

CASE OF THE FORTNIGHT



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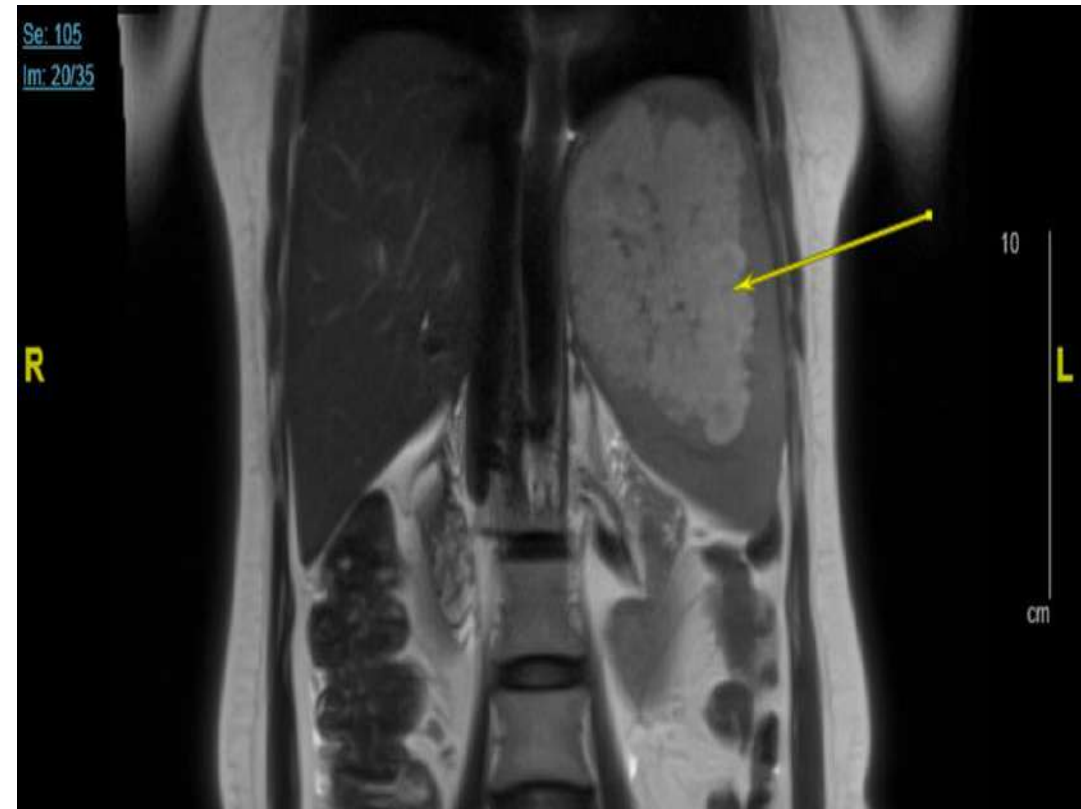
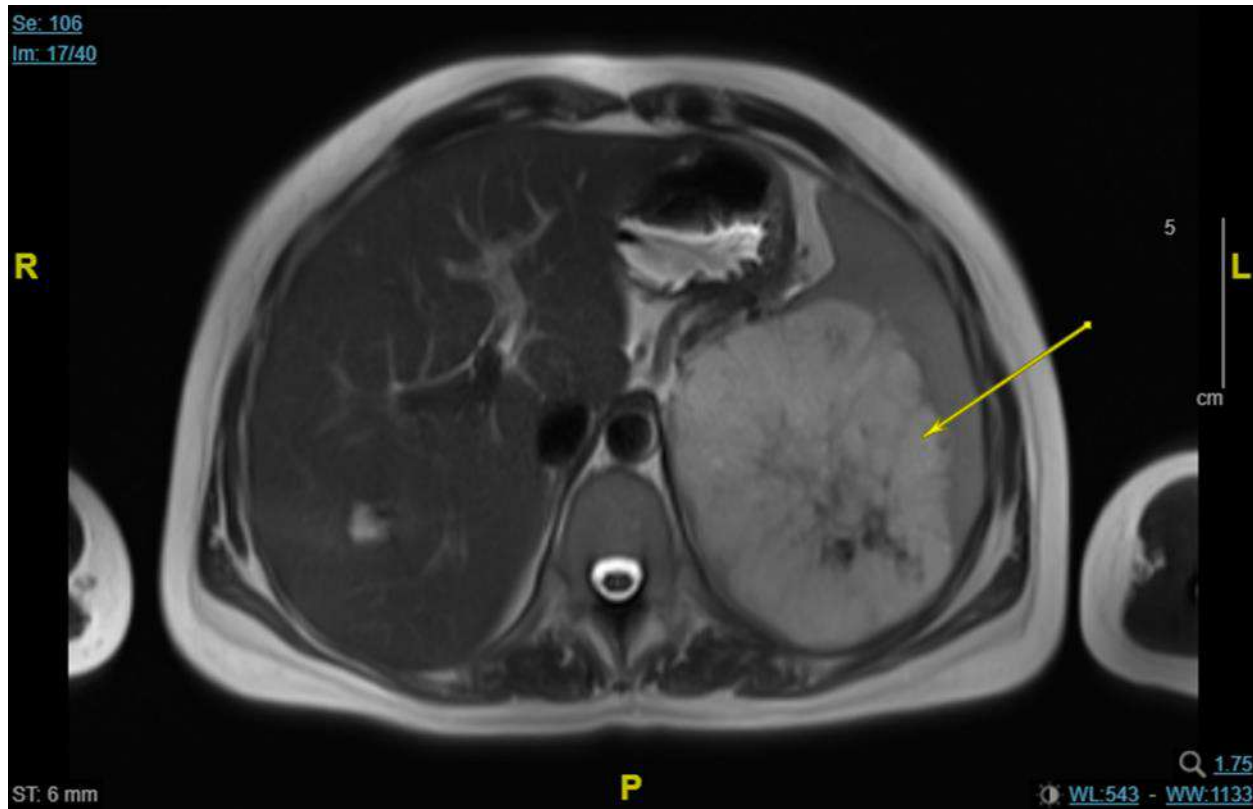
CASE HISTORY:

