# A *Littoral* Surprise: a Rare Case of Littoral Cell Angioma

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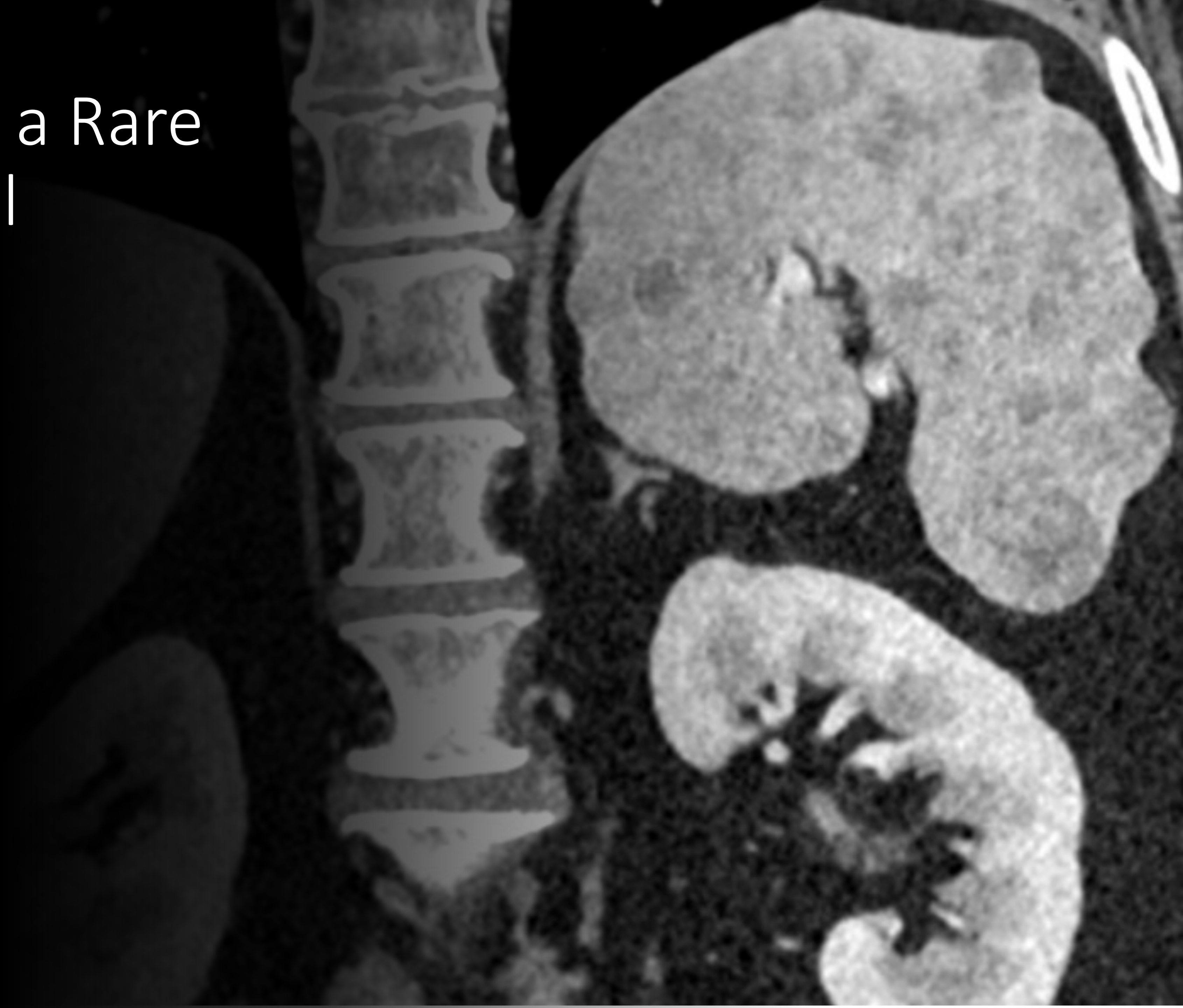


# Background

- **Littoral cell angioma (LCA) is** a benign primary splenic tumor which is classically difficult to differentiate from malignant pathologies on imaging alone.

