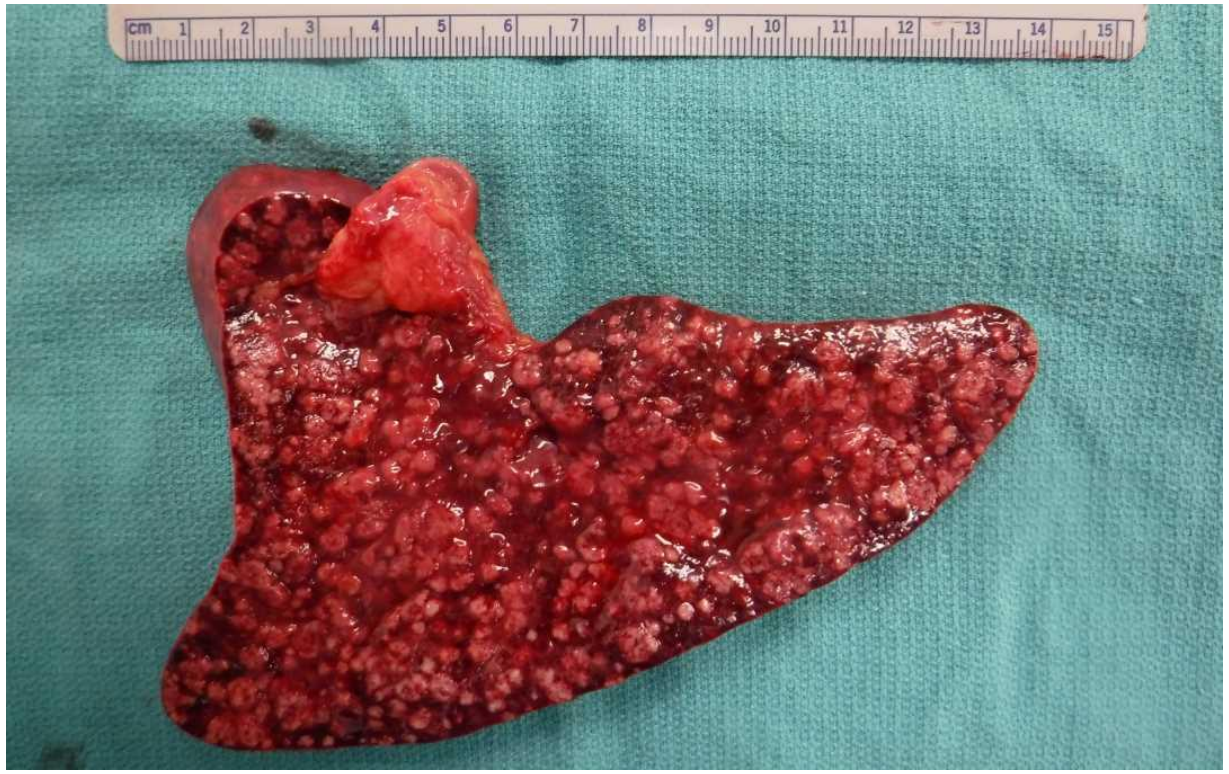
Case #32

NAME Educational Activities Committee

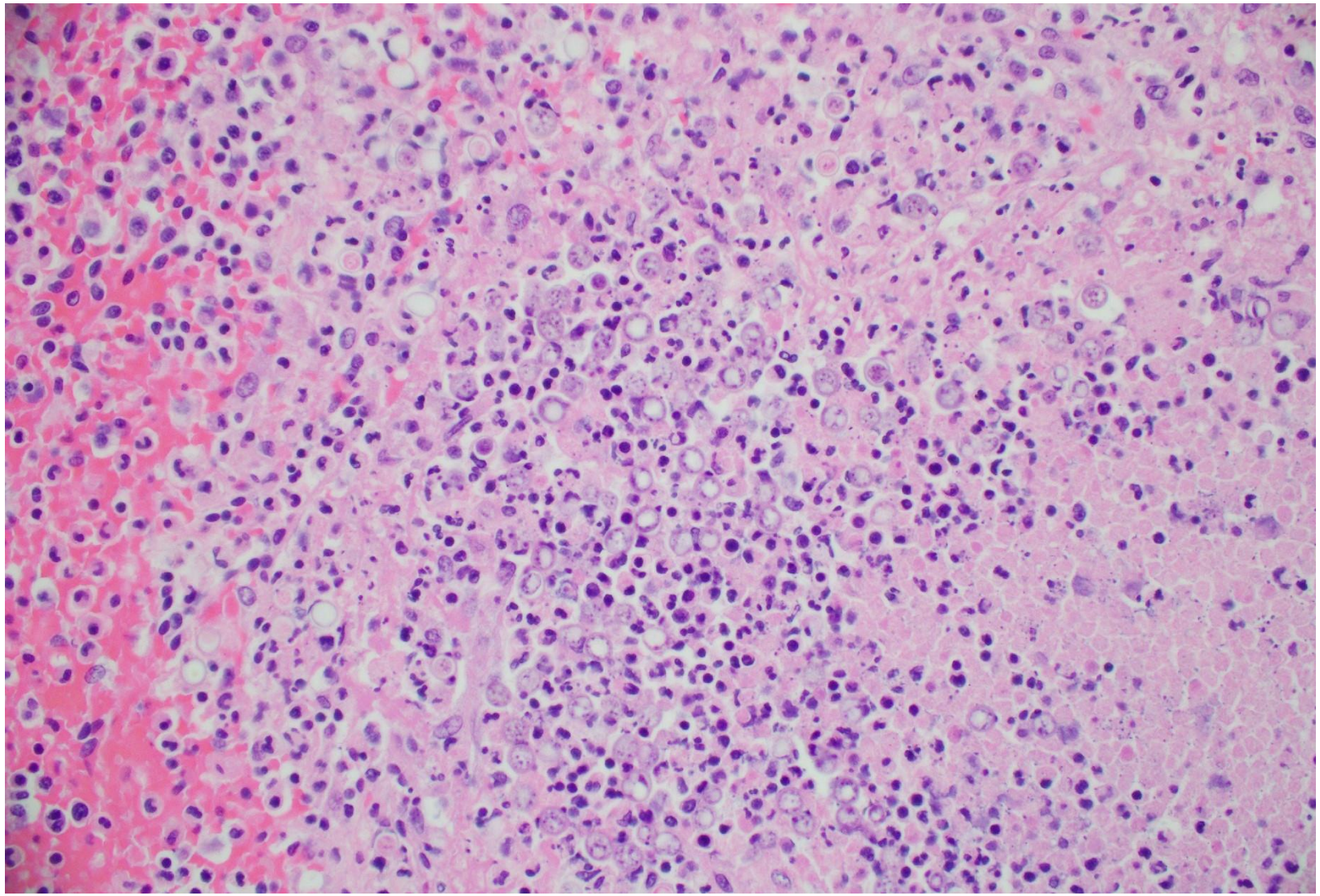
Case provided by:

Dr. Leo Yenwongfai (PGY 2)
Dr. Greg Davis (Professor)
Dr. Nathan Shelman (Assistant Professor)
Dr. Alexander Fenwick (Assistant Professor)

University of Kentucky College of Medicine.



Gross images, spleen



Microscopic image, spleen (H&E stain, 200x)

1. The images above are from a 52-year-old male with non-resolving pneumonia and marked splenomegaly. What is the most likely diagnosis?

- ◯ Primary splenic diffuse large B cell lymphoma
- ◯ Fungal infection
- ◯ Splenic marginal zone lymphoma
- ◯ Sarcoidosis
- ◯ Hyaline perisplenitis

Answer...

B. Fungal infection (Blastomycosis)

The decedent was a 52-year-old male in Kentucky with coal workers' pneumoconiosis hospitalized for non-resolving pneumonia after initial treatment and discharge for community-acquired pneumonia. Upon admission, he was septic with bilateral pulmonary lung consolidations. Despite appropriate antibiotic treatment, he remained in septic shock, and bronchoscopy demonstrated bilateral alveolar hemorrhage. He developed diffuse papulovesicular skin lesions with central ulceration and hemorrhage. Results received from pre-mortem fungal studies were positive for Blastomyces antigen (results finalized after the patient was deceased).

The gross autopsy findings confirmed the diffuse papulovesicular skin lesions with central ulceration and hemorrhage, bilateral cavitary lung lesions with diffuse consolidations, splenomegaly with congestion and diffuse granulomas, mild cardiomegaly and hepatomegaly. Microscopically, yeast forms with morphology consistent with Blastomyces were identified in the lungs, spleen, skin, heart, esophagus, larynx, liver, pancreas, kidneys, prostate, testes, spleen, and bone marrow. Fungal cultures isolated Blastomyces dermatitidis.