34- year - old female presented with postprandial bloating and abdominal distension, loss of weight and appetite.

Intra-operative findings: Large splenic hemangioma, occupying the upper 2/3rd of the spleen.

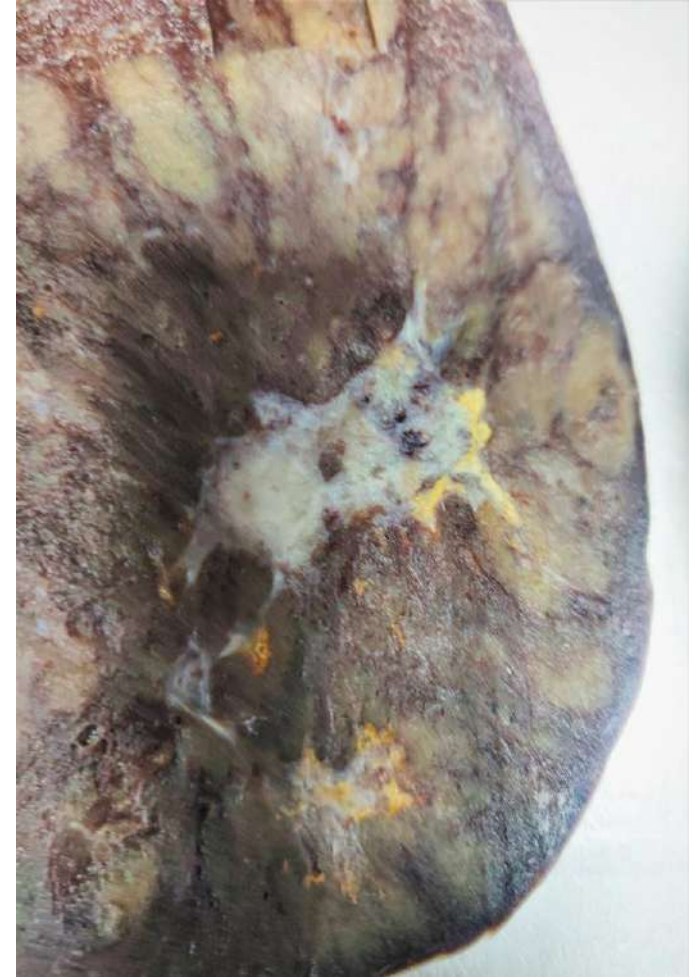
MRI:

Enlarged spleen with well-defined, large, T2 hyperintense, lobulated, heterogenous mass, 10.5 x 10 x 8.5 cm.

