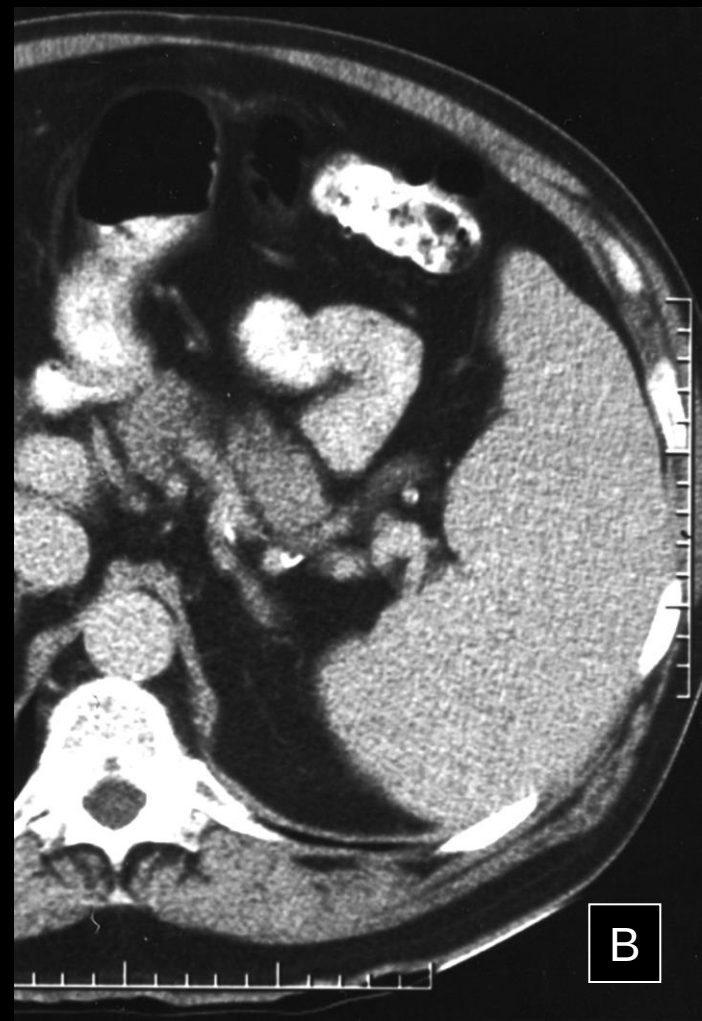
Cas 1



Cas 2

Dascalescu CM, Wendum D, Gorin NC: Littoral cell angioma as a cause of splenomegaly. New Engl J Med 2001, 345(10):772-773

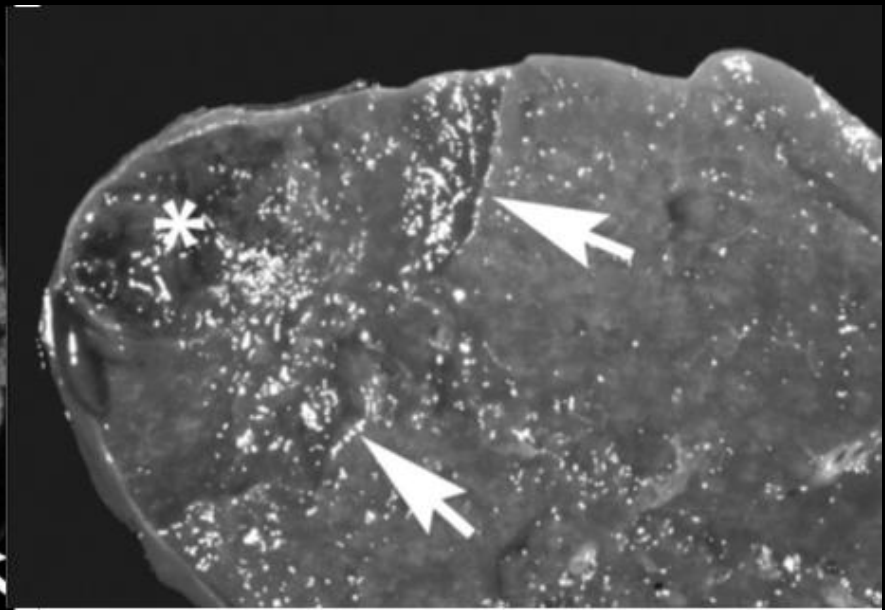
Cas 3



A, Early contrast-enhanced helical CT scan shows enlarged spleen containing innumerable small, focal, low-density lesions.

B, Delayed contrast-enhanced helical CT scan shows complete filling of splenic lesions seen in A. Lesions are now isodense with normal splenic parenchyma.

Littoral Cell Angioma of the Spleen: Imaging Features. Lisa L. Kinoshita et al. AJR 2000;174:467–469



Cas 4

Littoral cell angioma in 74-year-old woman who presented with persistent rectal bleeding 2 weeks after undergoing colonoscopy. She also had cirrhosis secondary to nonalcoholic steatohepatitis.

- (a) Transverse contrast-enhanced late portal venous phase CT image shows several low-attenuating masses (arrows) in a normal-sized spleen.
- (b) Resected surgical specimen shows a hemorrhagic nodule (arrows) at the periphery of the spleen. The nodule contains small hemorrhagic cystic spaces (*).

Littoral Cell Angioma of the Spleen: CT Features with Clinicopathologic Comparison. Angela D. Levy et al. Radiology 2004; 230:485–490

Cas 5



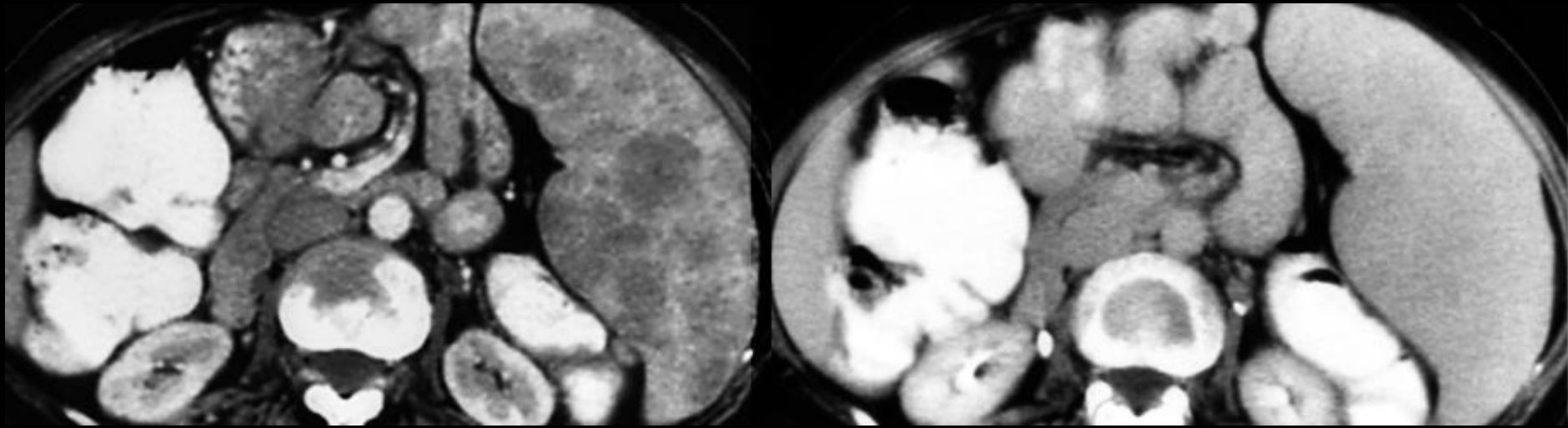
Littoral cell angioma in 41-year-old man found to have thrombocytopenia while being evaluated for potential blood donation.

Transverse contrast-enhanced early portal venous phase CT image shows multiple, large, partially confluent masses within an enlarged spleen.

The masses have partial areas of heterogeneous high attenuation.

Littoral Cell Angioma of the Spleen: CT Features with Clinicopathologic Comparison. Angela D. Levy et al. Radiology 2004; 230:485–490

Cas 6

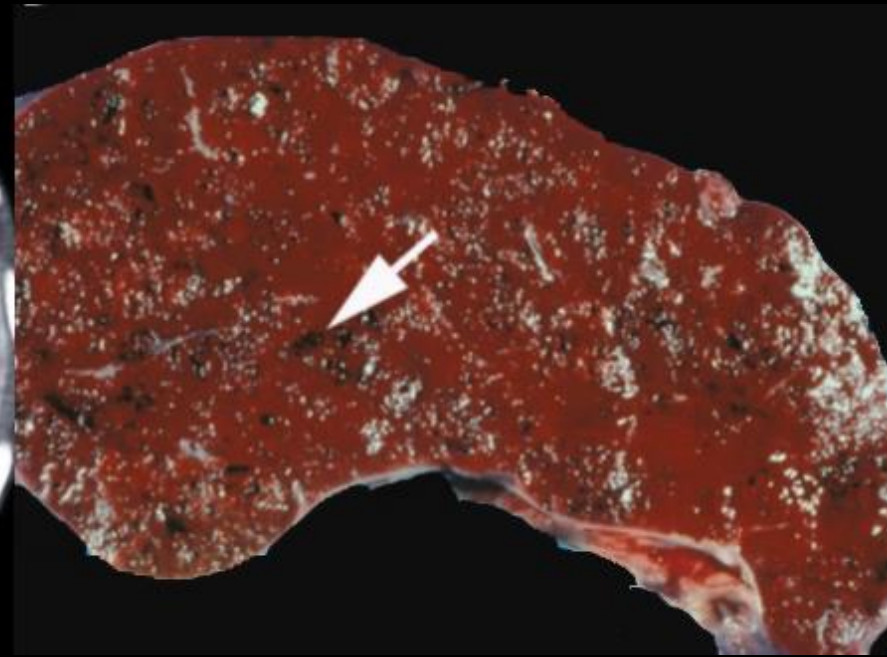


Littoral cell angioma in 55-year-old woman found to have splenomegaly at physical examination for evaluation of leg swelling.

