



Case #20

NAME Educational Activities Committee



Case courtesy of Dr. Kanayo Tatsumi (St. Louis City ME Office) and written by Dr. Nicolas Kostecky (Washington University, St. Louis)

1. The decedent was a 62-year-old African-American male involved in a motor vehicle collision. Minimal trauma was noted on external examination. At autopsy, 6000 mL of ascitic fluid was extracted, the liver was cirrhotic, and extensive hypertensive and atherosclerotic disease was documented. The decedent's spleen weighed 360 grams and had the following appearance.

What is the name of this finding (free text answer)?

spleen sugar spleen perisplenitis Sugar icing spleen Frosted
Sugar coating Sugar-coating
sugar coated spleen fibrosis Perisplenitis
Hyaline Hyaloseritis capsular fibrosis
Hyaline perisplenitis Chronic
Sugar spleen hyalinosis sugar-coated spleen
Splenic Icing sugar spleen Splenic capsular Zuckerguss

2. What is the most likely etiology of this finding?

- Sickle cell disease
- Portal hypertension
- Splenic infarct
- Normal anatomic variant
- Hematolymphoid malignancy

2. What is the most likely etiology?

A: Sickle-cell disease (13.31% responses)

Sickle cell trait may lead to splenic complications similar to those experienced by individuals with sickle cell disease, albeit typically at high altitudes with or without physical exertion. This would include splenic sequestration, leading to splenomegaly, or splenic infarction. It does not typically cause fibrosis of the capsule to the degree observed in the image.

B: Portal hypertension (CORRECT ANSWER, 47.4% responses)

Hyaline perisplenitis/hyaloserositis of the spleen is a common and incidental finding at autopsy. Colloquially referred to as “sugar-coated spleen” or “icing-sugar spleen”, it results from the deposition of collagen on the capsular surface of the spleen, which may occur secondarily to inflammation of the peritoneal cavity. It is often incidental but is found in association with cirrhosis and Fitz-Hugh-Curtis syndrome.

C: Splenic infarct (4.87% responses)

Splenic infarcts are typically subcapsular and wedge-shaped.

D: Normal anatomic variant (27.92% responses)

Though hyaline perisplenitis/hyaloserositis is a common and incidental finding at autopsy, it is not normal. The most likely cause in this patient is cirrhosis and portal hypertension.

E. Hematolymphoid malignancy (6.49% responses)

Splenic involvement by a hematolymphoid malignancy typically presents with an enlarged spleen, often with nodularity or an appreciable mass on sectioning.

REFERENCES

- 1) Goodman J, Hassell K, Irwin D, Witkowski EH, Nuss R. The splenic syndrome in individuals with sickle cell trait. High Alt Med Biol. 2014 Dec;15(4):468-71. doi: 10.1089/ham.2014.1034. PMID: 25361178; PMCID: PMC4273194.
- 2) Swami SY, Bhure AA, Narwade SB, & Valand AG. Icing sugar spleen/perisplenitis cartilaginea: a case report. J Cytol Histol. 2018. 9:1
- 3) Wick M, LiVolsi V, Pfeifer J, Stelow E, & Wakely P Jr. (Eds.). (2015). Silverberg's Principles and Practice of Surgical Pathology and Cytopathology (5th ed.), vol. 2. Cambridge: Cambridge University Press. doi:10.1017/9781139137201
- 4) Sens MA, and Hughes R. (2021). Diagnostic Pathology Forensic Autopsy. Philadelphia USA: Elsevier.