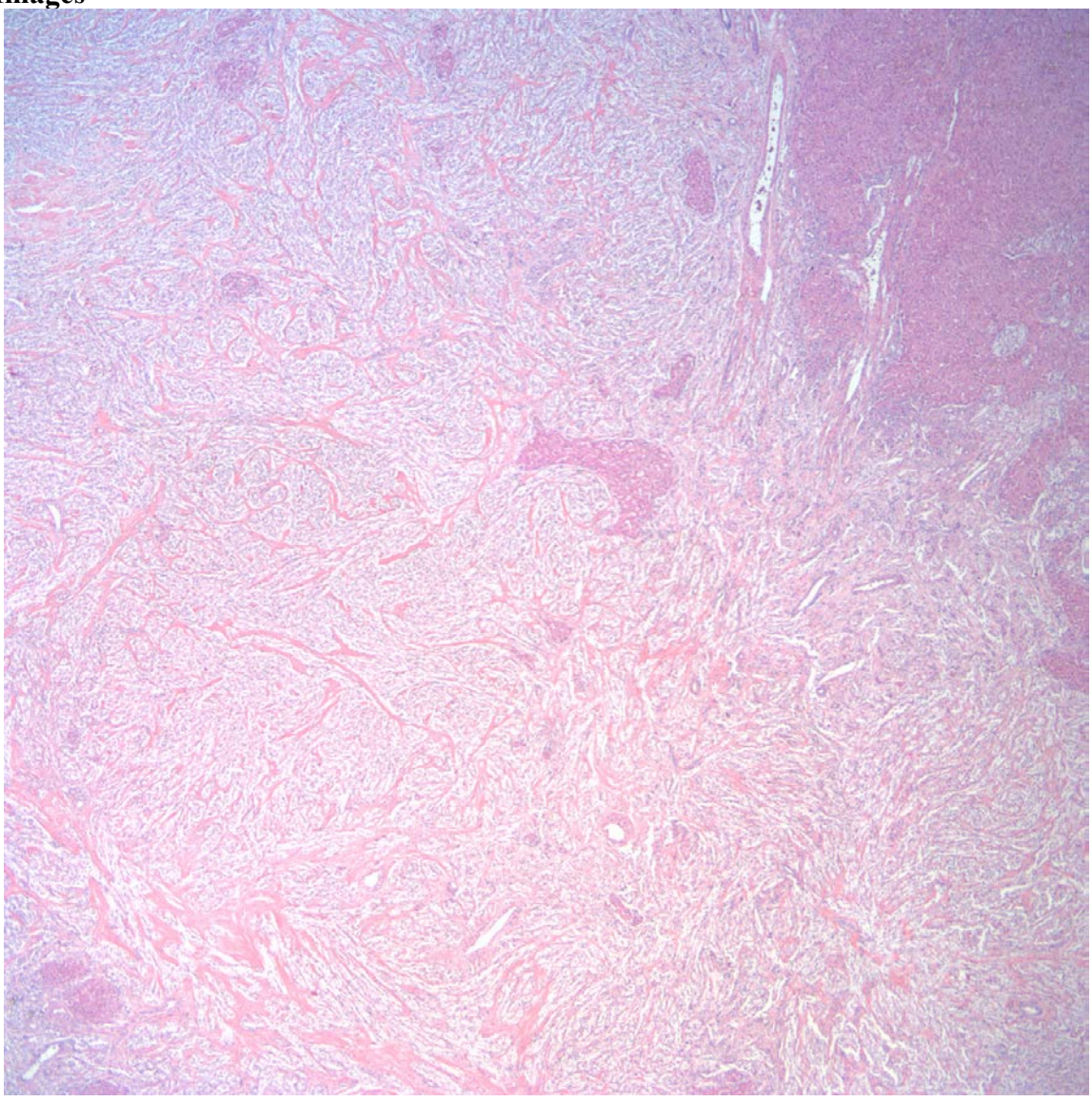
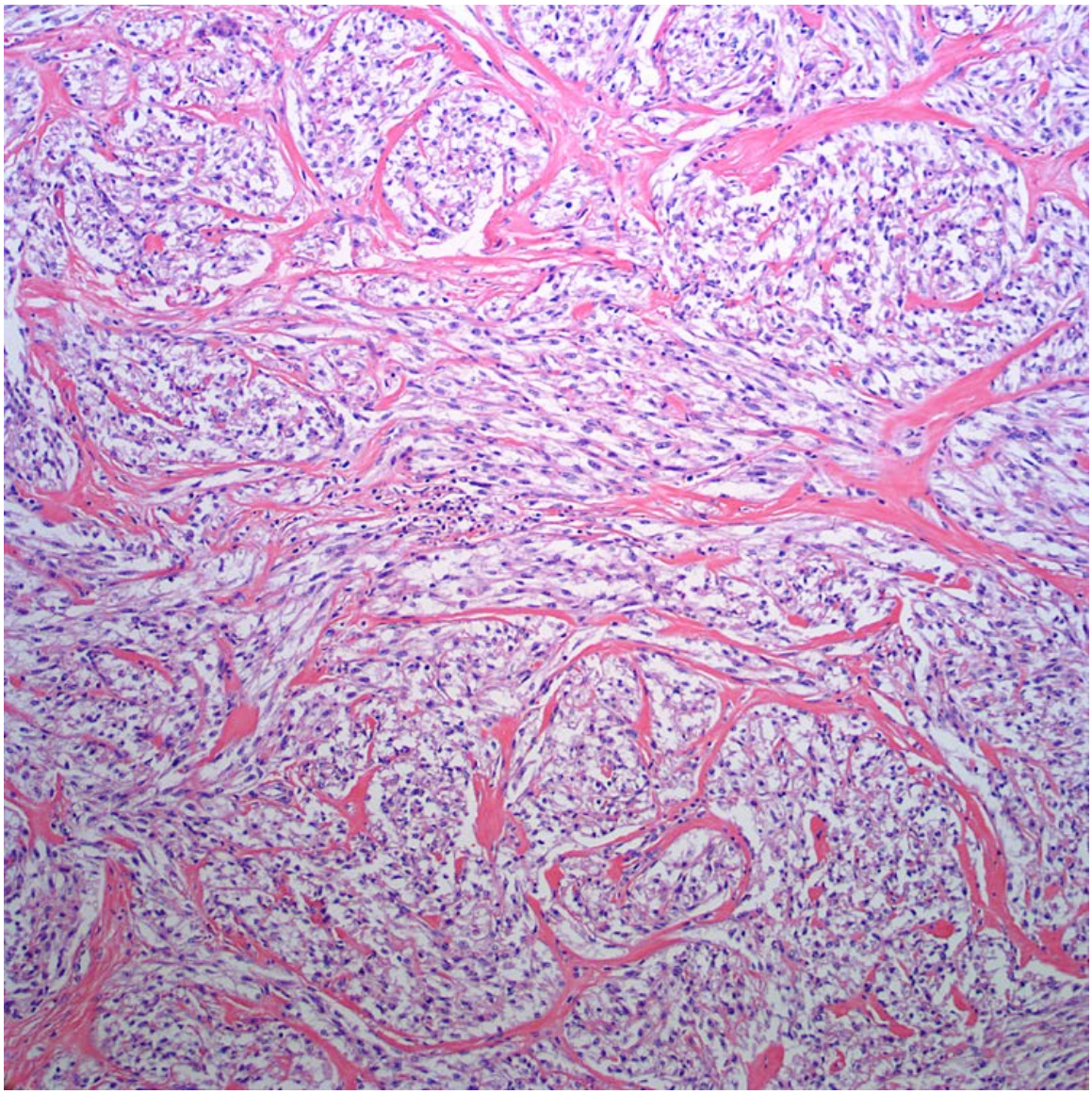
Case

The patient is a 17 year old woman who presented with abdominal pain. A CT-scan revealed numerous masses in the left lobe of the liver, and the patient underwent left hepatic lobectomy. The gross specimen revealed multiple tan-white nodules ranging from 0.5 cm to 7.0 cm. Three years prior, the patient had a 5 cm mass removed from her falciform ligament.