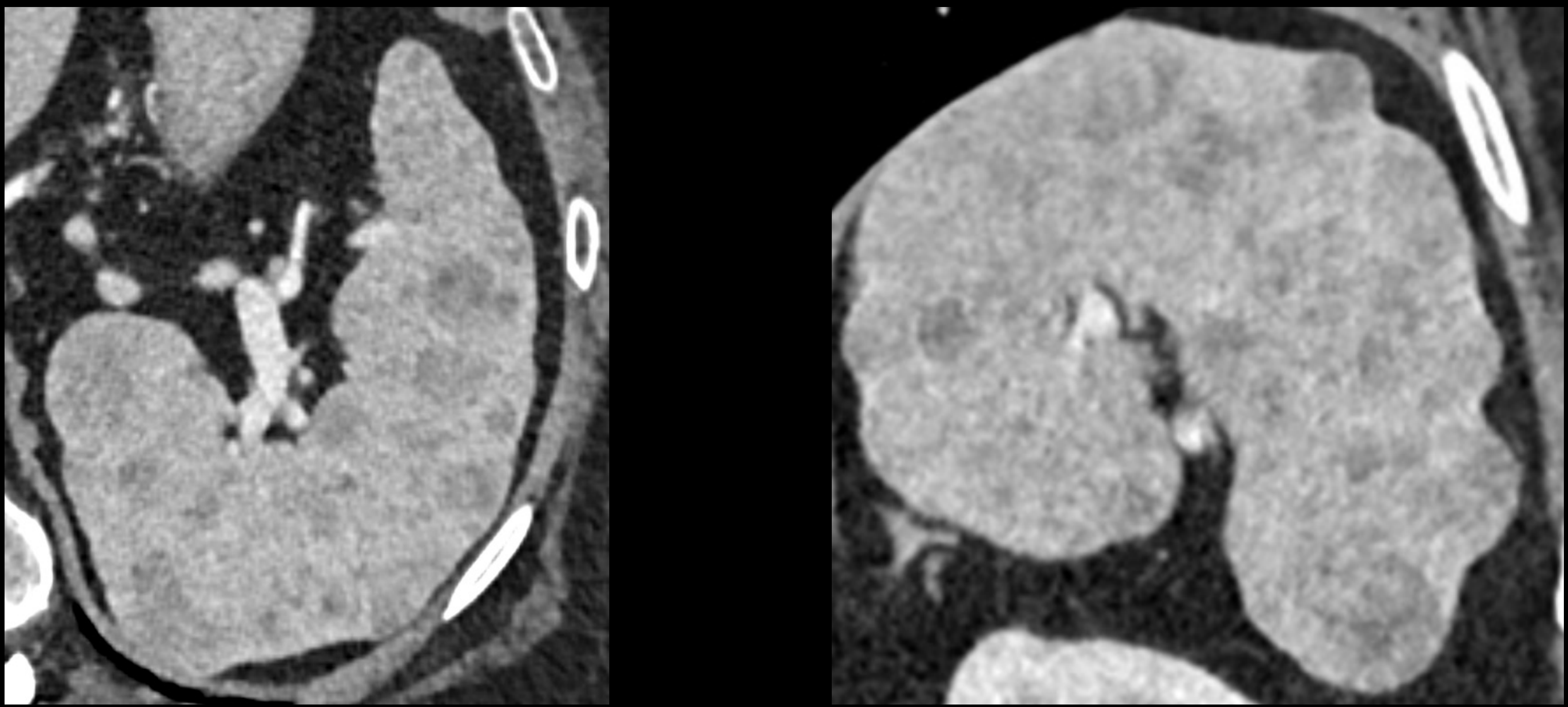
- It has a **vascular origin composed of connecting vessels** and is derived from the littoral cell in red pulp. [1]

- It is often an **incidental** finding and should be considered in the differential for multiple splenic rounded lesions.

# Case introduction

- A patient in their 70s presented to the ED and had contrast enhanced CT imaging for trauma.
- They had no relevant medical history and no history of weight loss, fever or night sweats.
- Full blood count was normal.