Cause of Death: Sepsis due to severe disseminated blastomycosis dermatitidis complicating underlying coal workers pneumoconiosis

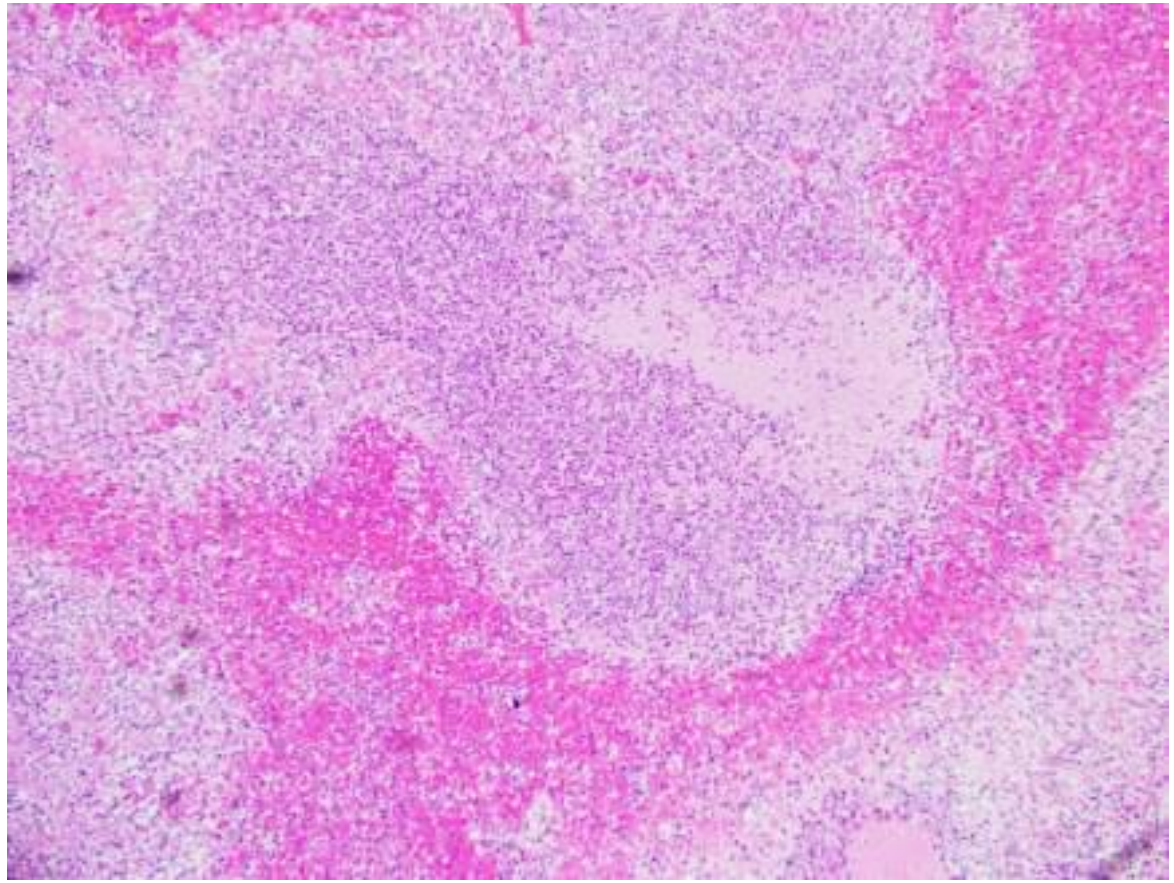
A. Primary splenic diffuse large B cell lymphoma (14.9% responses)
Mature B cell neoplasm usually presents in adults over 60 years as single or multiple distinct splenic nodules. Microscopy shows sheets of large hematolymphoid cells with variable cytologic features. Necrosis is identified in these nodules interspaced within normal splenic parenchyma. These cells are usually CD20, CD 19, and PAX-5 positive, confirming the B cell lineage.