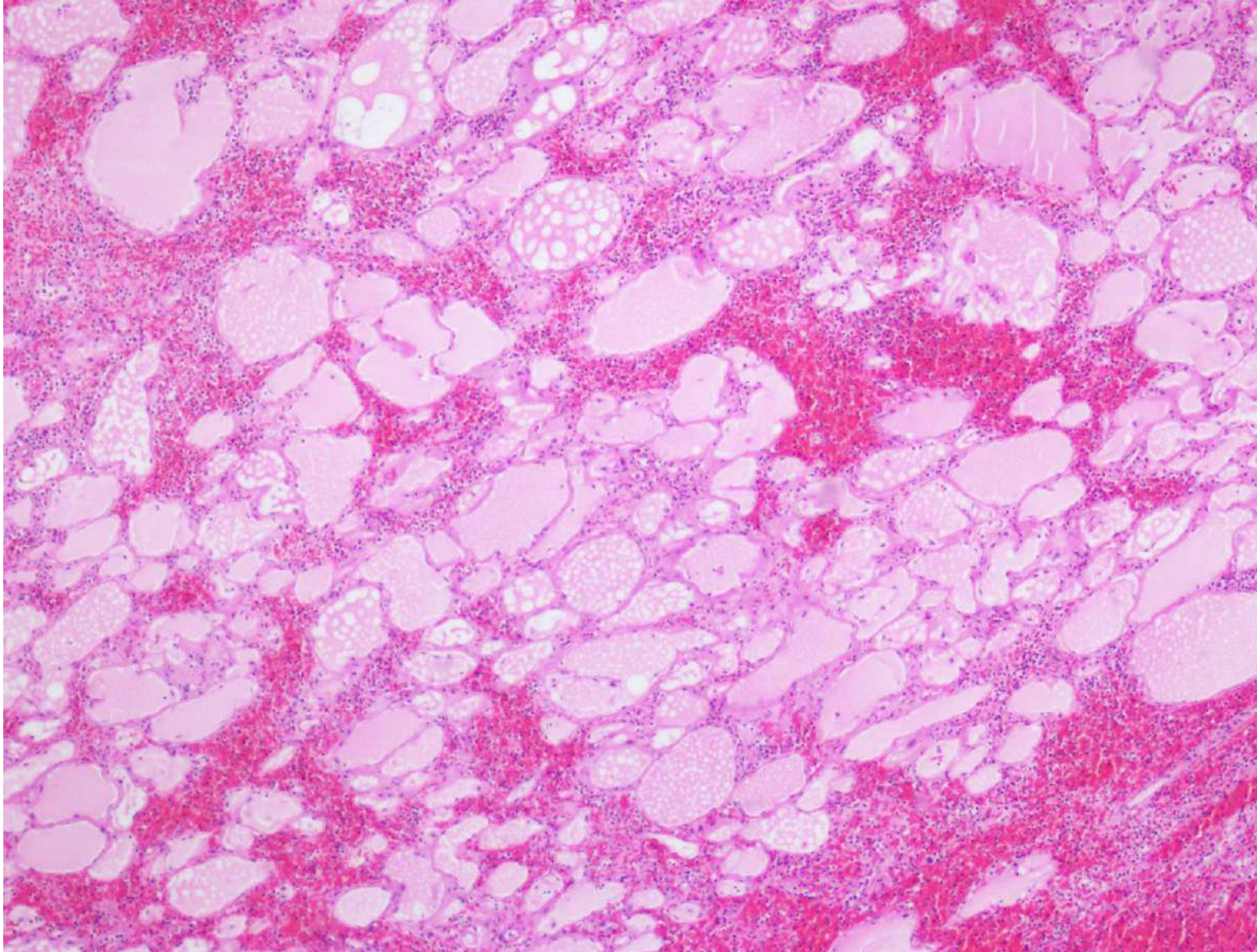


GROSS:

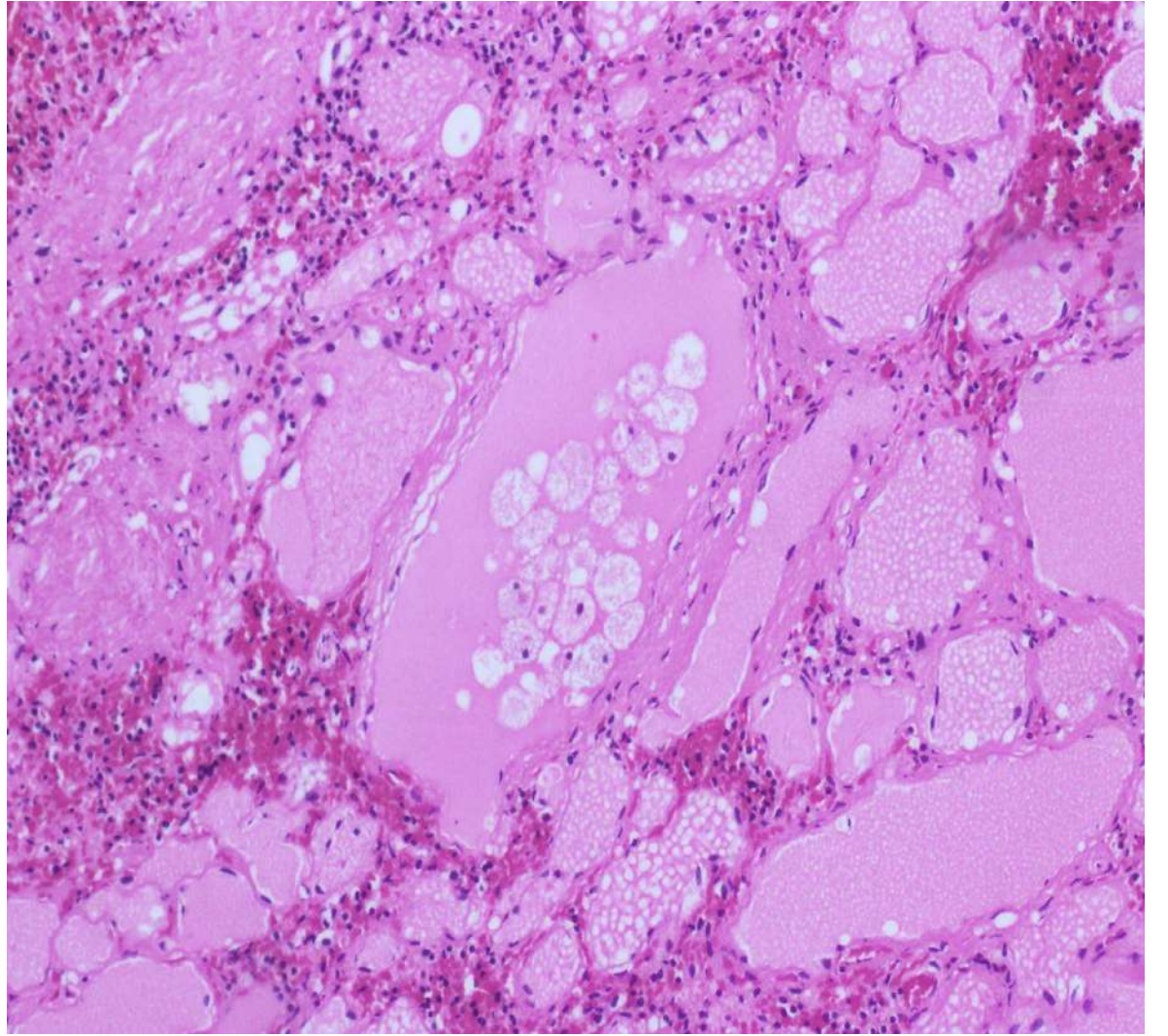
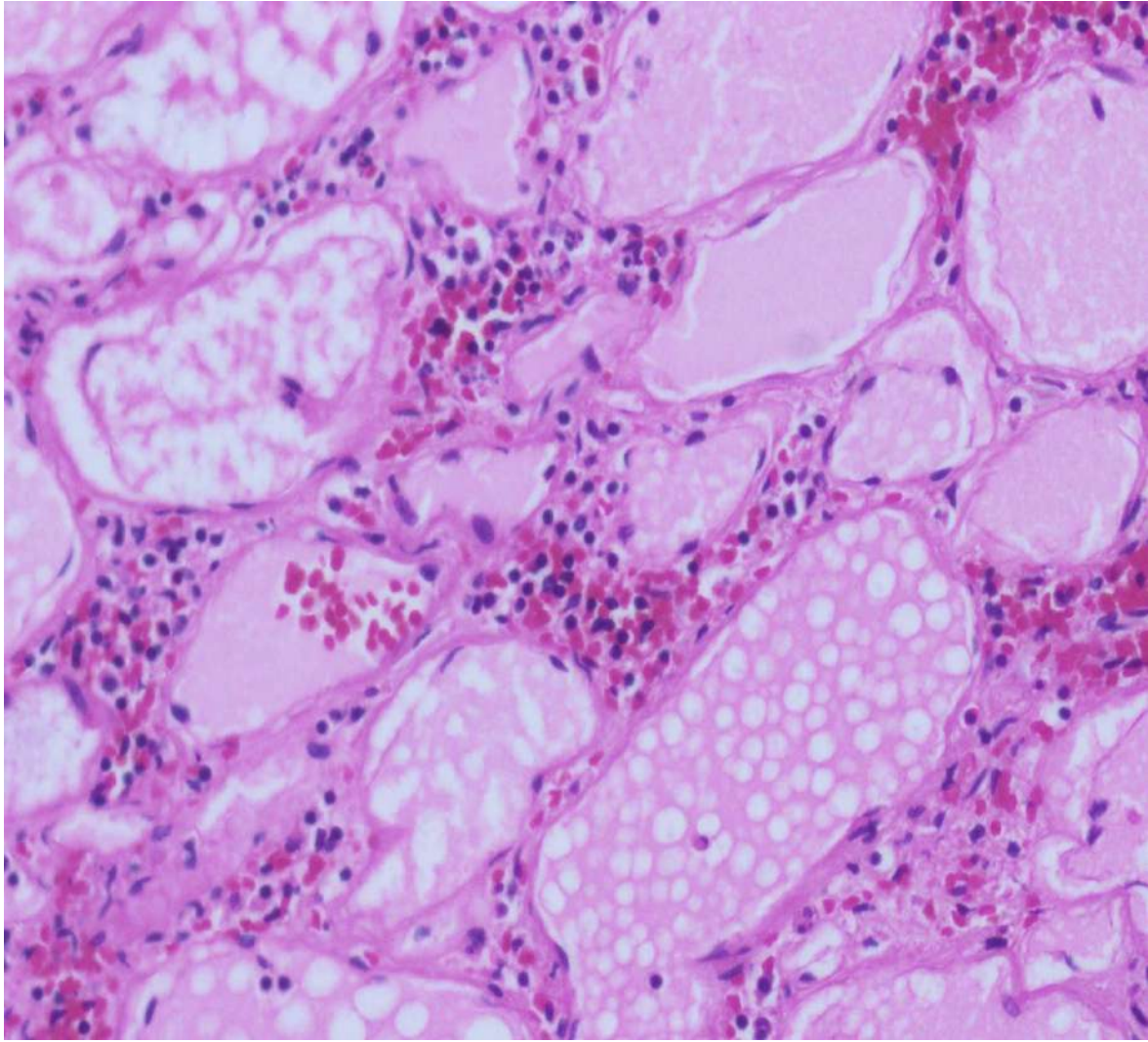
A well circumscribed lobulated tumour, 10.1x9x8cm with pale tan firm to spongy cut surface and central pale white fibrotic areas with focal yellow areas.