Images





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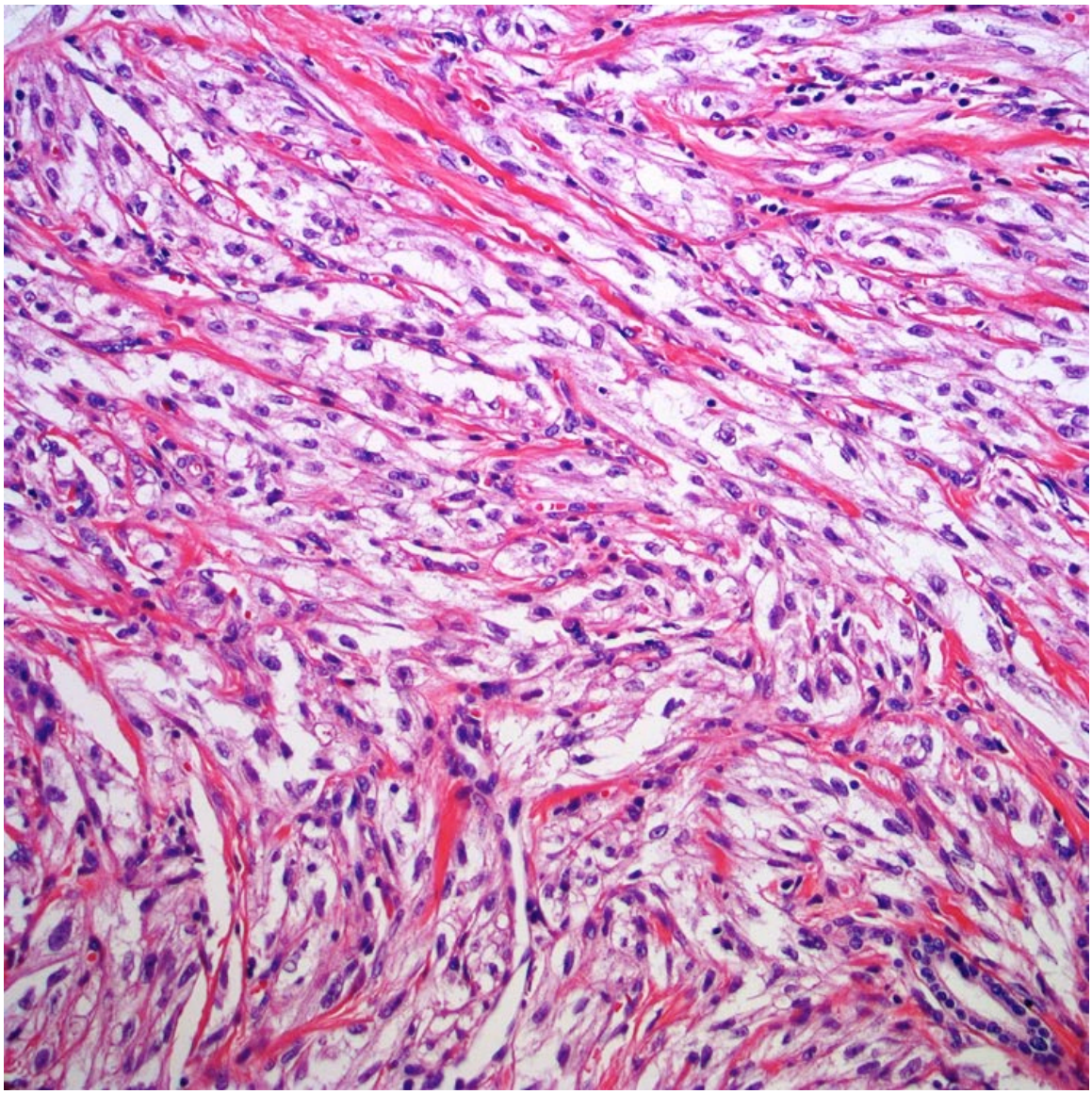