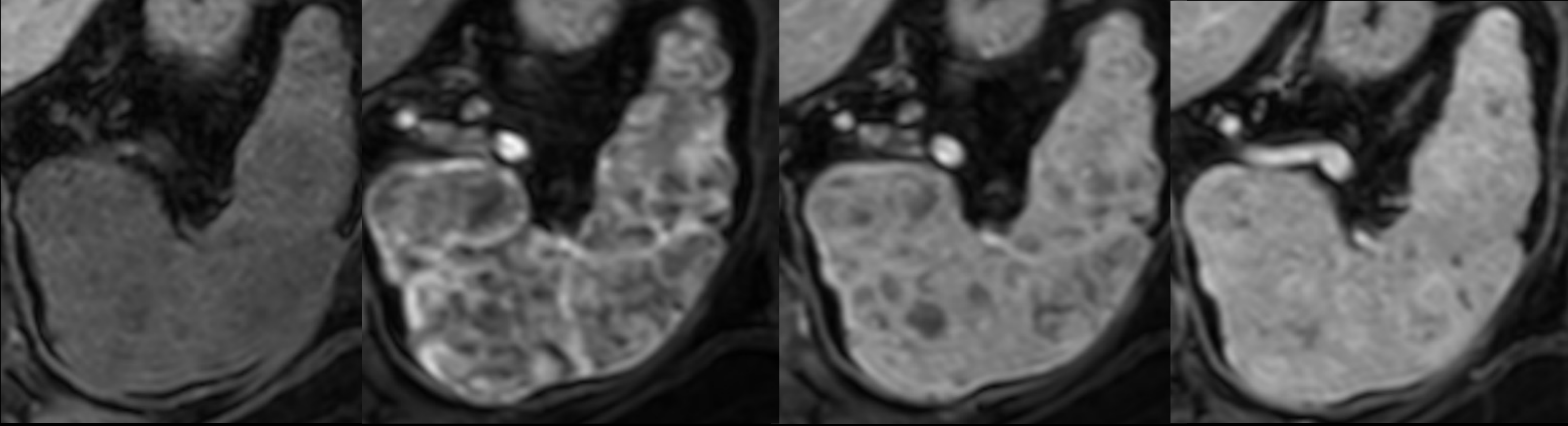
# Initial CT findings:



Axial and coronal CT images demonstrating **splenomegaly** to 14.5cm
with **multiple rounded hypodense nodules** scattered throughout the spleen.

# Subsequent contrast enhanced MRI:

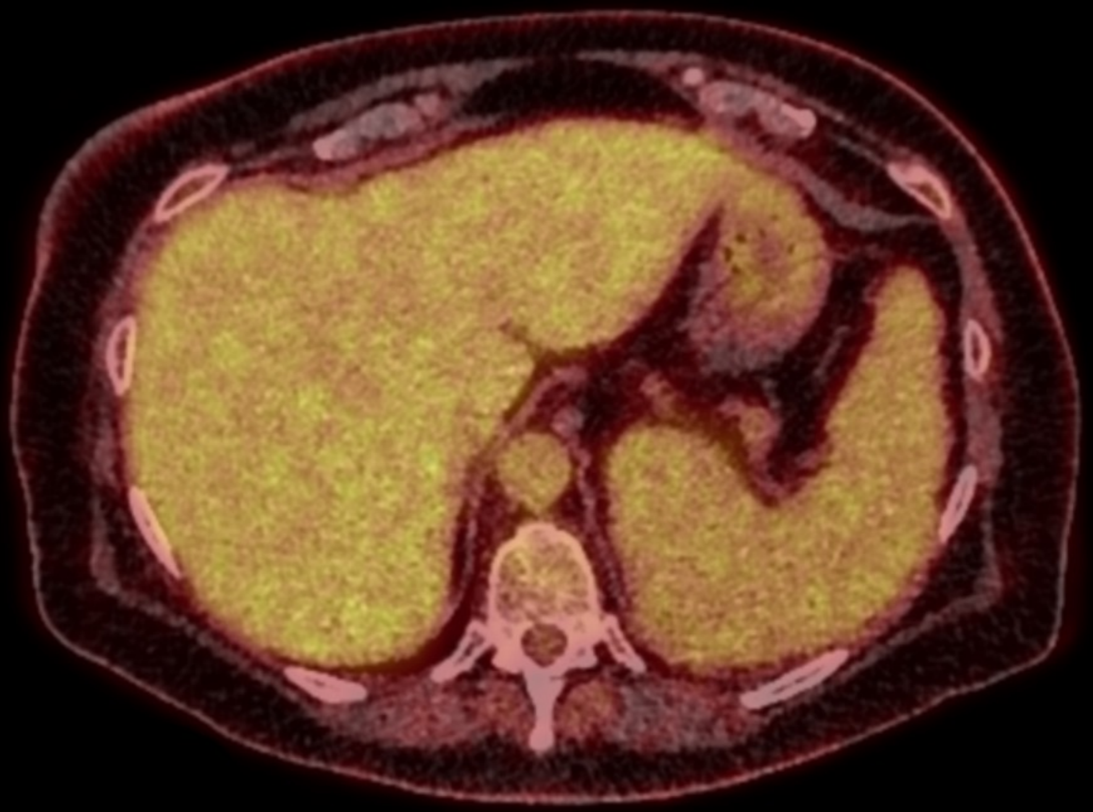
T2 axial and coronal images demonstrate **multiple splenic iso/hyperintense T2 signal** lesions with internal **punctate** foci of **low** signal.