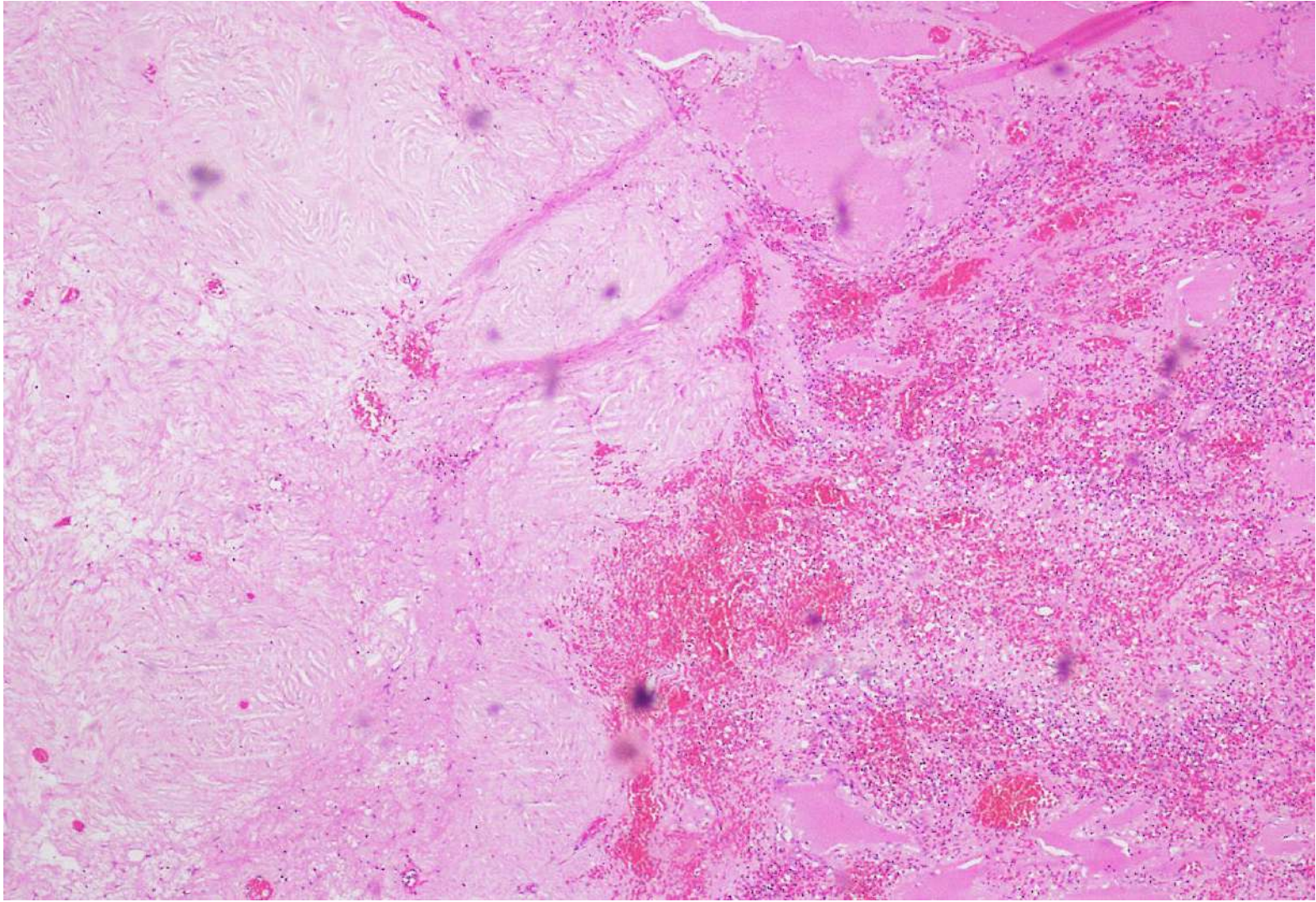


MICROSCOPY:



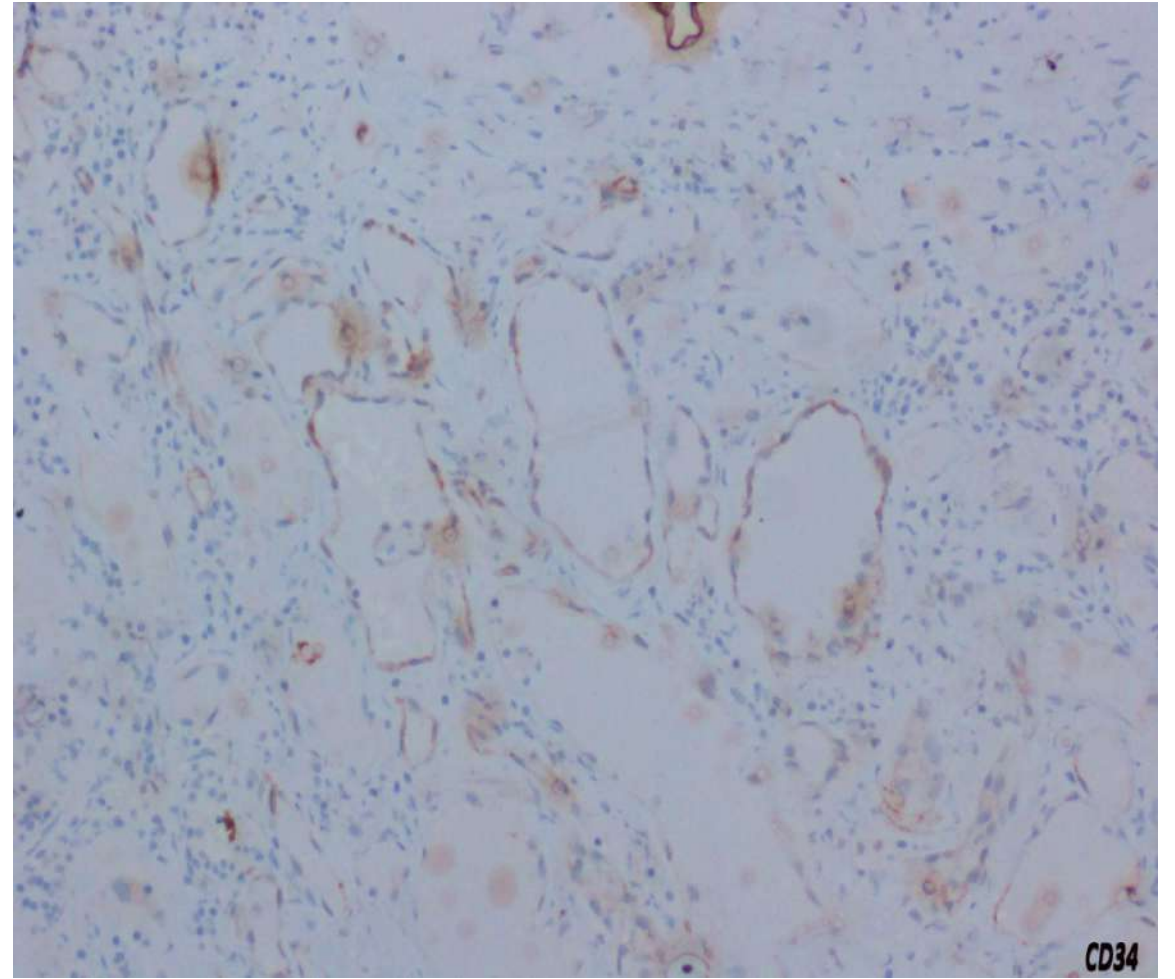
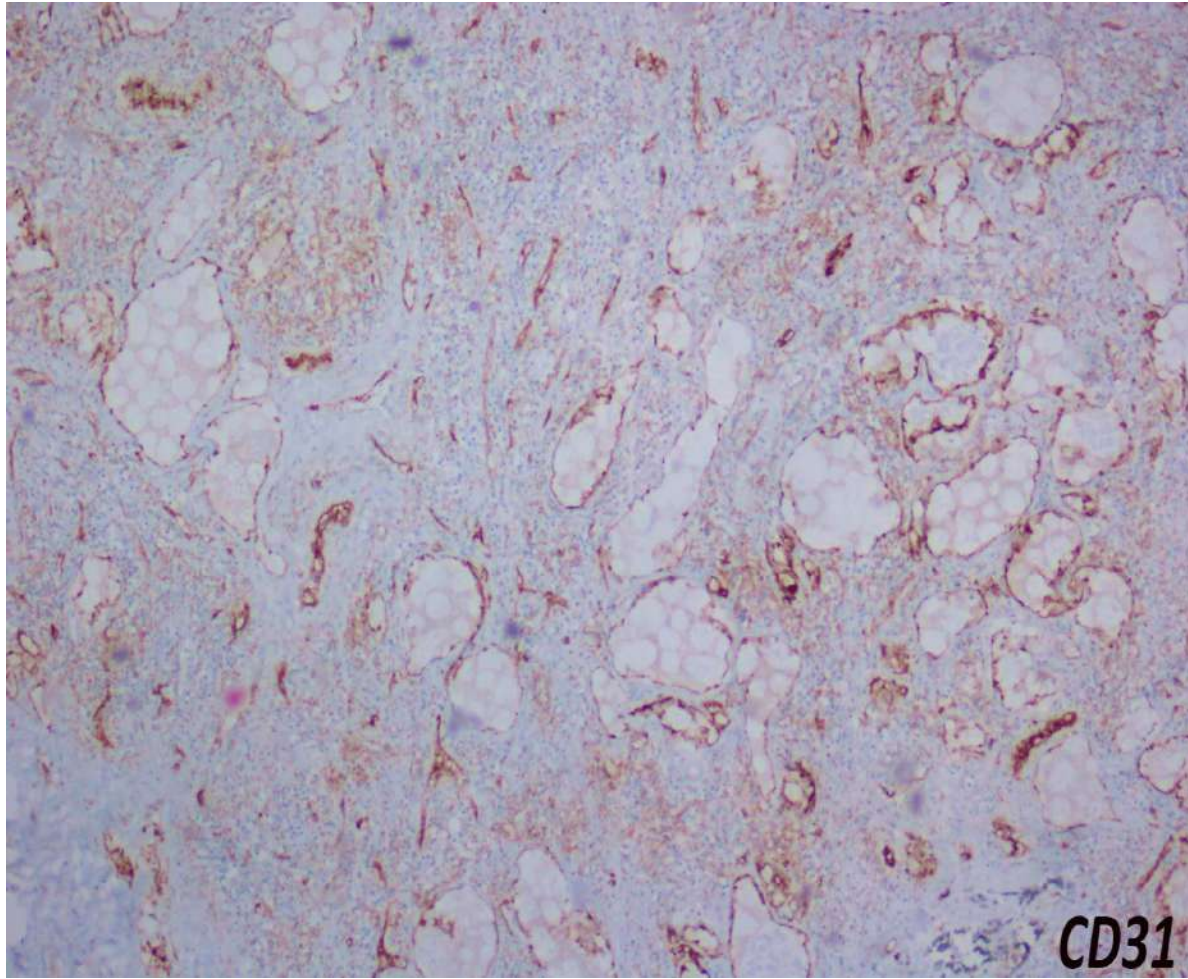
- *A lesion comprising proliferation of closely packed vascular channels of varying size.*
- *Lined by a single layer of bland endothelium*
- *Intraluminal amorphous pale eosinophilic proteinaceous material and aggregates of foamy macrophages.*
- *Central avascular fibrotic areas*