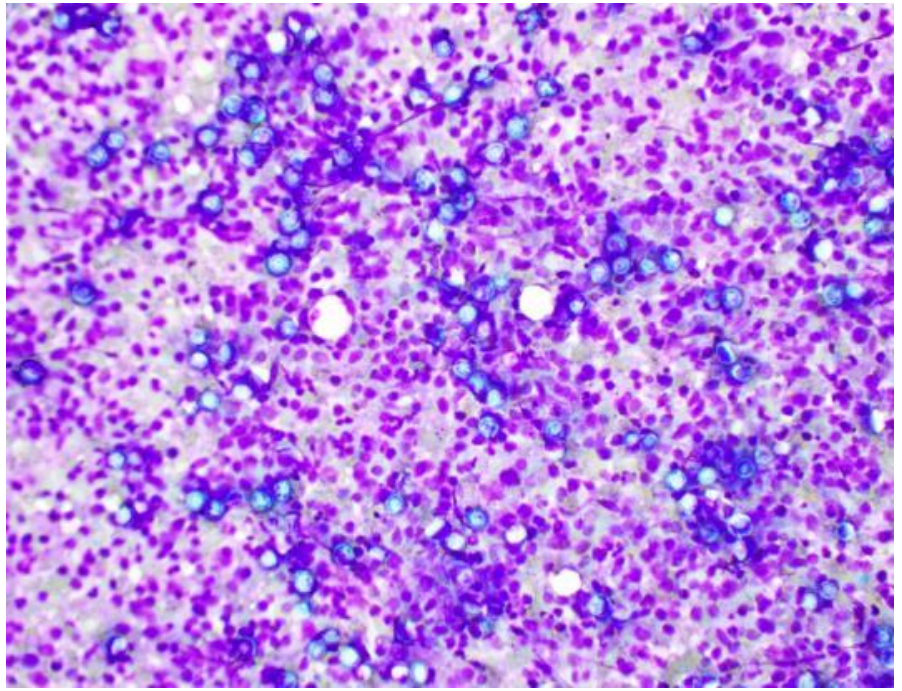
B. Fungal infection (CORRECT ANSWER, 67.96% responses)
Blastomyces is endemic in southeastern, central, and midwestern USA, with the lung, skin, and bone being the most common sites of infection with possible widespread dissemination. Immunocompromised patients typically have a worse prognosis. Microscopically, yeast forms are round (8-15 microns) with a thick wall and typically show broad-based budding. Yeast forms with similar morphologies were identified in this case, as demonstrated by the touch preparations and H&E. Moreover, these yeast forms typically cause necrotizing granulomas, compatible with similar lesions identified in the spleen and lungs in this case.

C. Splenic marginal zone lymphoma (6.53% responses)
Mature B cell neoplasm predominantly affecting the white pulp of the spleen, resulting in white pulp expansion by small B lymphocytes typically showing a biphasic appearance of the spleen. The leukemic phase typically shows lymphoid cells with polar villous projections. The typical immunophenotype for splenic marginal zone lymphoma includes CD 19, CD 20, CD 79 (a), CD 79 (b), and rarely CD 103.