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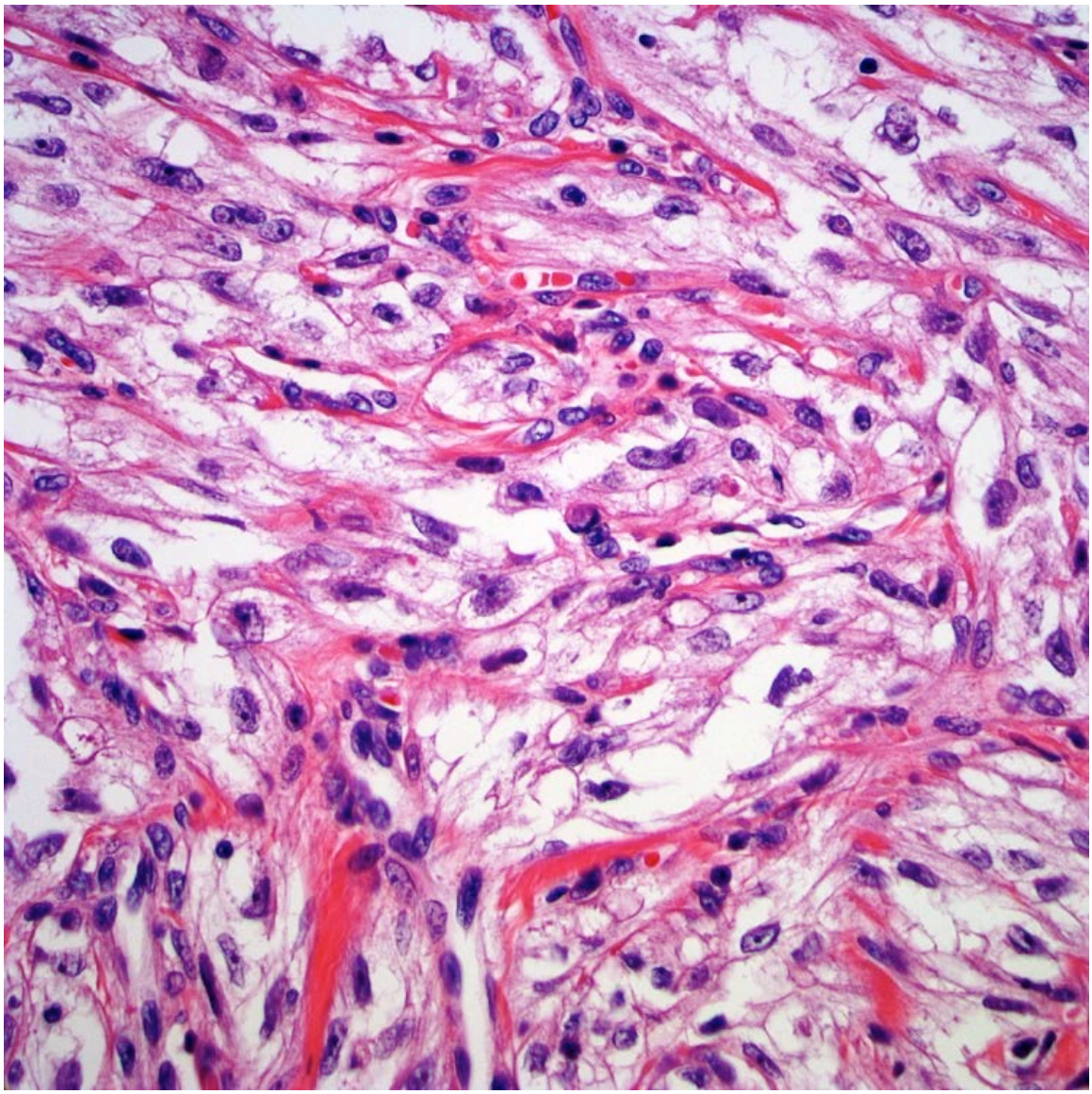

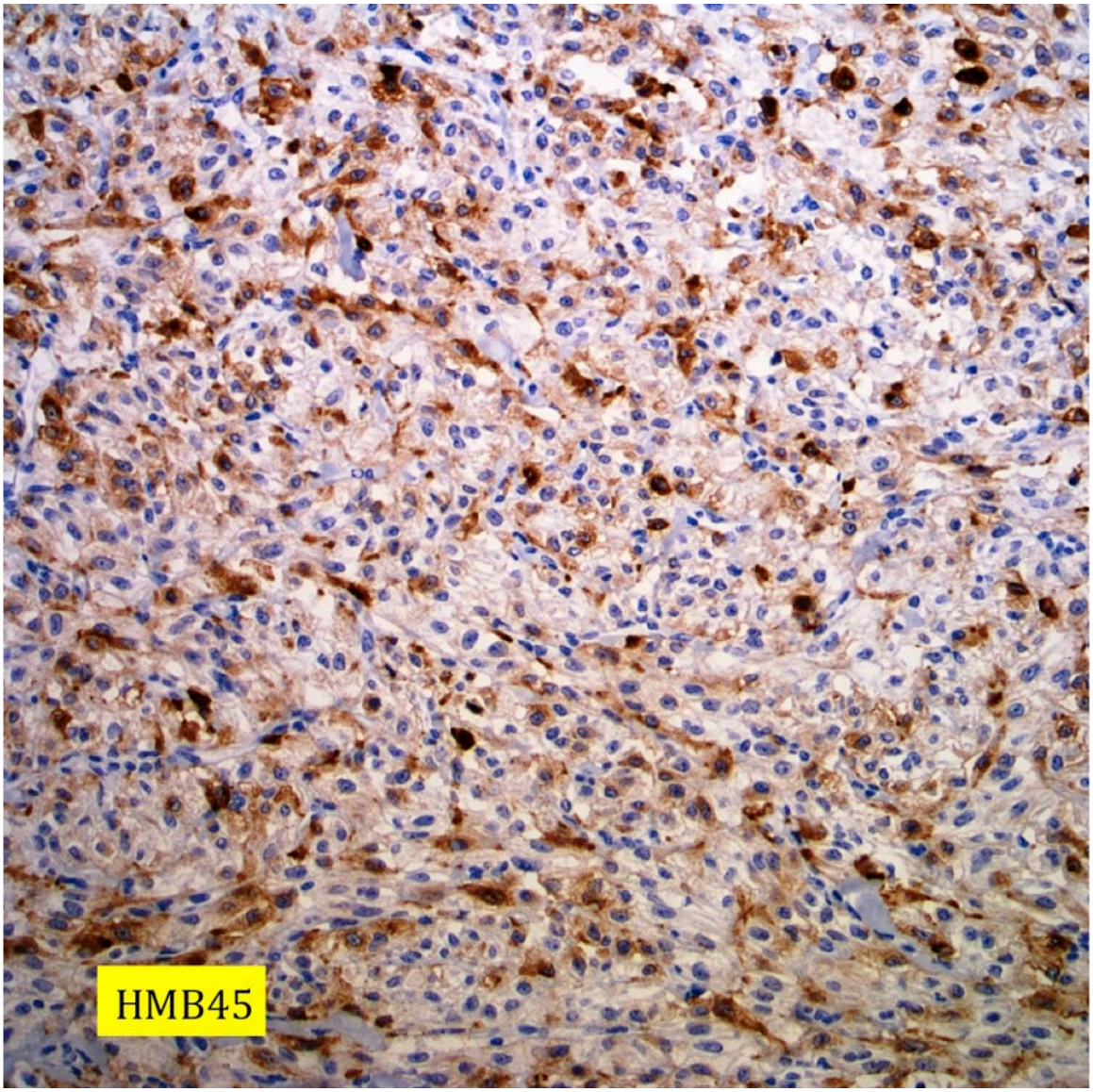