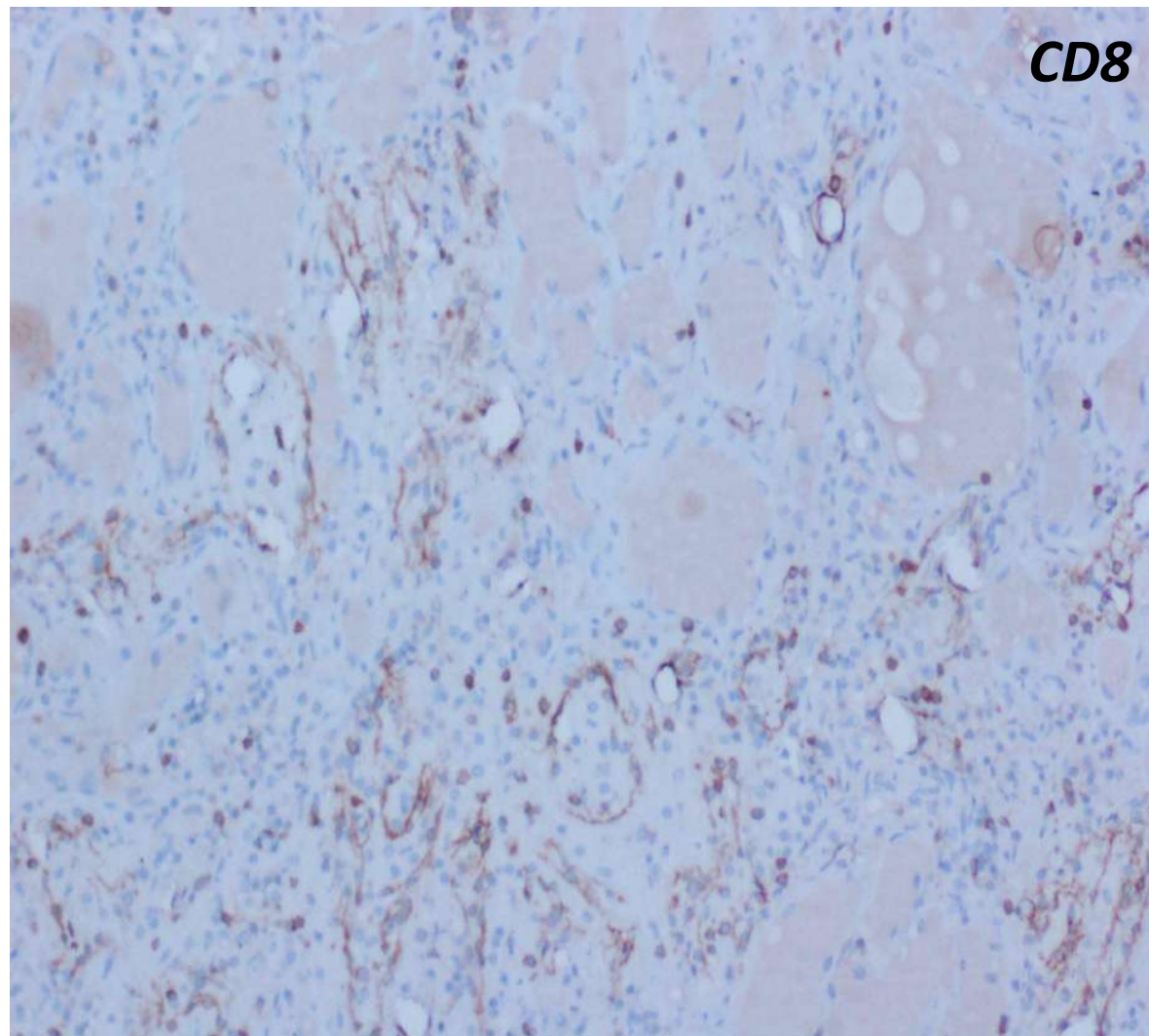
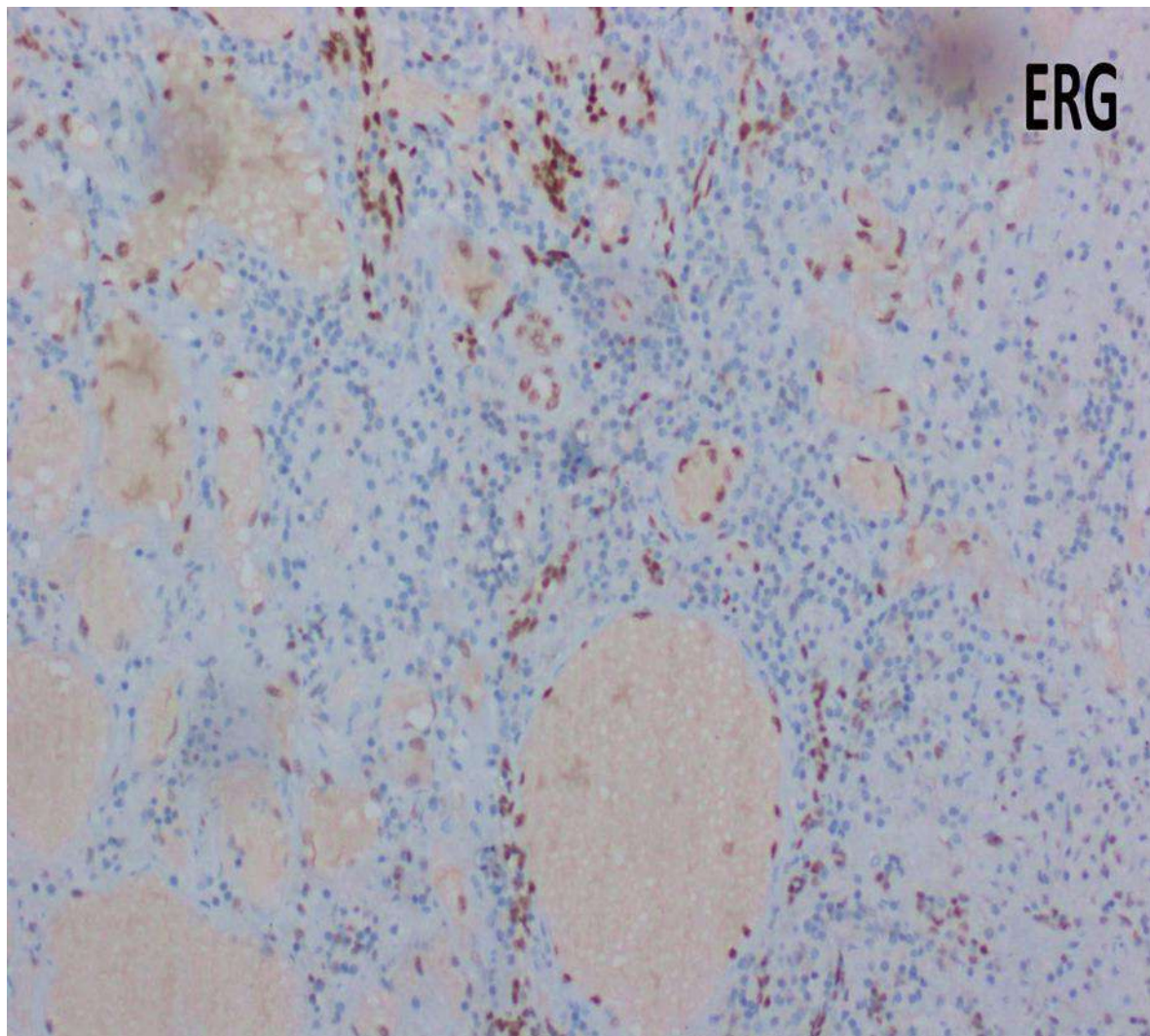