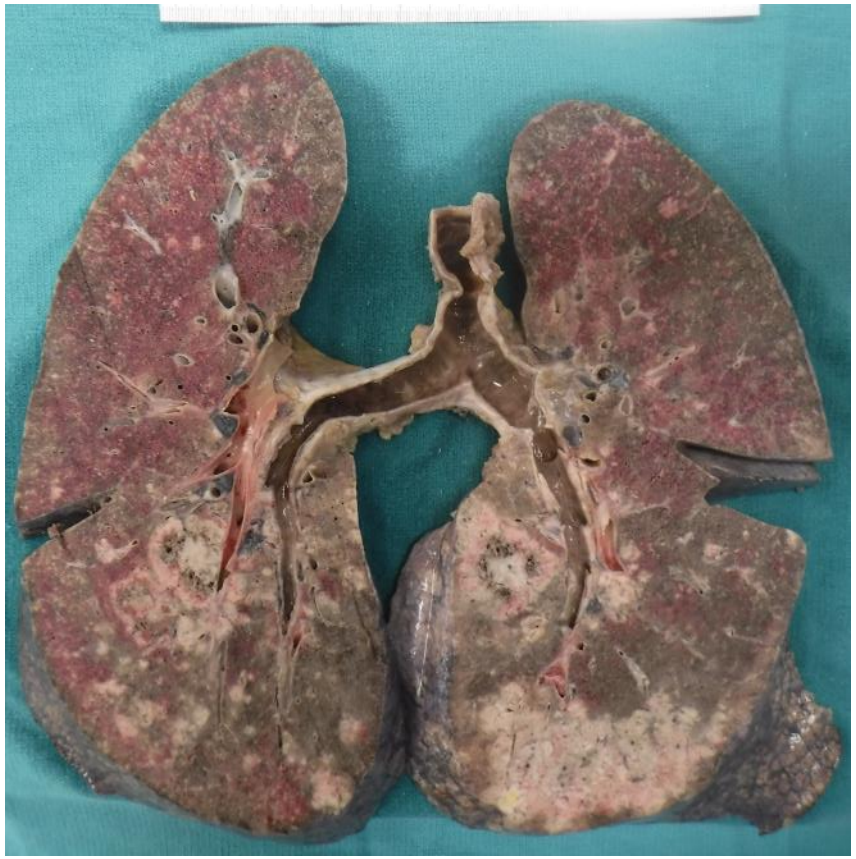
D. Sarcoidosis (7.76% responses)
Sarcoidosis commonly involves hilar lymph nodes, and can show diffuse involvement with nonnecrotizing granulomas, and can certainly involve the spleen. Splenic sarcoidosis can progress to splenic infarction or rupture. Diagnosis of sarcoidosis is based upon the presence of characteristic nonnecrotizing granulomas in combination with clinical presentation, laboratory studies, and imaging features. This case depicts necrotizing granulomas with yeast forms.

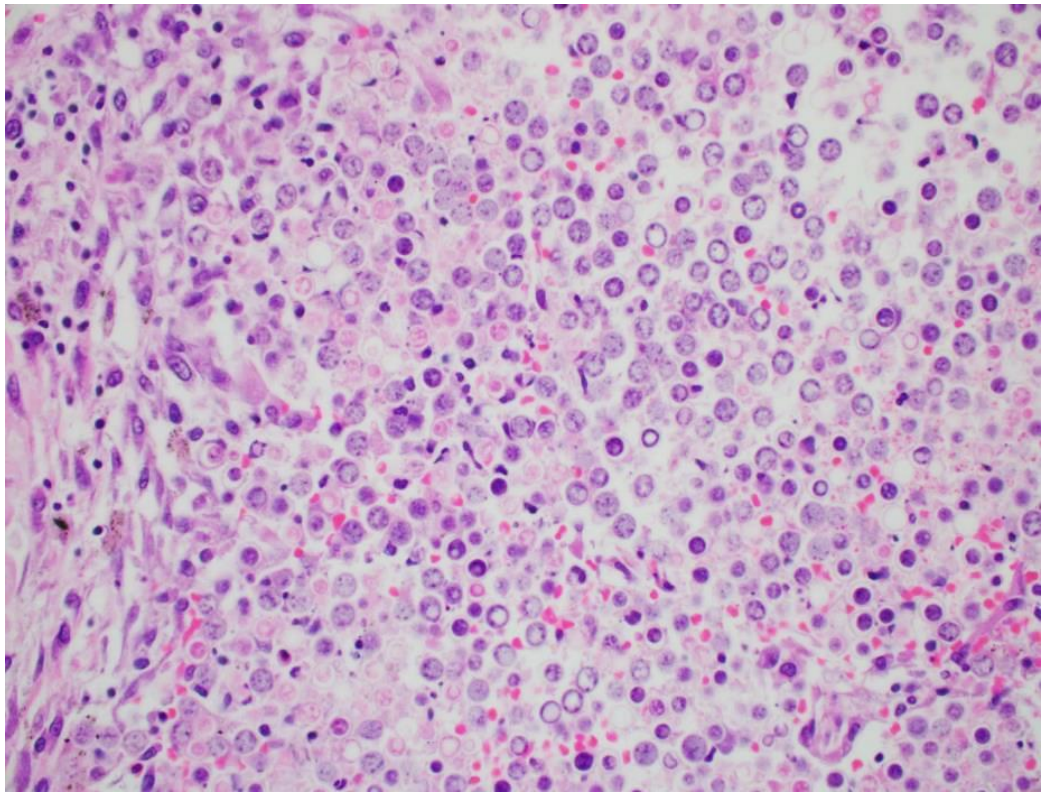
E. Hyaline Perisplenitis (2.86% responses)
Hyaline perisplenitis is often characterized by collagenous splenic nodules with a conspicuously thickened and similarly collagenous capsule. This process is simply the result of chronic and repeated episodes of splenic capsular inflammation. Some syndromic association has been reported, but it is not an uncommon incidental finding at autopsy.



