A Pediatric Case Report: Necrotizing Sarcoid Granulomatosis

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OBJECTIVES

- Recognize an uncommon clinical presentation of necrotizing sarcoid granulomatosis (NSG) in a pediatric patient
- Distinguish NSG from other granulomatous disease processes
- Outline the diagnosis, treatment, and management of NSG

INTRODUCTION

- NSG is a rare systemic disease characterized by confluent sarcoid-like granulomas with extensive necrosis and vasculitis, primarily affecting the lungs
- Since first description in 1973, etiology and classification of NSG remains unclear
- Rarely described in pediatrics. Median age 42 years. F>M.
- Symptoms non-specific: cough, dyspnea, chest pain, constitution symptoms include +fever, +fatigue, +night sweats

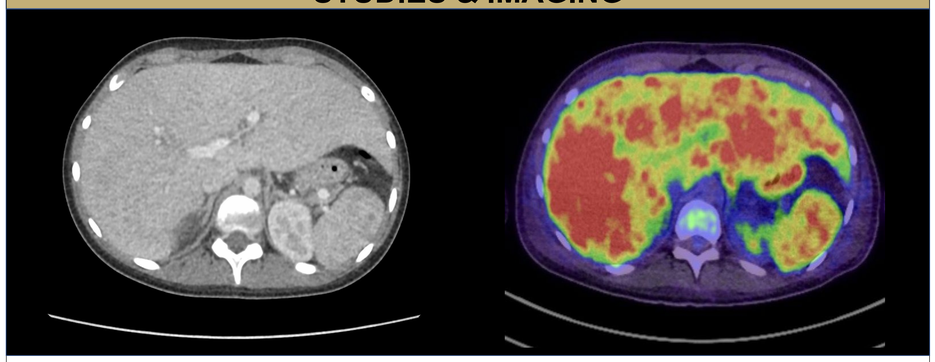
CASE PRESENTATION

- 17 yo female presents to ED with one week of daily low-grade fever, diarrhea, generalized abdominal pain, myalgia, fatigue, and 2x NBNB emesis
- PMH: One year of intermittent abdominal pain of unknown etiology. Previous workup only significant for hepatomegaly and enlarged abdominal LN
- In ED, patient experienced acute decompensation with fluid-refractory shock of unclear etiology requiring norepinephrine and steroids

HOSPITAL COURSE

- Multiple labs and imaging as workup significant for highly elevated inflammatory markers, mild transaminitis, and mild AKI
- Infectious disease, rheumatology, hematology, ophthalmology, and oncology services consulted
- Continued to have fevers on antibiotics: ceftriaxone, doxycycline, metronidazole, vancomycin
- After extensive infectious workup, no infectious source identified

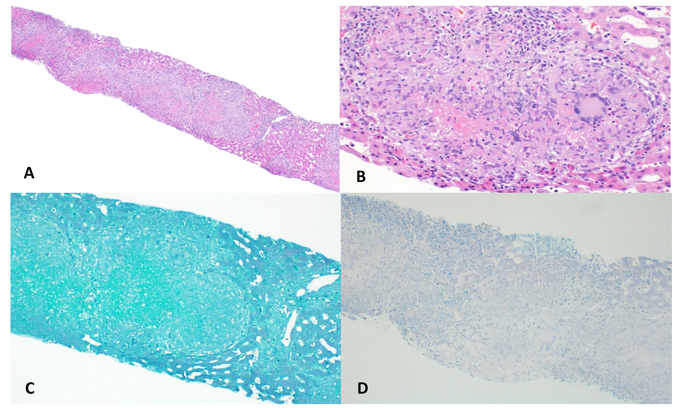
STUDIES & IMAGING



- **CT AP w/ contrast:** hepatomegaly with periportal edema, innumerable hypoattenuating splenic lesions, bilateral pleural effusions, and prominent intra-abdominal lymph nodes without pulmonary lesions
- **PET:** increased radiotracer uptake in the liver and spleen with increased number of scattered sub-centimeter lymph nodes in the mesentery and retroperitoneum

Selected references:
1. Churg, C. B. Carrington, and R. Gupta, "Necrotizing Sarcoid Granulomatosis," *Chest*, vol. 76, no. 4, pp. 406–413, Oct. 1979, doi: 10.1378/CHEST.76.4.406.
2. Y. Rosen, "Four Decades of Necrotizing Sarcoid Granulomatosis: What Do We Know Now?," *Arch Pathol Lab Med*, vol. 139, no. 2, pp. 252–262, Feb. 2015, doi: 10.5858/arpa.2014-0051-RA.

PATHOLOGY



A: Numerous confluent necrotizing granulomas, low power
B: Necrotizing granuloma, central areas of necrosis
C & **D:** Special AFB stain, Fites AFA stain, negative for organisms

DISCUSSION

- Varied clinical & radiologic features, thus important to obtain pathologic data to examine histologic features
- Can imitate various disease processes: granulomatous infection, autoimmune, oncologic processes, which must be excluded before diagnosis
- Treatment with steroids, however unclear duration

CONCLUSION

- Uncommon presentation of NSG without pulmonary involvement
- Rule out other disease processes prior to starting tx
- Patient was stable on prednisone, continues to follow-up with outpatient rheumatology