

Littoral Cell Angioma of Intrapancreatic Splenule within the Pancreatic Tail

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Introduction

- Littoral cell Angioma (LCA) is a rare, potentially malignant tumor which originates from the red pulp sinuses of the spleen.
- While typically benign, the rate of malignant transformation is unclear, and malignancy cannot be determined until final pathology.
- LCA has the potential to affect accessory spleens (splenules), which may exist in multiple locations within the abdomen, most commonly the splenic hilum.
- Few cases in the literature have reported LCA of splenules, and only describe splenules within the hilum of the spleen.
- We present a unique case of a patient with LCA affecting both the spleen and an intrapancreatic splenule within the pancreatic tail.

Case Presentation

- Patient presented to PCP with dyspnea and occasional Left Lower Quadrant pain.
- She underwent CT-PE to which revealed splenic lesions (Figure 1).
- Follow up MRI showed multiple enhancing lesions of the spleen , as well as as lesion in the pancreatic tail (Figure 2).
- This was suspected to be a neuroendocrine tumor with metastases to the spleen
- IR Biopsy showed dual histiocytic and endothelial expression confirming the condition of Littoral cell angioma (Table 1).
- Later PET scan showed no evidence of hypermetabolic primary or metastatic disease.

Workup

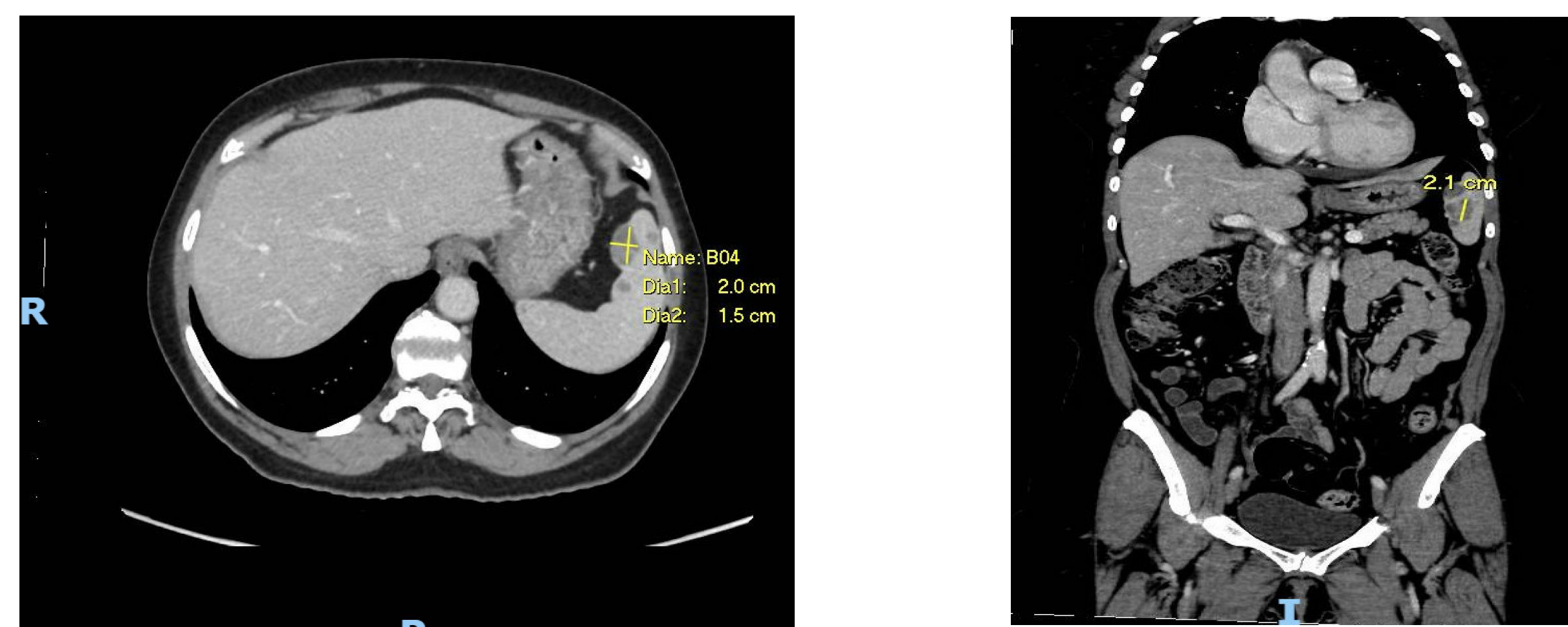


Figure 1. Transverse and Coronal CT scan of abdomen, with splenic lesion marked.