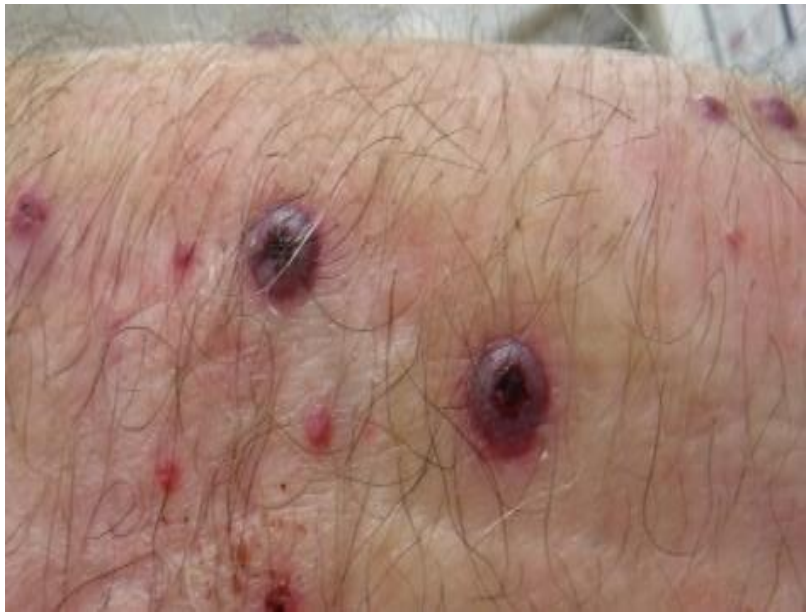


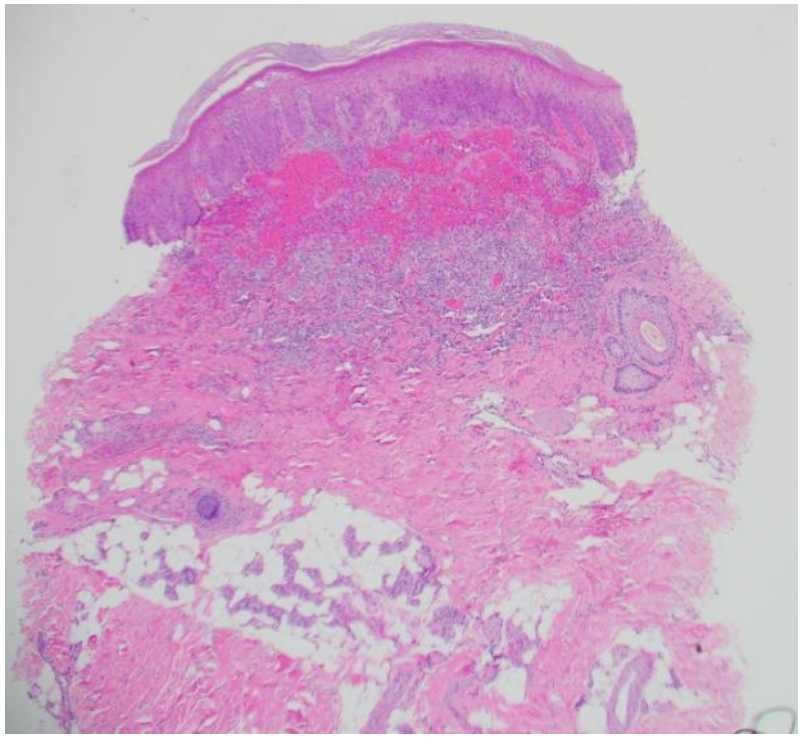
Additional spleen images (low power H&E and touch prep)