Pre contrast axial T1 | Arterial phase | Portal venous phase | Delayed phase

Pre and post contrast T1 images of the spleen demonstrate **progressive delayed centripetal enhancement.**

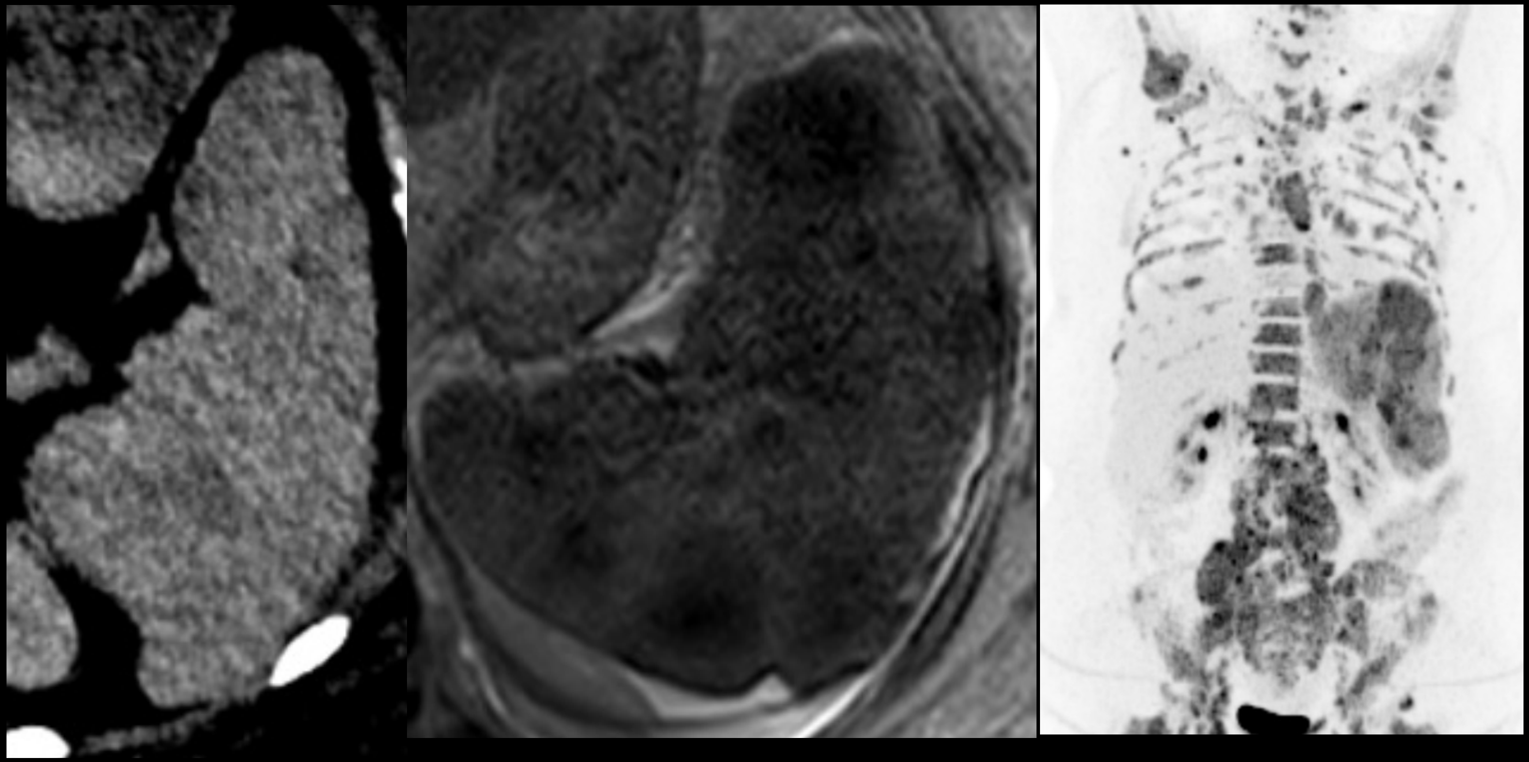
# Subsequent FDG PET- CT:



Fused axial PET-CT and coronal MIP images showing only physiological background activity in the spleen **without focal increased uptake** in the splenic lesions.

# Differential diagnosis:

The differential for multiple splenic lesions includes **lymphoma, metastases, haemangiomas, sarcoidosis if solid; if cystic,** differentials include **lymphangioma and infection.**



**Lymphoma:**

Axial contrast enhanced CT (left): multiple ill defined **hypodense** lesions, with splenomegaly
MRI T2 axial (middle): multiple **hypointense** lesions relative to background splenic parenchyma.
PET-CT coronal MIP (right): these splenic lesions are **FDG avid** amongst extensive nodal disease elsewhere.
Lesions typically show absent or mild enhancement (not shown)

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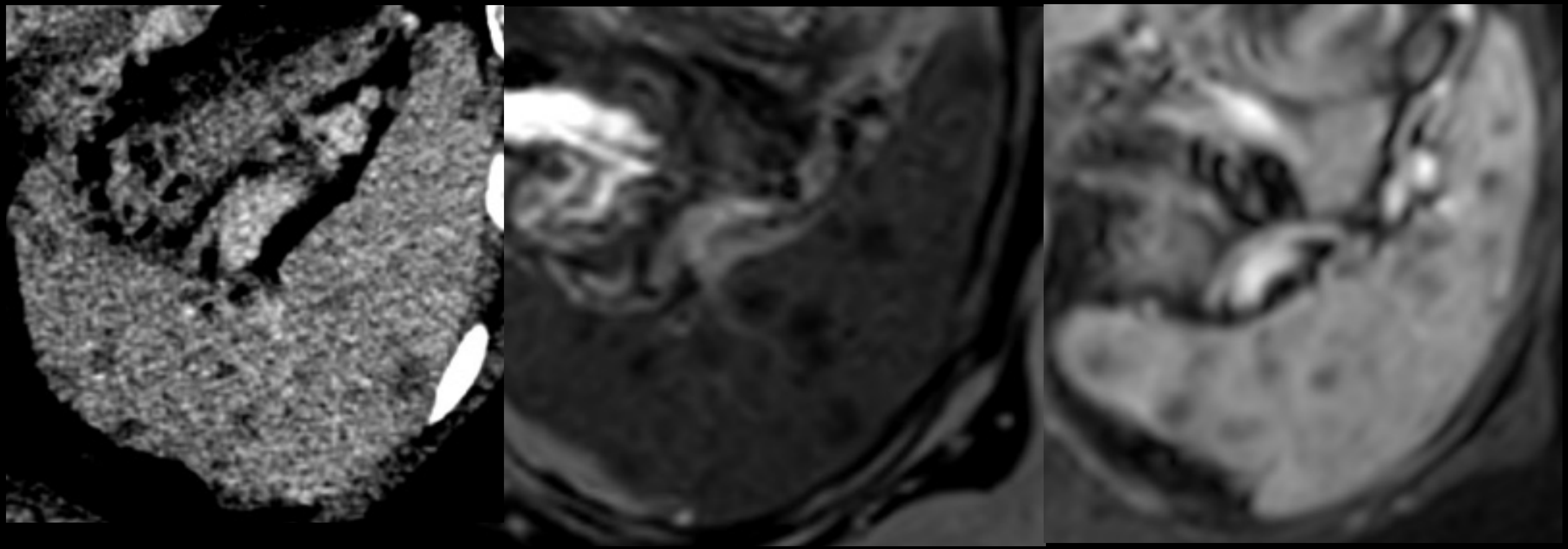
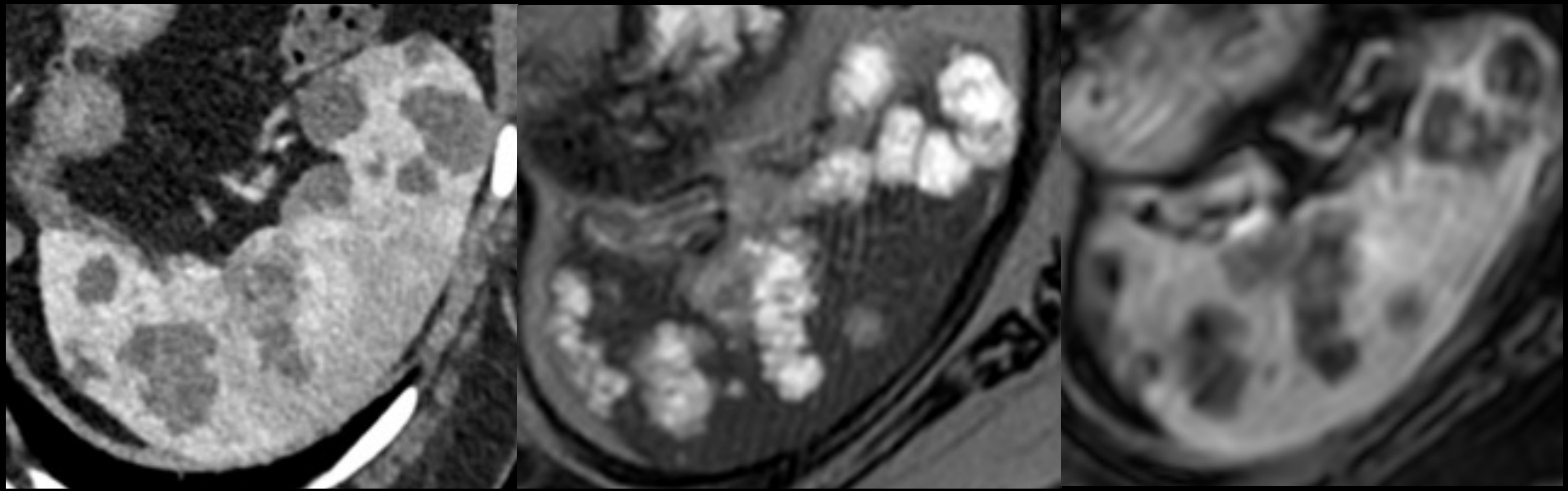