- (a) Transverse contrast-enhanced early portal venous phase CT image shows multiple, partially confluent hypoattenuating masses in the spleen.
- (b) Transverse contrast-enhanced late portal venous phase CT image shows that the masses are homogeneously enhanced and have become imperceptible.

Littoral Cell Angioma of the Spleen: CT Features with Clinicopathologic Comparison. Angela D. Levy et al. Radiology 2004; 230:485–490

Cas 7



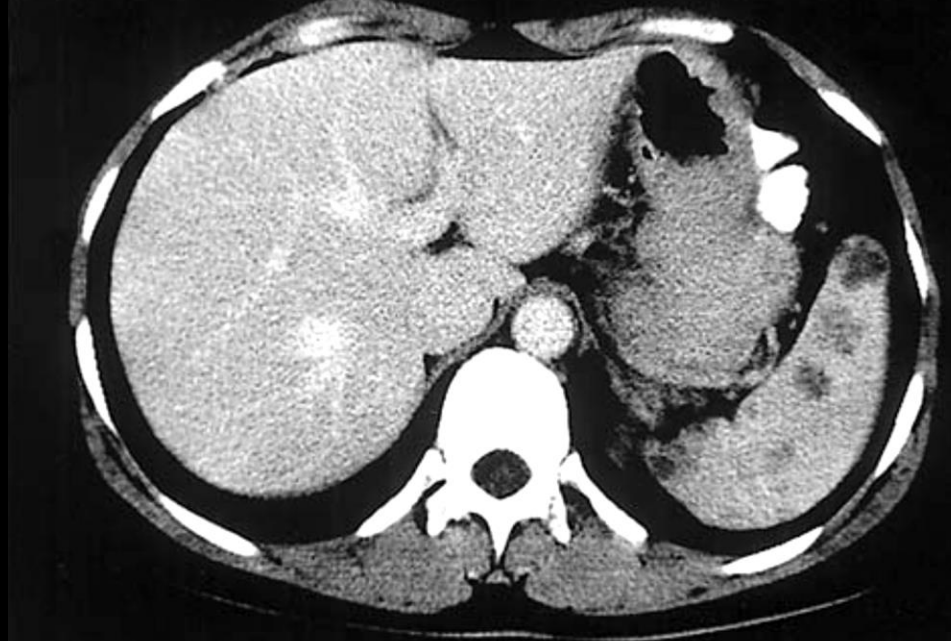
Littoral cell angioma in a 39-year-old woman who was found to have splenomegaly when she was being evaluated for peptic ulcer disease.

- (a) Contrast-enhanced CT scan shows the enlarged spleen, which contains innumerable hypoattenuating masses.
- (b) Photograph of the cut surface of the resected spleen shows multiple blood-filled spaces (arrow).

Primary Vascular Neoplasms of the Spleen: Radiologic-Pathologic Correlation. Robert M. Abbott et al. RadioGraphics 2004; 24:1137–1163



Axial pre-contrast CT scan showed multiple hypodense nodules in the spleen



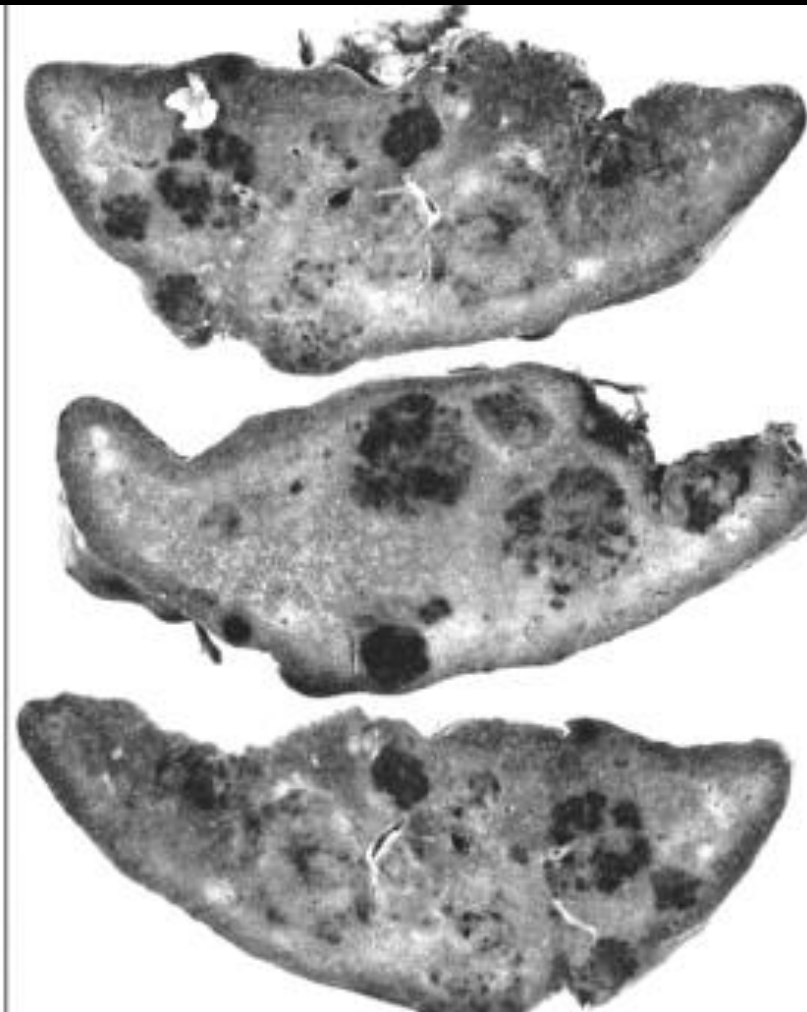
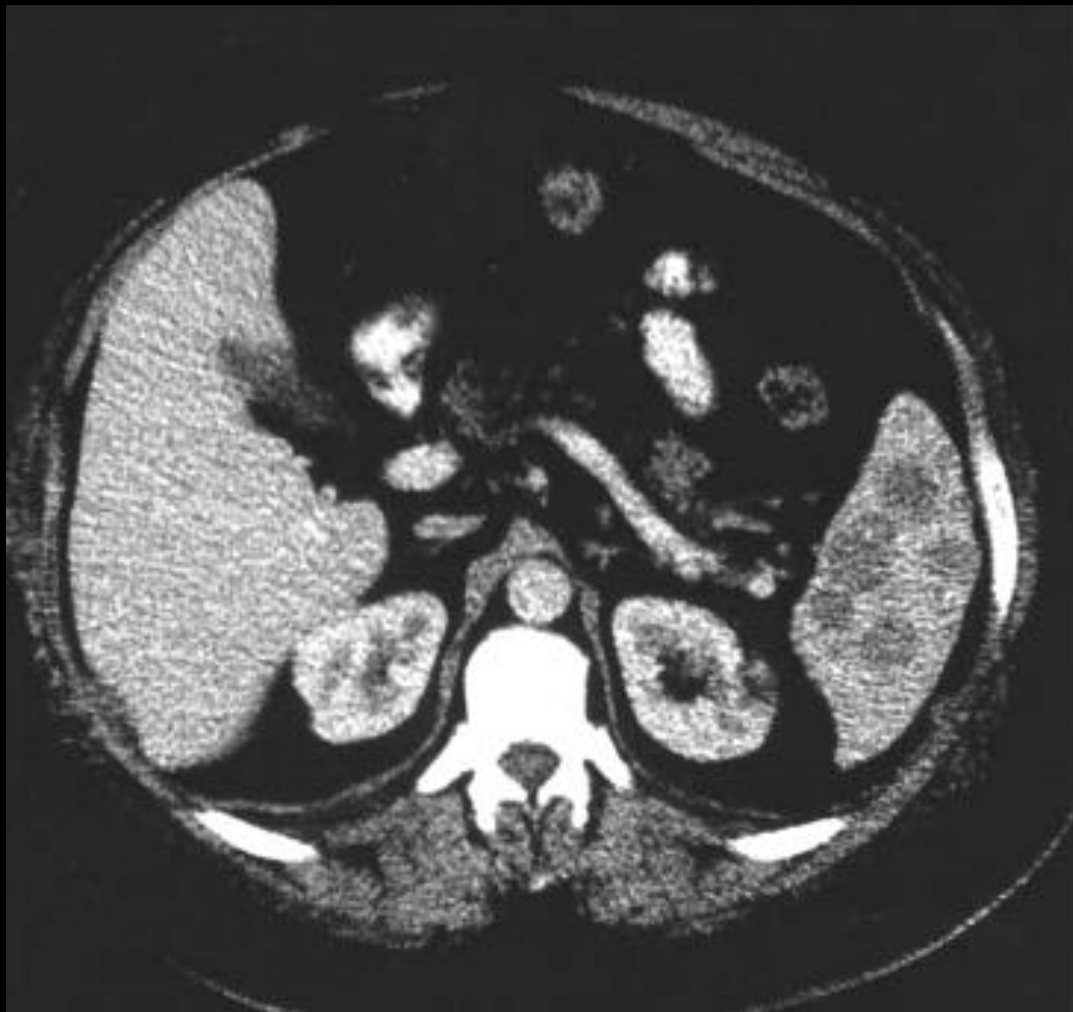
Axial and coronal contrast-enhanced CT images at late portal venous phase showed multiple welldefined, heterogeneously hypoattenuating lesions with partial areas of high density corresponding with the vascular spaces inside the lesions.