Central fibrotic stroma

IMMUNOHISTOCHEMISTRY:





Final diagnosis

- An unencapsulated lesion comprising proliferation of closely packed vascular channels of varying size, lined by a single layer of bland endothelium, positive for CD31(diffuse), D240(patchily) and focally for CD34, ERG(faint) and negative for CD8.
- Benign angiomatous neoplasm, consistent with lymphangioma, partial splenectomy.

Lymphangioma of spleen.

- Epidemiology: Extremely rare, usually an incidental finding, any age, often children and women.
- Cause: Abnormal congenital development, bleeding or inflammation of the lymphatic system may cause progressive dilatation of the lymphatic channels.
- Clinical features: Usually asymptomatic, if symptomatic, upper left quadrant pain is most common symptom, frequently followed by fever, nausea, vomiting, weight loss. Larger lesions are a/w bleeding, consumptive coagulopathy, hypersplenism, portal hypertension.
- The macroscopic appearance is heterogeneous, as lesions can be solid, cystic or even microcystic. When a single lymphangioma is present, this is generally subcapsular and sometimes surrounded by smaller satellite lesions. Sometimes lymphangiomas can involve the whole spleen, a condition regarded as diffuse lymphangiomatosis.
- Light microscopy: Subcapsular, multicystic vascular spaces lined by single layer of flattened endothelium lined spaces, filled with eosinophilic proteinaceous material, not red blood cells. Occasionally endothelium may form small papillary projections.
- IHC= Positive: CD31, D2-40, CD34(variable), ERG(variable) and CD8 negative.

Useful immunohistochemical markers in the diagnostic workup of vascular lesions of the spleen.

	Hemangioma	Lymphangioma	Littoral cell angioma	Hamartoma	Peliosis	Angiosarcoma
CD31	+	+	+	+	+	+
CD34	+	+/-	-	+/-	+	+/-
CD8	-	-	-	+	+	+/-
D2-40	-	+	-	-	-	-

REFERENCES:

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- ✓ Arber DA, Strickler JG, Chen YY, Weiss LM. Splenic vascular tumors: a histologic, immunophenotypic, and virologic study. *Am J Surg Pathol*. 1997;21(7):827-835.
- ✓ Dr. Christopher Fletcher's Diagnostic Histopathology of Tumors, 5th Edition.
- ✓ Vascular neoplasms and non-neoplastic vascular lesions of the spleen Valentina Fabiola Ilenia Sangiorgio a, Daniel A Arber.