**Splenic sarcoidosis:**

Axial contrast enhanced CT (left): multiple **hypodense** lesions relative to background splenic parenchyma.
MRI T2 axial (middle): multiple **hypointense** lesions relative to background splenic parenchyma.
MRI axial post gadolinium portal venous phase (right): these lesions are **non-enhancing**.

# Differential diagnosis:

The differential for multiple splenic lesions includes **lymphoma, metastases, haemangiomas, sarcoidosis if solid; if cystic,** differentials include **lymphangioma and infection.**



**Lymphangiomas:**

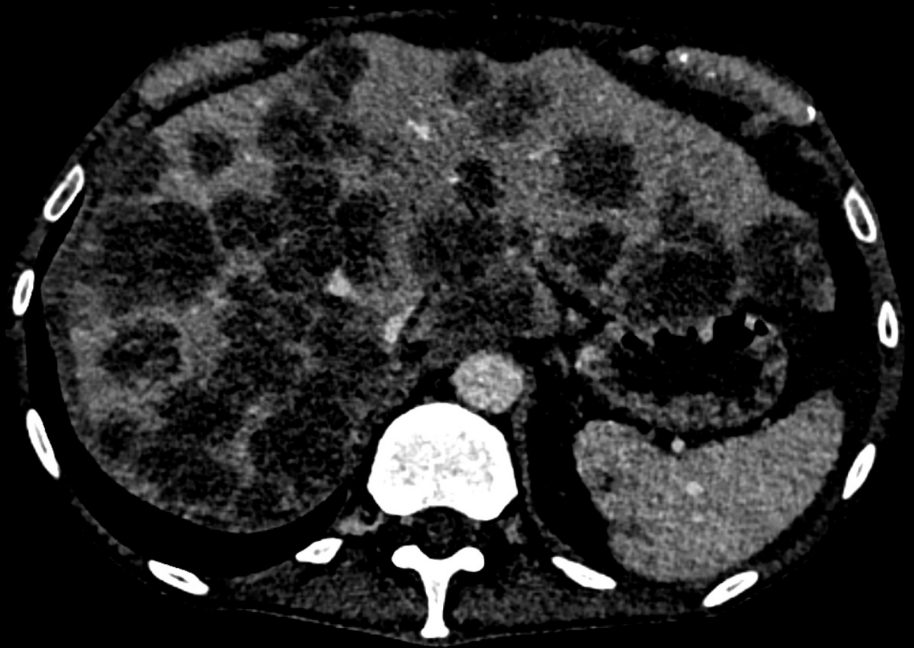
Axial contrast enhanced CT (left): multiple **cystic hypodense** lesions relative to background splenic parenchyma.