Cas 8

Littoral Cell Angioma of the Spleen: a case report. YU-PENG LIU et al. Chin J Radiol 2005; 30: 241-246

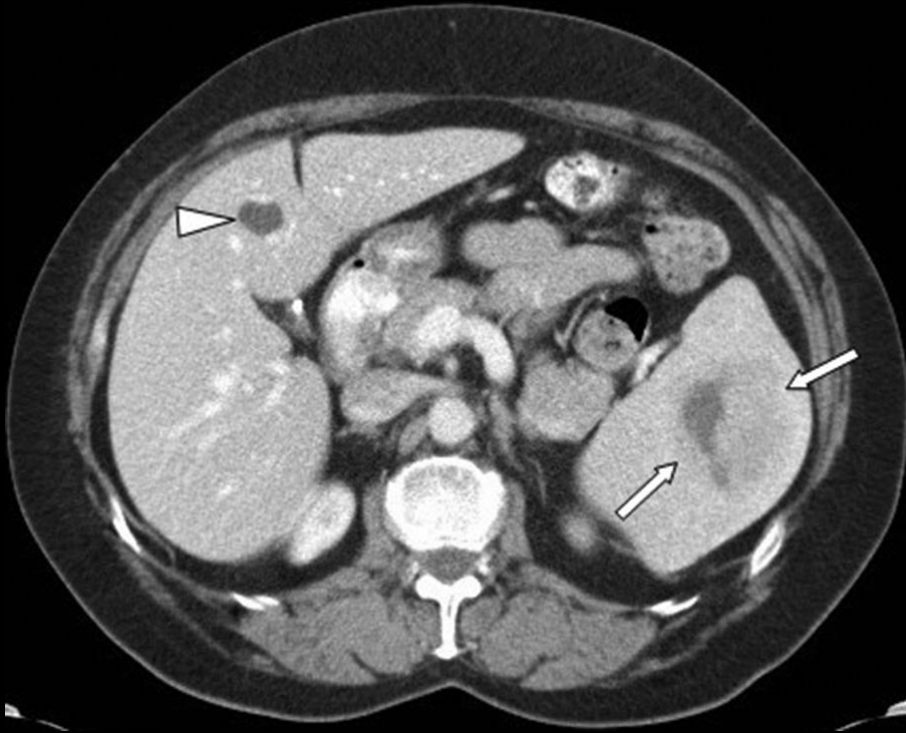
Cas 9



Collins PJ, Ettler H, Amann J, Rajgopal C: Soft-tissue images: splenic littoral cell angioma. Can J Surg 2003, 46(3):204-205

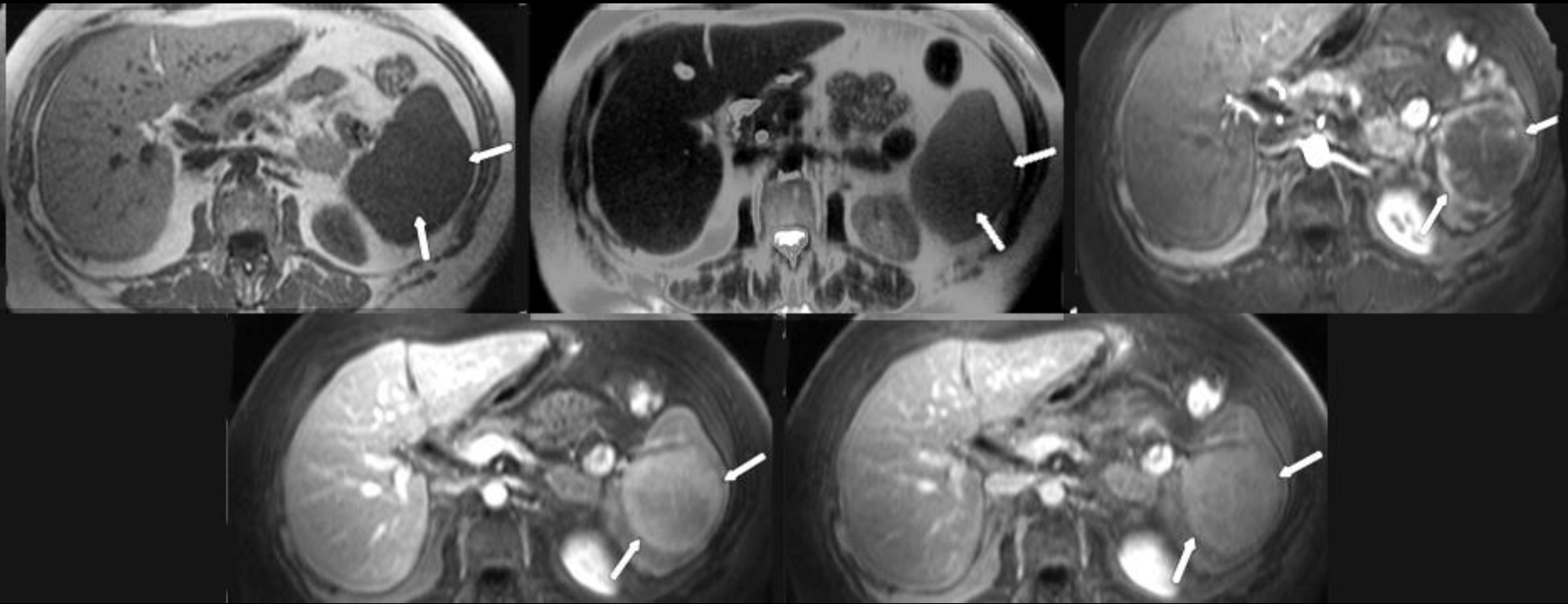
Cas atypiques de lésions uniques

Cas 1



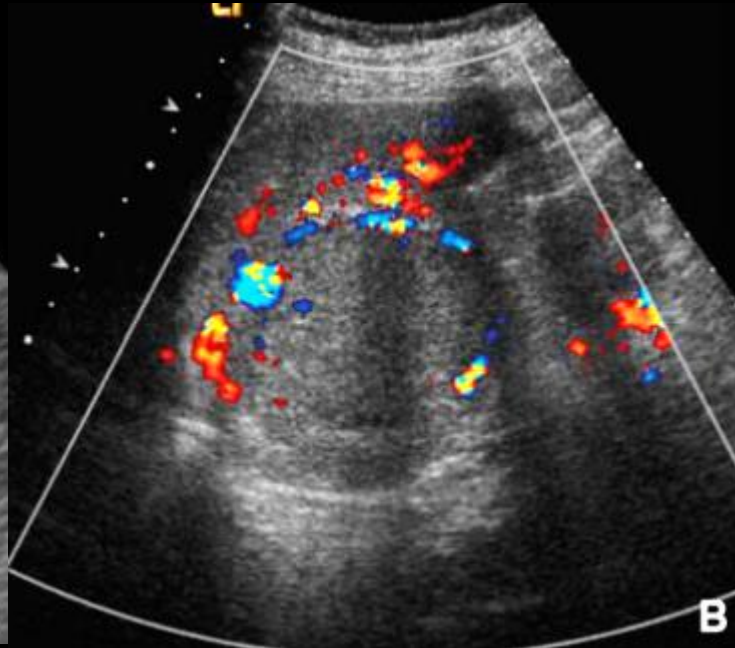
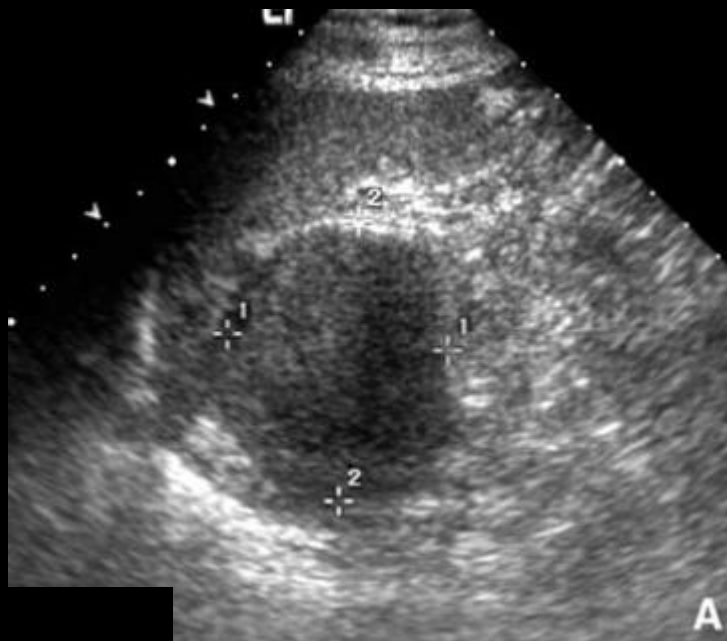
Contrast-enhanced axial computed tomogram image obtained during portal venous phase shows a large mass (arrows) in the spleen, the mass is showing diffuse homogeneous enhancement on the lateral aspect and the medial aspect is hypoenhanced.