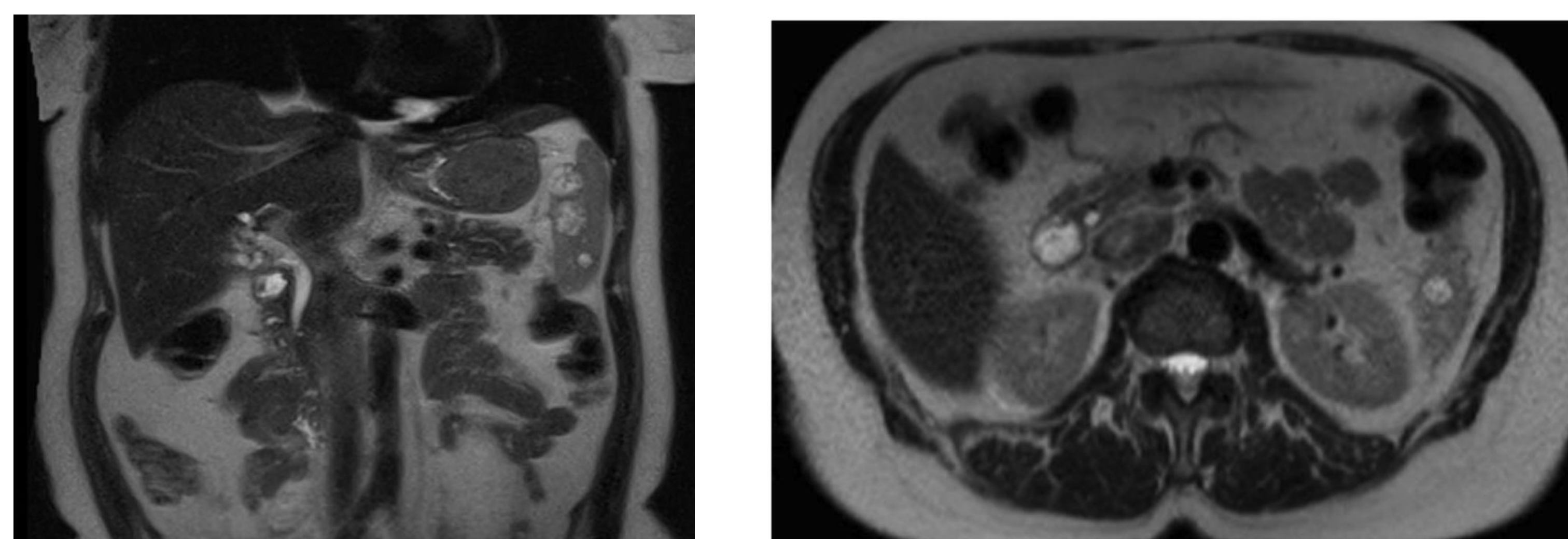


Figure 2. Magnetic Resonance Imaging showing T2 hyperintense/ mildly T1 hypointense heterogeneously/ progressively enhancing lesions in the spleen with the largest in the anterior aspect of the spleen measuring 2.1cm x1.5cmx1.7cm.

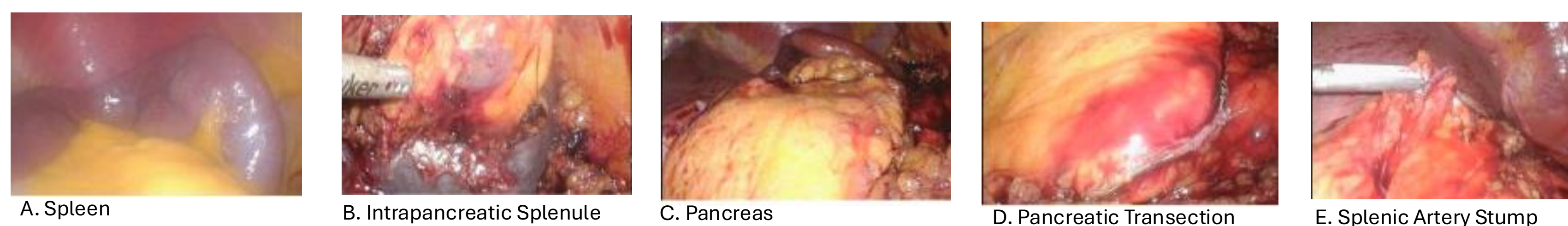


Figure 3. Images captured during laparoscopic splenectomy.

Table 1. Pathology Results from IR Biopsy of the Spleen, April 2023

Immunostain	Result	Typical LCA Result
CD8	Negative	Negative
CD31	Positive	Positive
CD34	Negative	Negative
CD68	Positive	Positive
ERG	Positive	Positive
HHV8	Negative	Negative
Langerhan	Negative	Positive

Management

- The tumor board recommended laparoscopic splenectomy and distal pancreatectomy.
- Patient elected for this surgery over the alternative of serial imaging and underwent a Splenectomy and Distal Pancreatectomy two months later.
- Diagnostic Laparoscopy showed no evidence of metastatic disease or any other abnormalities.

Discussion

- There are no official guidelines for treatment or surveillance of LCA.
- The options of surveillance and resection should be discussed thoroughly with patients presenting with LCA to help guide a patient-centered management approach.
- There are rare cases of LCA progressing to malignancy, and recurrence in remaining splenic tissue.

Conclusion

- Patient tolerated procedure well and surveillance imaging has not demonstrated recurrence
- Minimally-invasive management of LCA should be considered as a treatment option when faced with presented with this rare diagnosis

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