Primary undifferentiated pleomorphic sarcoma of spleen - a rare localization and review of literature

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Introduction:

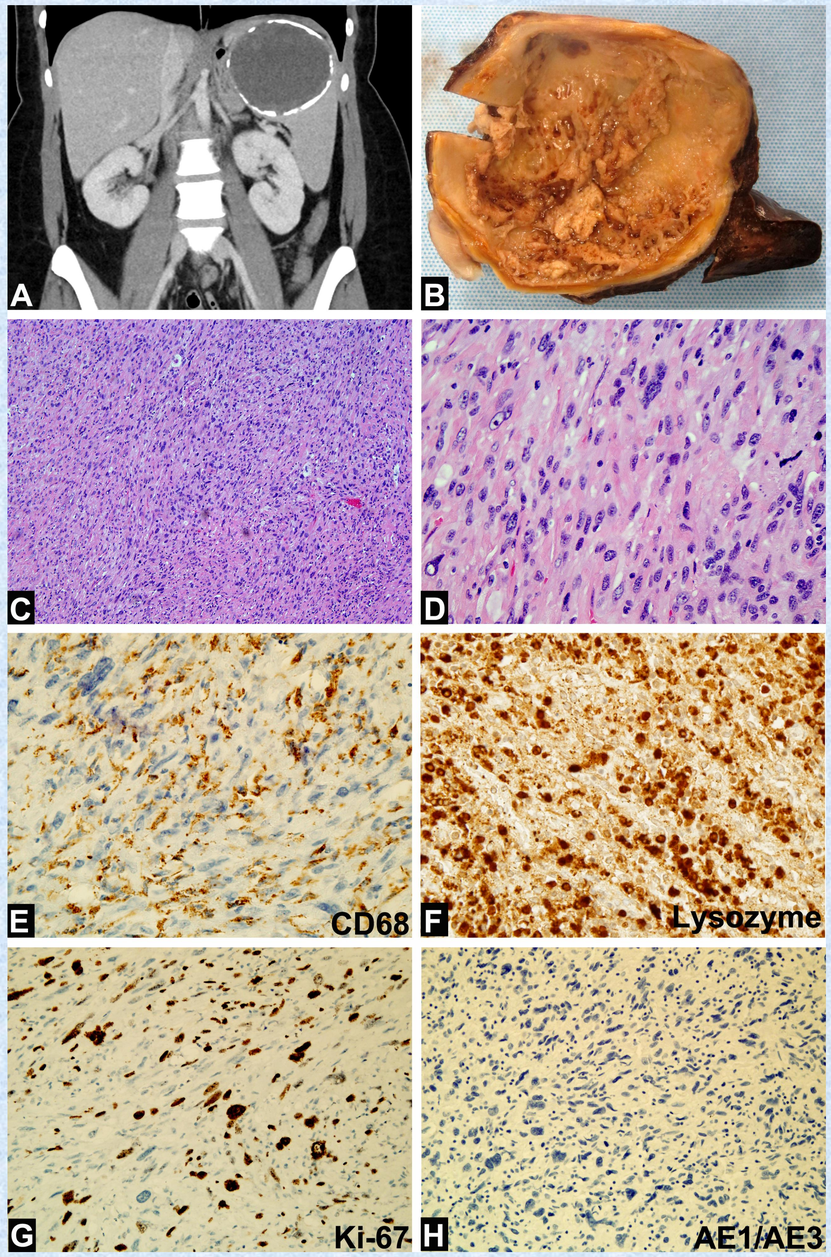
Undifferentiated pleomorphic sarcoma (UPS), formerly known as pleomorphic malignant fibrous histiocytoma, is now designated as a tumor showing no epithelial, melanotic, or lymphoid differentiation, and no definitive mesenchymal differentiation. It is typically found in soft tissues of the extremities or trunk. Primary UPS of the spleen is extremely rare.

Case Features:

The patient is 35-year-old female with a history significant for hypothyroidism, depression, asthma, and a presumed diagnosis of echinococcal infection in February 2014. CT scan (**A**) identified large calcified cysts in the enlarged spleen (16.7 cm) and hepatomegaly (22.5 cm). A diagnosis of Echinococcus was made per the imaging and her extensive travel history including Laos, Nigeria, Egypt. She presented to her PCP in April for fever, cough and abdominal pain, and again in May for fever. The patient reported night sweats, red/orange urine, 20 lb weight loss over 3 months. No lymphadenopathy was found. Splenectomy was started laparoscopically and converted to an open midline laparotomy due to the large size of the cysts and dense adhesions to the upper aspect of the spleen.

Gross Finding:

The specimen is a 980 gram, 16.0 x 15.0 x 9.0 cm spleen with a large 15.0 x 9.0 x 8.0 cm cystic mass. There are several 1.2 to 2.0 cm lobulated small masses protruding from the large cystic mass. The cystic cavity is surrounded by a thick wall with peripheral calcifications. The cystic cavity contains cloudy, yellow fluid with suspended soft necrotic debris. The remaining splenic parenchyma is pink to red, firm, and the splenic capsule appears intact. No hilar lymph nodes are identified (**B**).



Pathology and Ancillar Tests:

Histopathological examination revealed high-grade malignancy with a mixture of spindled and marked pleomorphic large cells (**C-D**), and focal necrosis associated with cystic degeneration. By immunohistochemistry, the neoplastic cells were positive for histiocytic markers including CD68 (**E**), CD163, and lysozyme (**F**), but did not show other lineage differentiation. Twenty percent of the noeplastic cells are postive for Ki-67 (**G**). Tumor cells are negative for AE1/AE3 (**H**).

Conclusions:

A diagnosis of primary UPS was rendered after consultation with two leading expert pathologists. The diagnosis of primary splenic UPS, which lacks distinctive clinical or imaging features, is essentially a process of exclusion. For therapeutic purposes, the complete immunohistochemistry panel is important to exclude specific lineage differentiation and rule out mimickers with cystic changes. Genetic study may show complex karyotype without specific chromosomal abnormalities. The curative treatment of UPS is radical surgical excision +/- chemotherapy or radiation.

References:

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