A cyst in the liver is also demonstrated (arrowhead).



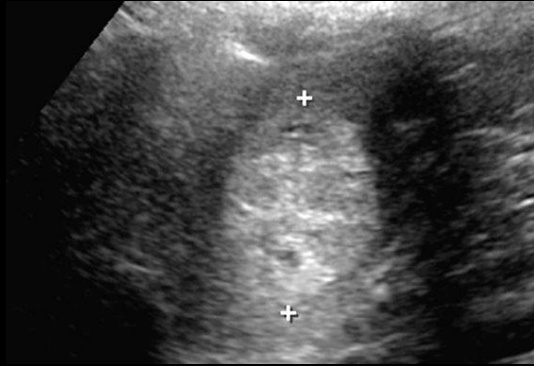
Axial magnetic resonance imaging images at the level of mid spleen show a large mass (arrows) which is isointense to the rest of splenic parenchyma before contrast injection on (a) T1-weighted and (b) T2-weighted images. (c) It shows peripheral nodular and internal septal enhancement during arterial phase on T1-weighted fat suppressed pulse sequence. (d) Diffuse enhancement during portal venous phase and (e) remains enhanced on delayed 3-min image.

Cas 2



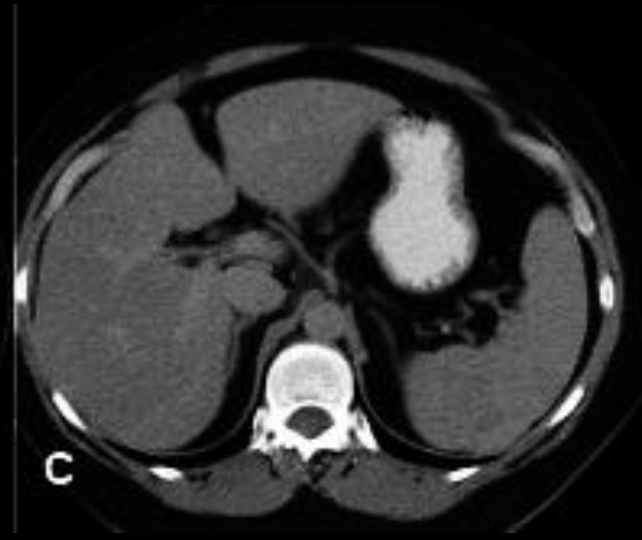
*Imagerie de la rate
pathologique (II). F.Mignon et
al. EMC 33-605-A-12*





Ultrasound of the spleen demonstrates a well-defined hyperechoic lesion. Tee et al. World Journal of Surgical Oncology 2008 6:87

CT scan after oral and iv-contrast in the arterial phase **A** and portal venous phase **B** demonstrates a hypodense well defined round lesion in the posterior portion of the spleen with some linear internal enhancement. **C**. The lesion is isodense compared to the normal spleen on the 5 minutes delayed image.



A. The lesion is hyperintense on the T2 weighted fast recovery fast spin echo (FRFSE) image. **B.** Internal linear enhancement is noted on the T1 weighted fat saturated image after iv-gadolinium in the portal venous phase

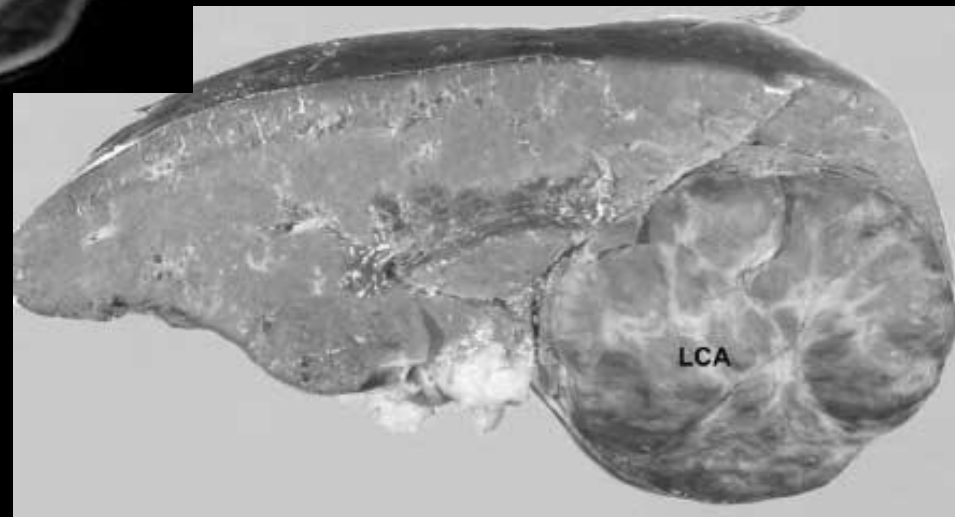
Cas 3

Cas 4



Contrast-enhanced CT at the level of the spleen reveals an enhancing intrasplenic mass (arrow) corresponding to the lesion shown on sonography

Gross appearance of the spleen. The tumor (LCA) appears as a lobulated mass, which was dark tan, adjacent to the splenic capsule.



Littoral Cell Angioma Sonographic and Color Doppler. Features Shweta Bhatt et al. J Ultrasound Med 2007; 26:539–542