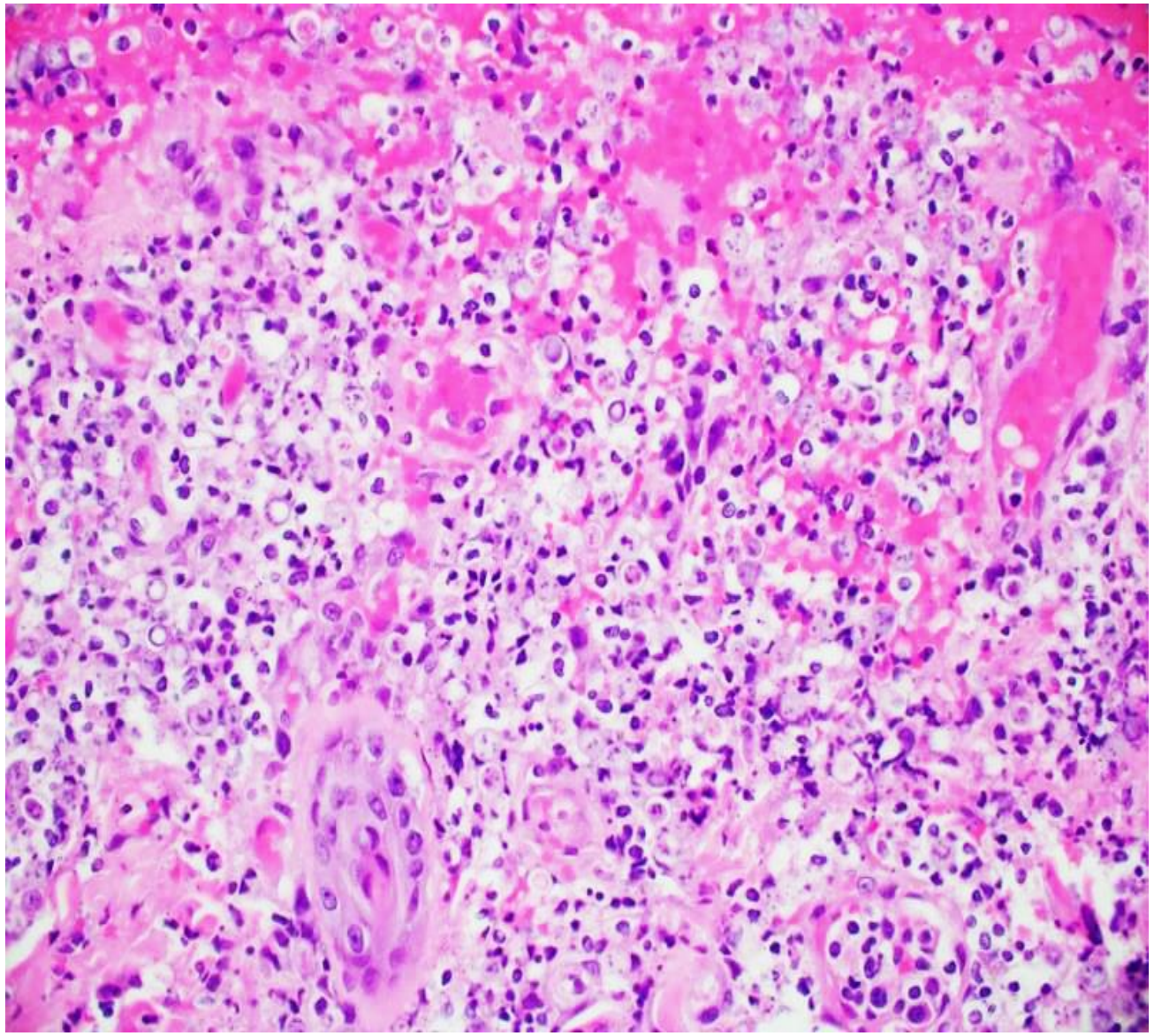




Lung lesions







Skin lesions

References:

- Wani NA, Parray FQ. Primary lymphoma of the spleen: an experience with seven patients. Int
- T. Rinda Soong, Dan A Milner. https://app.expertpath.com/document/blastomycosis/c57d878f-a543-419b-b589-47888cc3967f?searchTerm=blastomycosis%20der
- https://app.expertpath.com/document/splenic-marginal-zone-lymphoma/1d576cb1-f9d4-45f2-9978-8474846784b7?searchTerm=splenic%20marginal%20zone%20lymphoma
- Swami SY et al: Icing sugar spleen/perisplenitis cartilaginea: a case report. J Cytol Histol. 9:1, 2018
- Judson MA: Extrapulmonary sarcoidosis. Semin Respir Crit Care Med. 28(1):83-101, 2007