Learning Objectives

- Pleomorphic Undifferentiated Sarcoma (PUS) is a rare cancer typically found in the soft tissues of the extremities or trunk.
- PUS of the spleen is very rare, with only one case occurring in someone under the age of 40 in the literature.

HISTORY OF PRESENT ILLNESS

A 35 year old Caucasian woman with travel to the Middle East 6 months ago presents with fever and diffuse myalgias. Patient initially had cough after returning from abroad. CXR obtained following failed antibiotic therapy showed LUQ calcified structure, prompting further imaging. CT and MRI revealed several splenic lesions concerning for Echinococcus. Despite negative Echinococcal titers, patient was treated empirically with albendazole per ID without improvement.

PHYSICAL EXAM

Patient had a temperature of 38.2°C and a heart rate of 130. Patient appeared pale and anxious. Abdominal exam remarkable for LUQ tenderness and hepatosplenomegaly. The rest of the exam was unremarkable.

LABORATORY DATA

- WBC 17 and Hgb 6.6
- BMP and AST/ALT were normal
- Alk phos was 350 (normal <150)
- Lactic acid 1.2

Figure 1: Coronal and sagittal views of CT Abdomen demonstrating large calcified splenic cyst (9.2cm) surrounded by smaller cysts.

Repeat CT confirmed presence of enlarging cystic lesions in the spleen. Due to profound anemia and concern for intraparenchymal hemorrhage, surgery was consulted and splenectomy recommended. Post-splenectomy, she had rapid clinical improvement. Preliminary pathology revealed sarcoma without infection. After discharge, analysis of the tumor by two independent pathologists failed to show evidence of differentiation and patient was diagnosed with PUS. An outpatient PET scan showed LUQ hypermetabolic activity and patient was started on chemotherapy.

Teaching Points

- Formerly known as malignant fibrous histiocytoma, PUS most commonly affects people in the 6th and 7th decade of life. High grade pleomorphic sarcomas have an overall 5-year survival rate of 50 – 60% and age, subtype and staging of the sarcoma are among the most important prognostic factors.1-2
- Macroscopically, these tumors are heterogeneous, well-circumscribed masses with most measuring 5 – 15cm in diameter.3
- The diagnosis is one of exclusion. These undifferentiated tumors do not express antigens suggestive of other types of sarcomas on immunohistochemistry.4
- Treatment consists of surgery +/- chemotherapy or radiation, but the best treatment has yet to be elucidated.

References