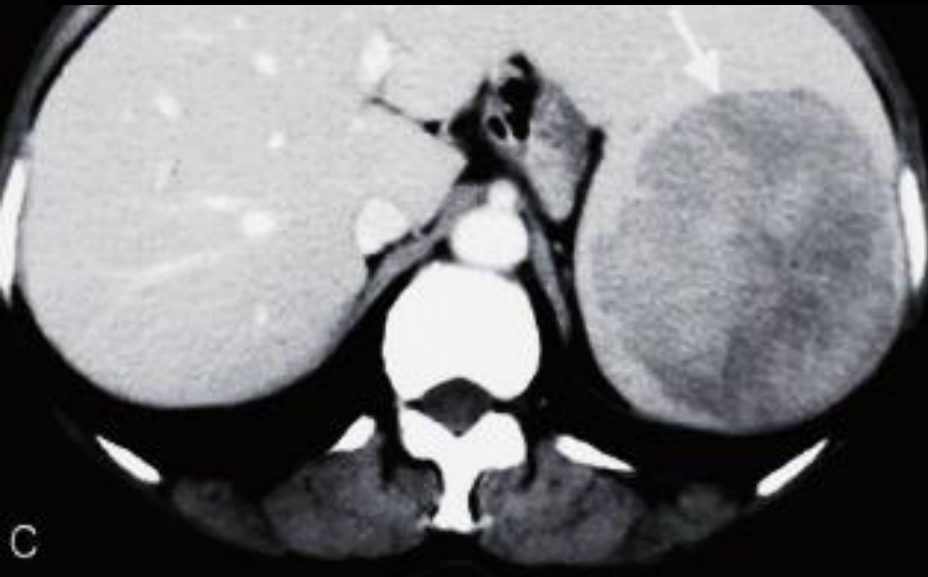
Cas 5

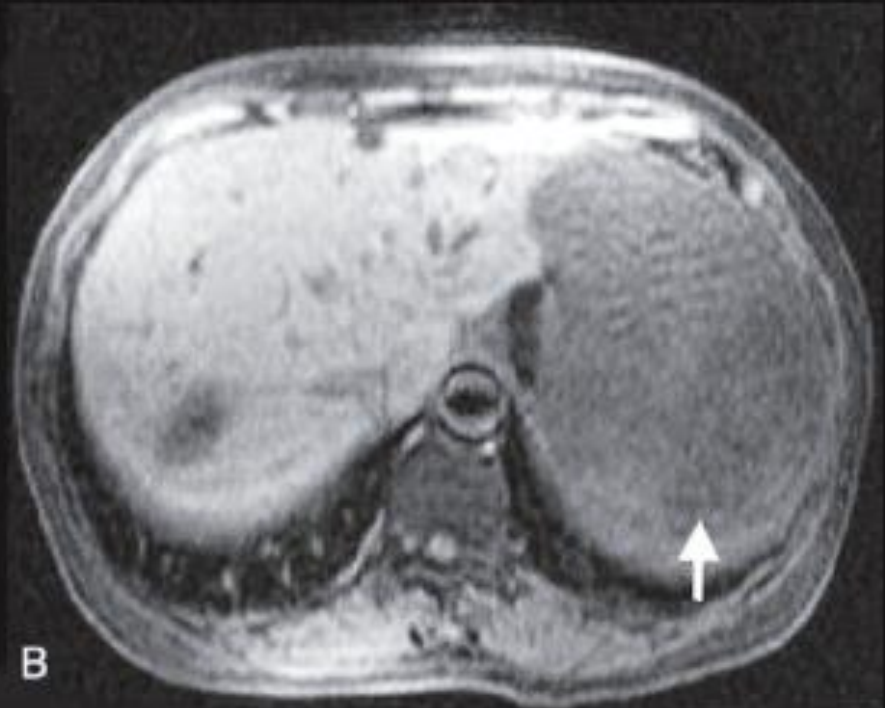


No splenic mass was seen on nonenhanced CT (A)

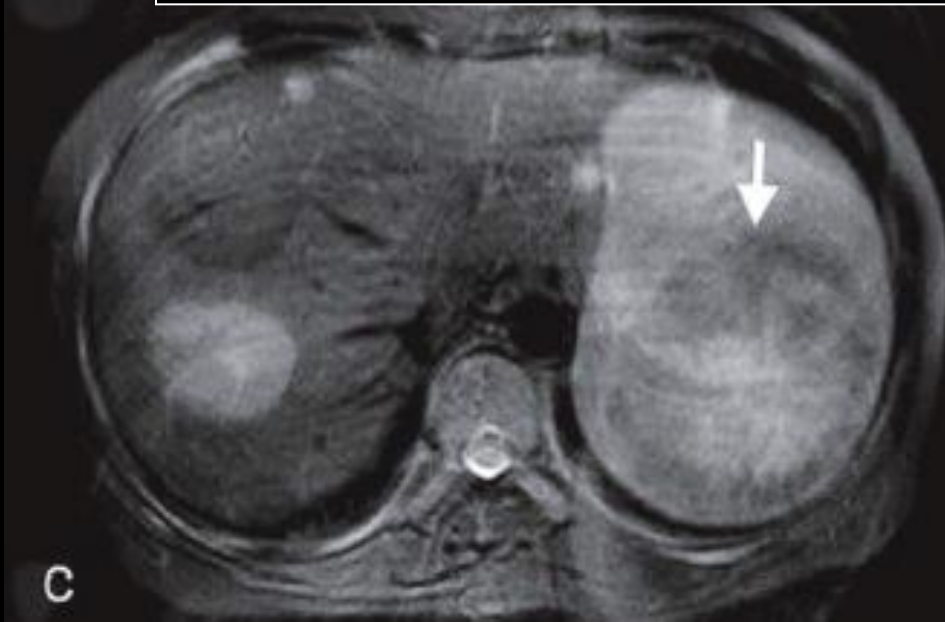
However, a large, solitary, low-density lesion appeared in the early portal venous phase of the contrast-enhanced CT (B and C)

This mass was isodense to the surrounding, enhancing normal splenic tissue on the delayed phase (D)





MRI showed the mass to be hypointense on both T1W and T2W images A and C



Anatomie pathologique

Macroscopie

- Macroscopiquement, lésion uni ou pluri-nodulaire, d'aspect spongieux.



Splenectomy-specimen revealing multiple nodular lesions with spongy appearance (arrows)