MRI T2 axial (middle): multiple heavily septated **hyperintense** lesions.

MRI axial post gadolinium portal venous phase (right): these lesions are **non-enhancing.**

# Differential diagnosis:

The differential for multiple splenic lesions includes **lymphoma, metastases, haemangiomas, sarcoidosis if solid; if cystic,** differentials include **lymphangioma and infection.**



**Splenic metastasis:**

Axial contrast enhanced CT portal venous phase: shows multiple ill defined **hypodense** splenic lesions, in a patient with colorectal cancer and **widespread metastatic disease** elsewhere (for example, in the liver on this slice).

Lesions can be **solitary** or **multiple** (as in this case).

On MRI, lesions are typically T1 hypointense (except melanoma), T2 hyperintense and show variable enhancement characteristics based on the primary tumour.

# Management and histological confirmation

- **Splenectomy** is often **favoured** over biopsy to confirm LCA with no atypia due to risk of bleeding and to avoid tumour seeding if subsequently found to have malignant features as in littoral cell angiosarcoma. [2]
- MDT in this case recommended elective splenectomy given imaging suggestive of LCA, which was performed without complication.
- Histological analysis confirmed morphological and immunohistochemical features, such as expression of **CD31 and CD68**, in keeping with **LCA without atypia**. [3]

# Learning points

1. Our case highlights the **non-specific but classical imaging** findings of LCA.
2. The radiologist's **suspicion** should remain **high** with careful attention to the **T2 and enhancement characteristics** to include LCA in the **differential** for multiple splenic lesions as well as more common causes such as lymphoma, metastases, haemangiomas and sarcoidosis.
3. T2 signal characteristics are dependent on the **amount of hemosiderin** in each lesion.[4]
4. Morphological characteristics on imaging such as **direct invasion** of adjacent structures may be able to distinguish benign Littoral cell angioma from malignant Littoral cell angiosarcoma- a diagnosis of LCA should **invariably be confirmed on histology.**
5. Although often incidental, LCA can present with **complications** such as **rupture** giving acute pain. [5]

# References


1. Rai, M. and Gupta, N., 2023. Littoral cell splenic angioma. In *StatPearls [Internet]*. StatPearls Publishing.
2. Machado Júnior, P.A.B., Pereira, C.D.O., Bortolo, J.L.D., Martins, A.L.C.L., Campos, H.G., Aguiar, A.J.D., Postai, R.P.T.A., Wiederkehr, J.C. and Wiederkehr, H.D.A., 2024. Littoral cell angioma of the spleen: case report and literature review. *einstein (São Paulo)*, *22*, p.eRC0267.
3. Borch, W.R., Aguilera, N.S., Brissette, M.D., O'Malley, D.P. and Auerbach, A., 2019. Practical applications in immunohistochemistry: an immunophenotypic approach to the spleen. *Archives of pathology & laboratory medicine*, *143*(9), pp.1093-1105.
4. Zeng, Q.Y., Liao, J., Song, Y., Zhang, D.Q., Li, H. and Wang, J., 2023. Multimodality imaging findings of splenic littoral cell angioma: a case description and analysis of literature. *Quantitative Imaging in Medicine and Surgery*, *13*(4), p.2740.
5. McHugh, A., O'Gorman, C., Cooley, S., McEntee, G. and Burke, N., 2018. Littoral cell angioma with splenic rupture in pregnancy. Case Reports in Perinatal Medicine, 7(1), p.20170064.