Epithelioid Hemangioendothelioma

- Vascular tumors run the gamut from benign hemangiomas to intermediate grade of malignancy epithelioid hemangioendotheliomas, to highly aggressive angiosarcomas.

- Epithelioid hemangioendothelioma (EHE) is a rare vascular neoplasm with distinctive morphologic appearance, presenting as a deep painful soft tissue mass, although can be found primarily in lung, bone, and liver.

- The tumor is most commonly seen as multifocal disease affecting liver, lung, pleura, or several of these sites simultaneously

- Only a minority of these tumors progress over the course of 1–3 years with many appearing largely dominant for a decade or more, suggesting observation as a viable option for management of such patients with unresectable multifocal disease.

- Approximately 20% of EHE are more aggressive and thus may require intervention.
- Liver-only disease has been treated with chemotherapy, bland embolization, chemoembolization, or even liver transplant in those rare patients who develop liver failure from disease.

- A high mitotic and profound nuclei atypia are independent predictors of survival, and the WHO classification suggests the use of “malignant EHE” terminology for this group of lesions, to distinguish from the more bland-appearing examples.
- The primary treatment is surgical excision with an uncertain role for radiation and chemotherapy.