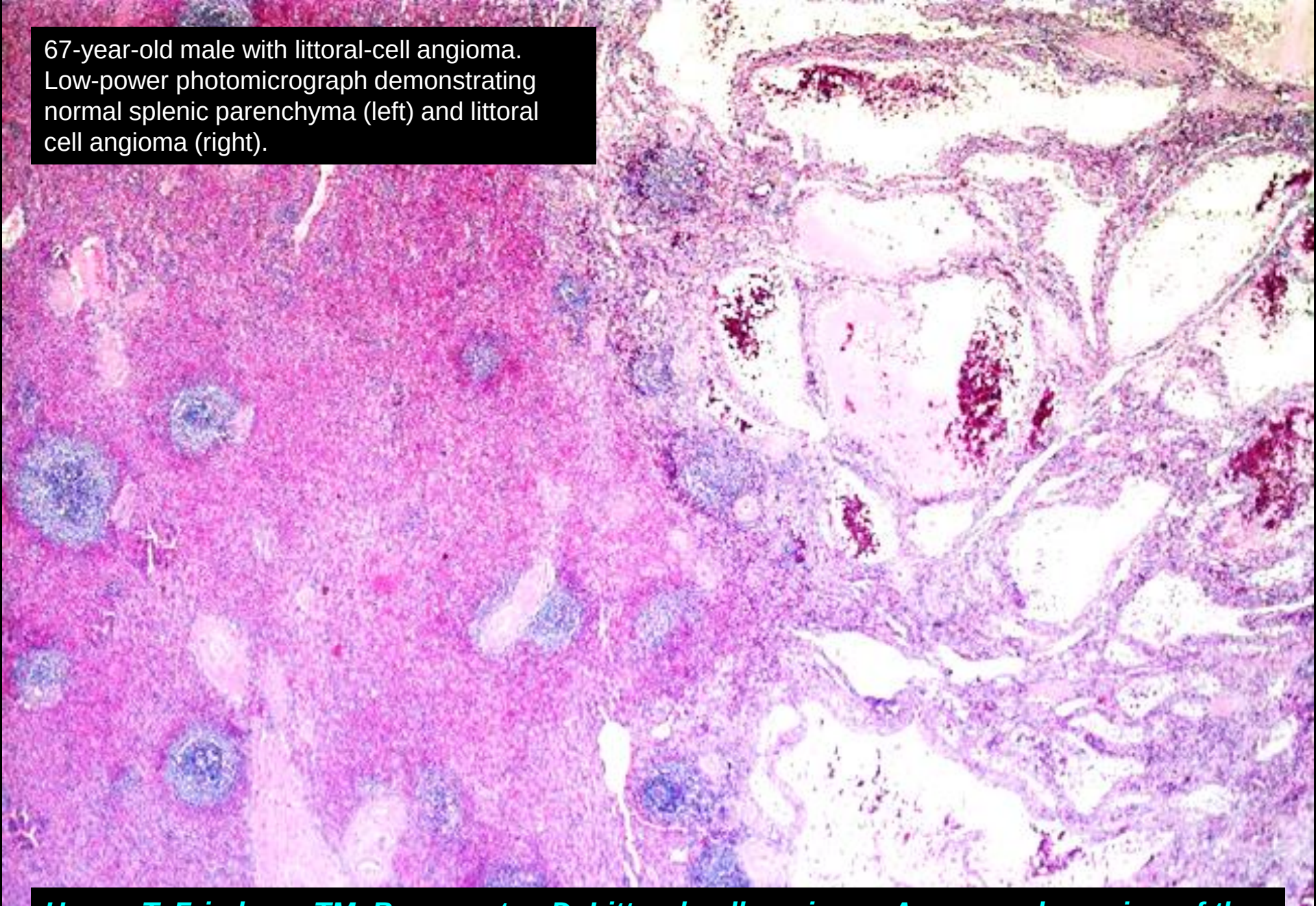


Cut surface of spleen studded with multiple well-defined tumor nodules

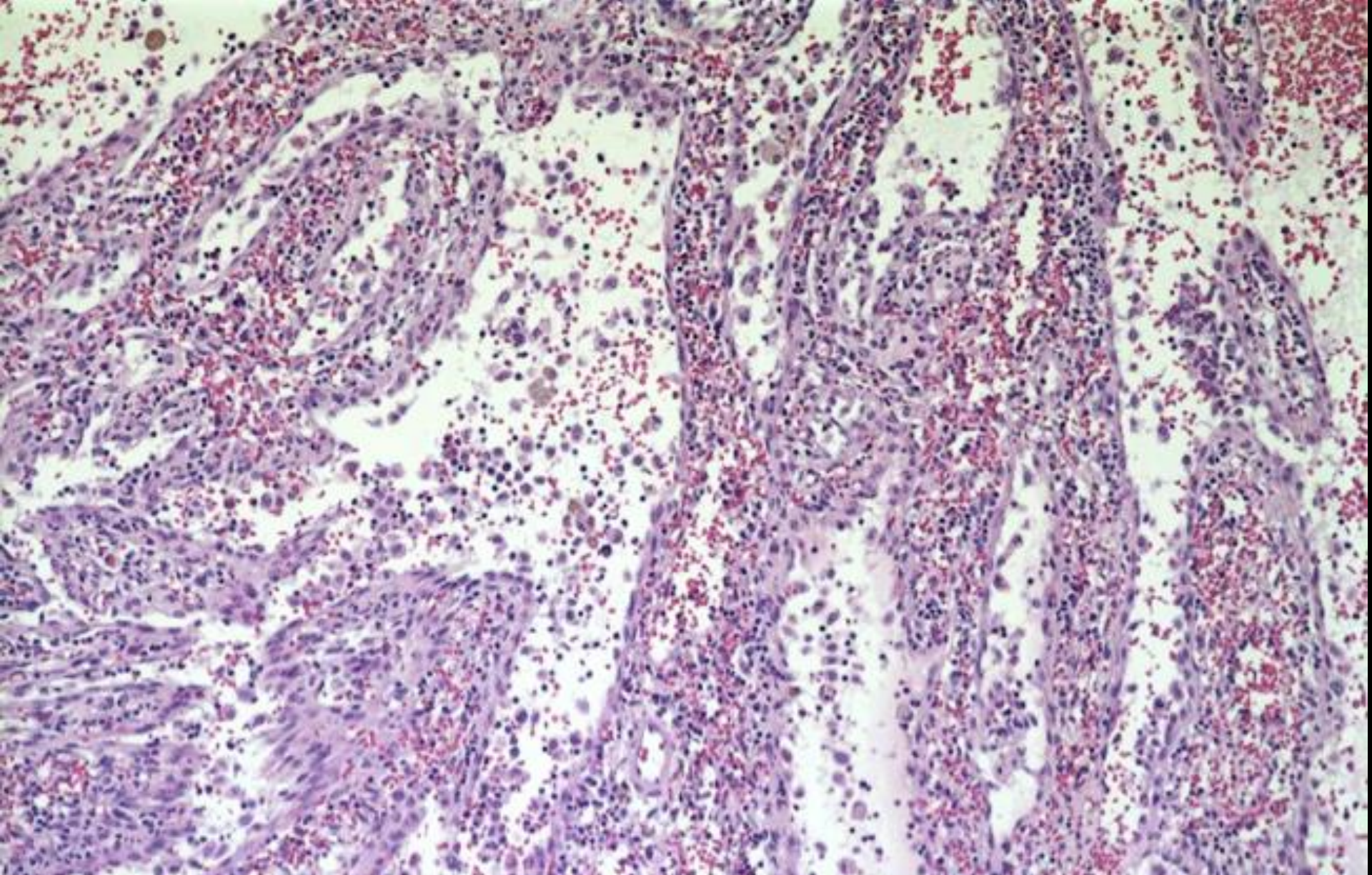
Microscopie

- Sur le plan morphologique, l'angiome à cellules littorales se caractérise par une **prolifération composée d'espaces vasculaires de taille variable, interconnectés**, comportant au sein de leur lumière **de longues projections papillaires**.
- Celles-ci sont tapissées de deux types de cellules. Les premières sont des **cellules endothéliales hautes** et sont au contact des lumières. Elles présentent de vastes cytoplasmes éosinophiles et des noyaux vésiculeux et nucléolés. Elles possèdent une tendance à l'exfoliation dans les lumières vasculaires où elles prennent alors un aspect presque histiocytaire. Les **autres, basales, sont endothéliiformes**, situées sous les premières au contact du chorion sous-jacent et présentent des noyaux allongés, sans atypie et des cytoplasmes peu abondants.
- L'angiome à cellules littorales se différencie de l'angiosarcome par l'absence de cellules atypiques, de nécrose ou d'invasion d'organes adjacents.

67-year-old male with littoral-cell angioma. Low-power photomicrograph demonstrating normal splenic parenchyma (left) and littoral cell angioma (right).

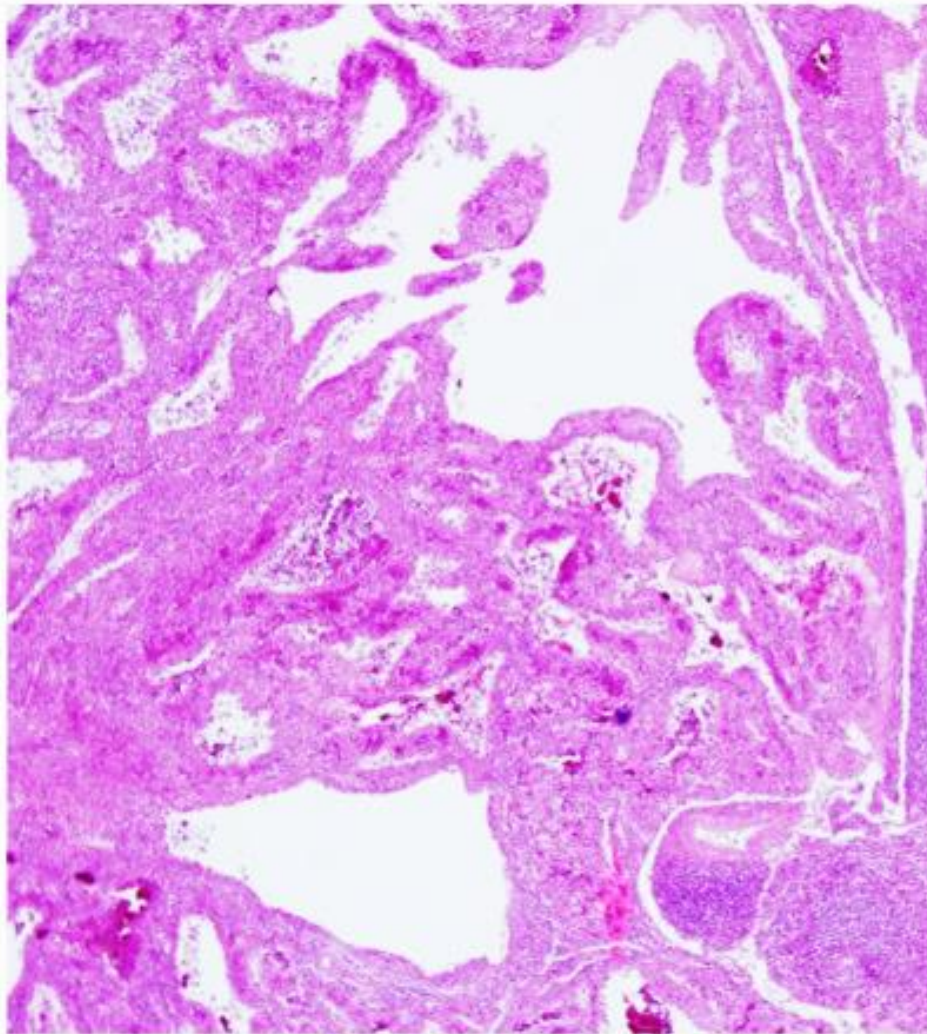


Hanna T, Friedman TM, Baumgarten D. Littoral-cell angioma: A case and a review of the literature. Radiology Case Reports. (Online) 2011;6:324.

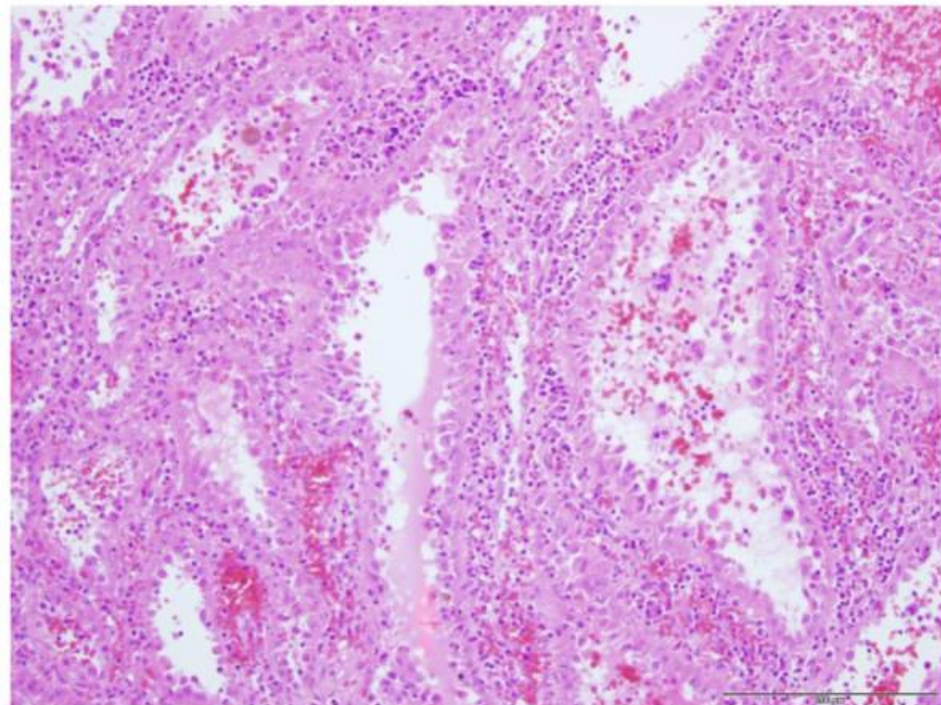
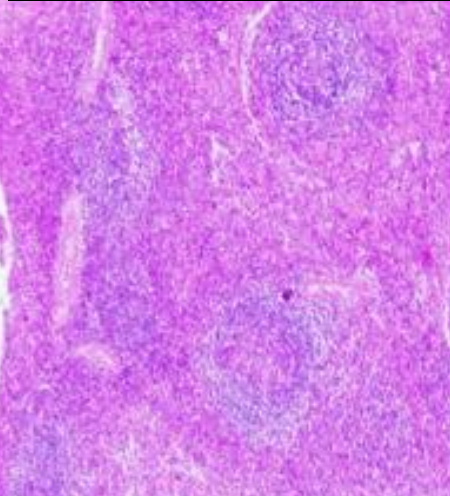


67-year-old male with littoral-cell angioma.

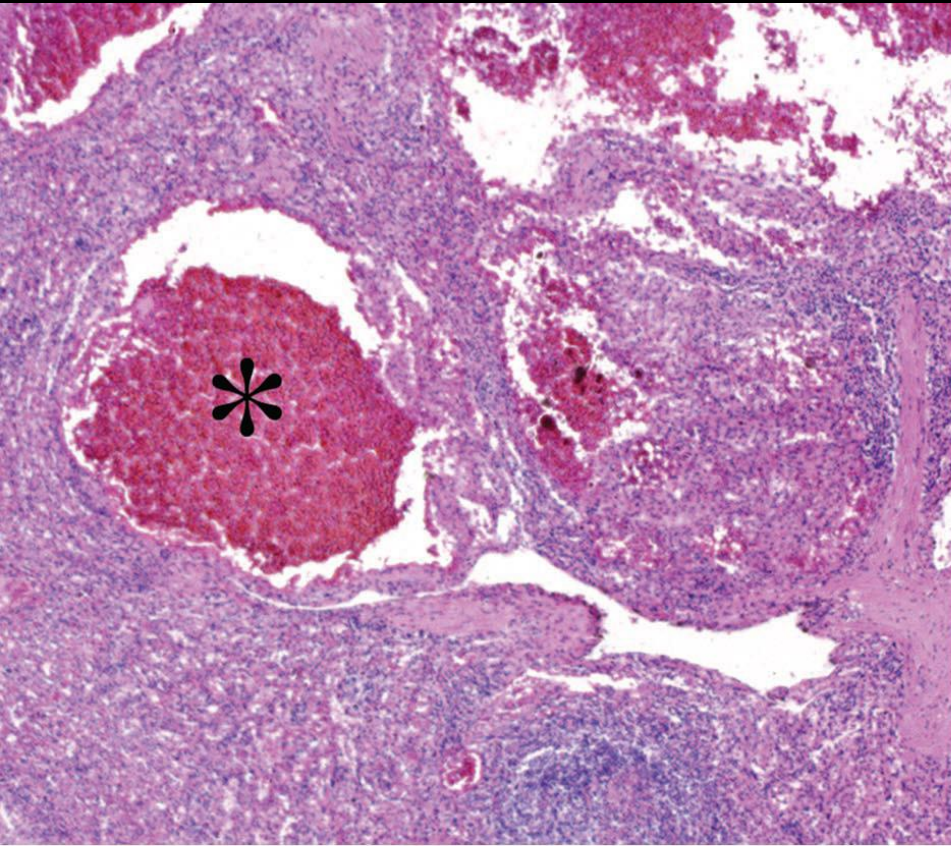
High-power photomicrograph of the lesion demonstrates anastomosing vascular channels lined by tall endothelial cells that slough into the lumen, characteristic of littoral-cell angioma.



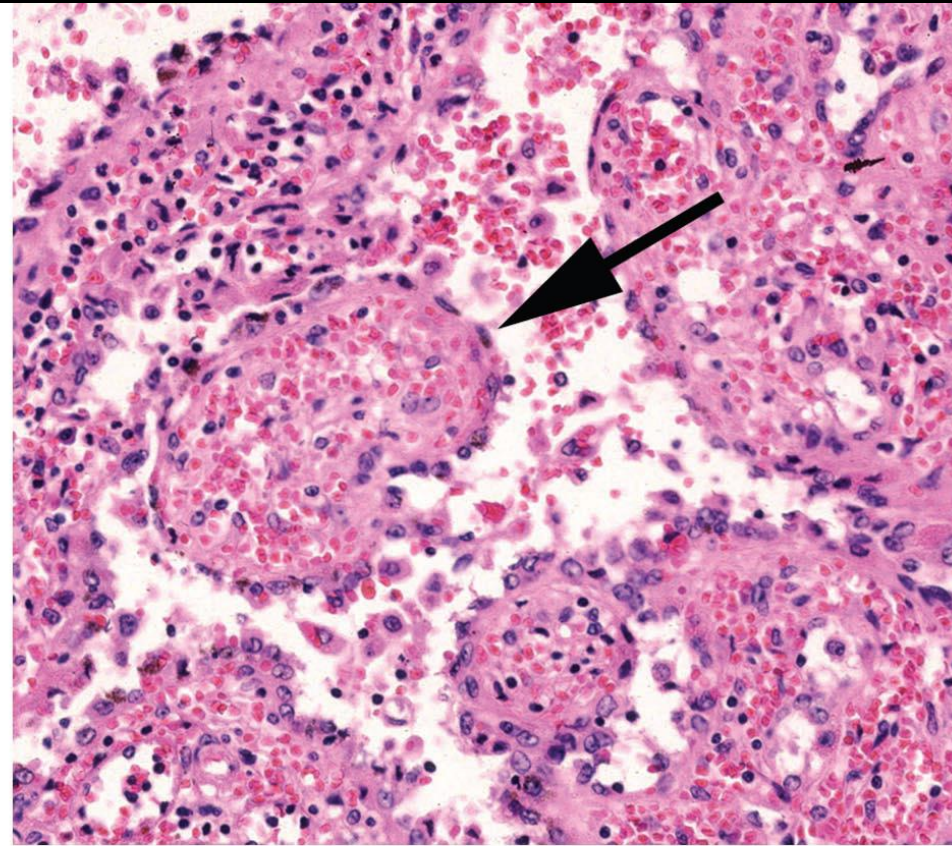
Regular splenic parenchyma (right) and tumor (left) composed of lacunae filled with oedematous fluid and blood (4× obj, HE-staining).



Neoplastic sinuses lined by a single cell layer (20× obj, HE-staining).



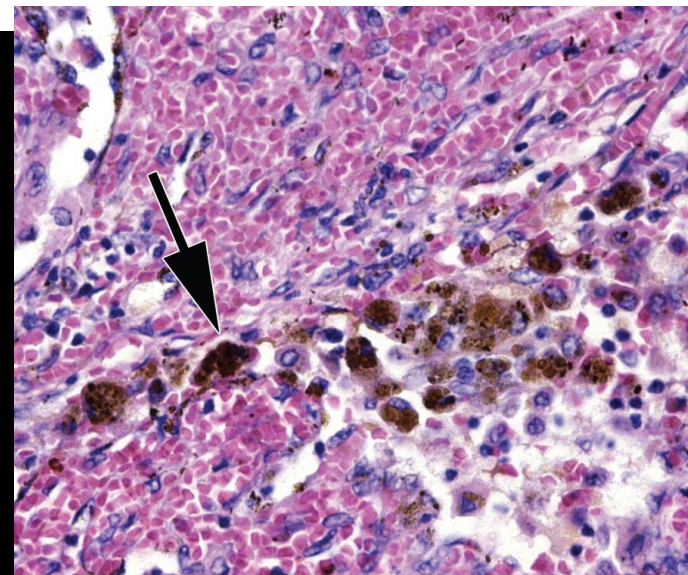
a.



b.

Littoral cell angioma

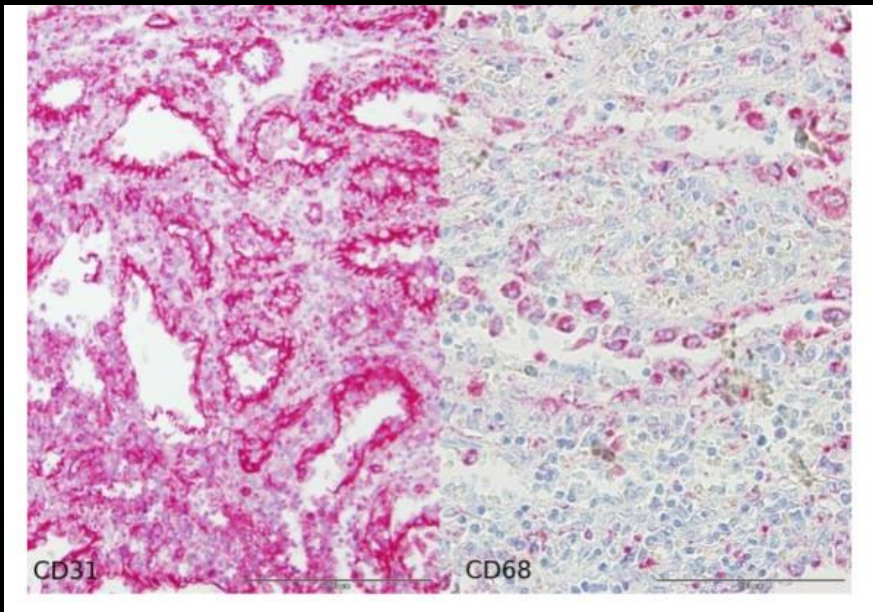
- (a) Photomicrograph (original magnification, 10; H-E stain) shows multiple blood-filled spaces (*) within a focus of littoral cell angioma.
- (b) Photomicrograph (original magnification, 20; H-E stain) shows papillary projections (arrow) extending into channels containing exfoliated cells.
- (c) Photomicrograph (original magnification, 40; H-E stain) shows hemosiderin pigment (arrow) within littoral cell angioma.



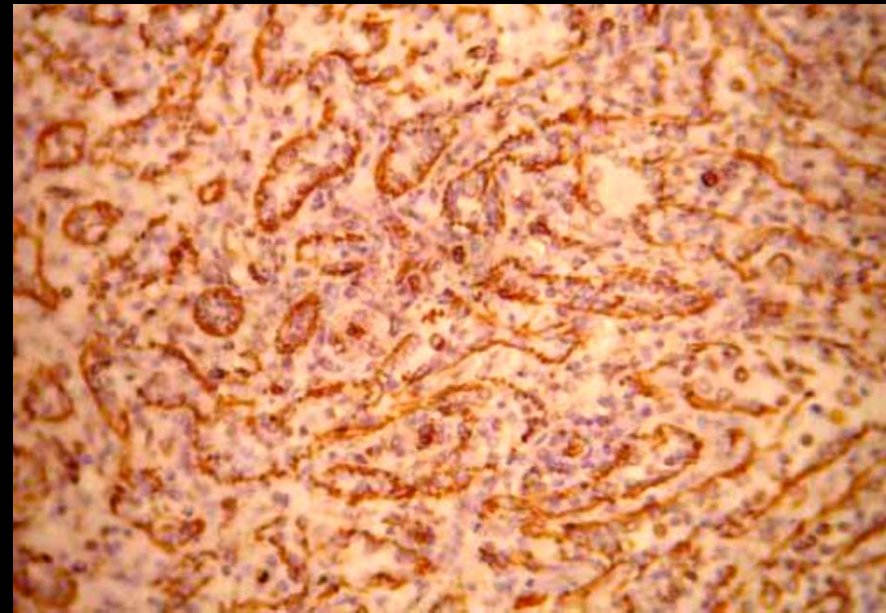
Immunohistochimie

- Le phénotype de ces tumeurs est très particulier associant un **double phénotype vasculaire et histiocytaire (CD31 et CD68/KP1)**
- La composante basale exprime également le CD34 alors que les grandes cellules sont négatives
- Le CD21 est parfois positif, le CD8 est négatif

Littoral Cell Angioma: A Morphologic and Immunohistochemical Study. Mohsen Emami Aleagha¹ et al. Iranian Journal of Pathology (2009)4 (2), 88 - 91

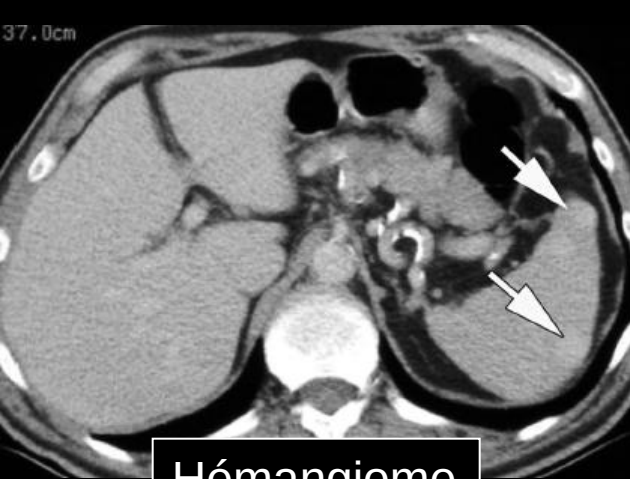


Combined expression of endothelial (CD31) and histiocytic (CD68) markers in immunohistochemical staining.

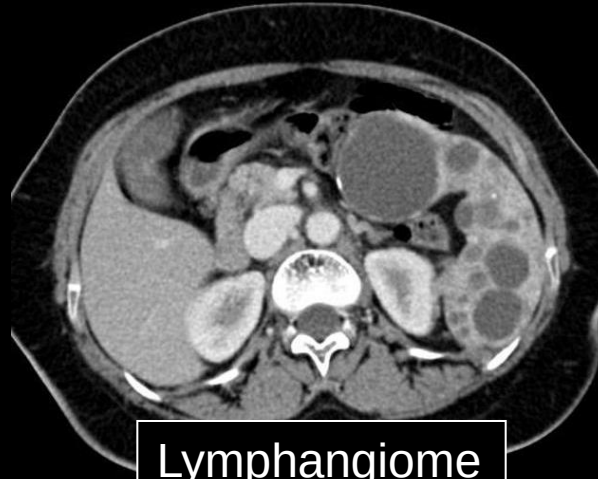


The lining cells expressed CD34 (*100 magnification. IHC Staining).

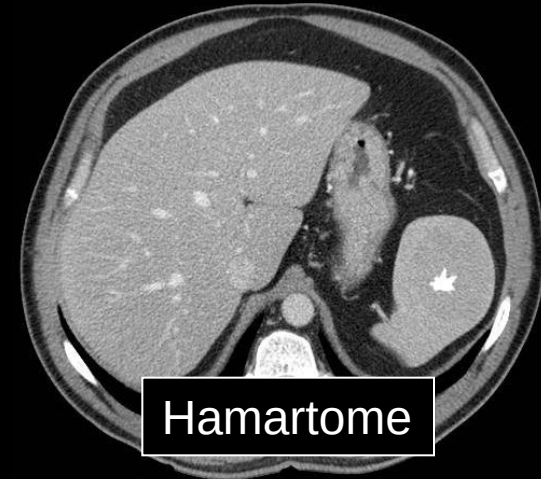
Principaux diagnostics différentiels



Hémangiome



Lymphangiome



Hamartome



Hémangioendothéliome



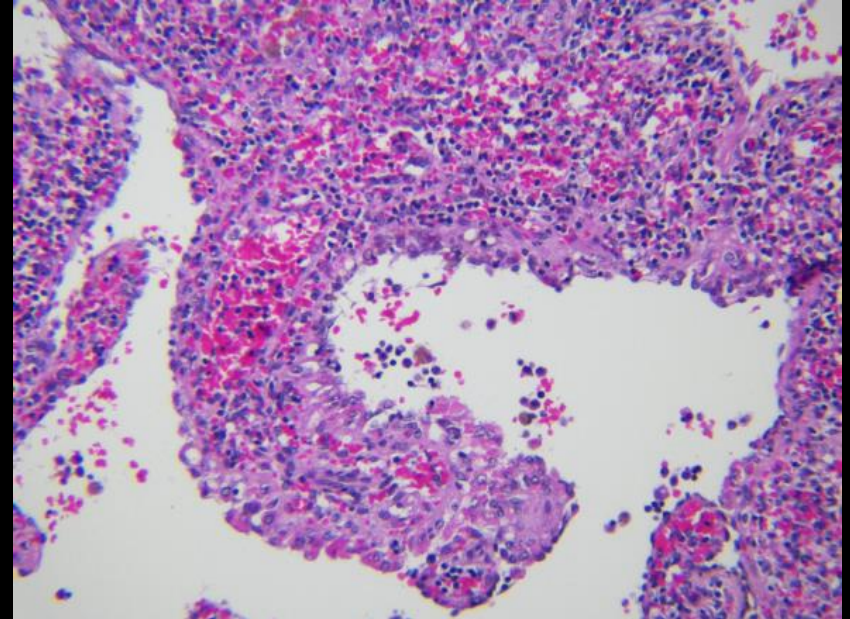
Angiosarcome



Péliose

Take home message

- Tumeur vasculaire rare, bénigne
- Se développe à partir des cellules littorales des sinus de la pulpe rouge de la rate
- SR = 1; Pas d'âge de prédilection
- En général asymptomatique; possibles manifestations d'hypersplénisme
- Diagnostic anatomopathologique ...



- **Echo** : Aspect marbré de la rate avec nodules pouvant être iso-échogènes, hypo-échogènes ou hyper-échogènes; Vascularisation centrale et périphérique au doppler
- **TDM** : Splénomégalie; 2 formes :
 - multiples nodules de taille variable, hypodenses spontanément et au temps portal avec homogénéisation à la phase tardive (la + fréquente);
 - lésion unique (plus rarement)
- **IRM** : Nodules spléniques **hypo-intenses en pondérations T1 et T2, reflétant la présence d'hemosidérine**