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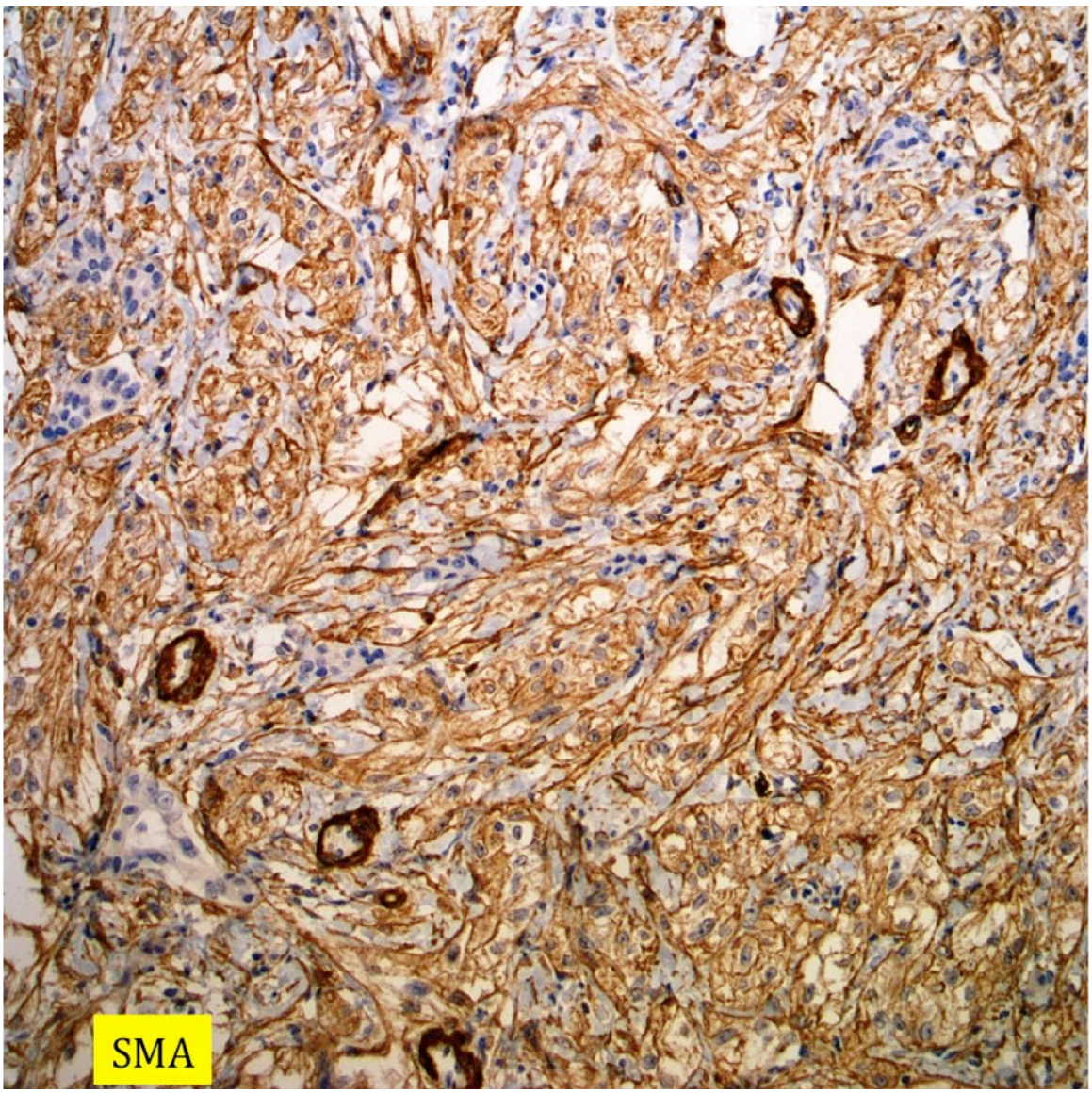

What is your diagnosis?

A) Hepatocellular carcinoma
B) Metastatic clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres (PEComa)
C) Metastatic clear cell sarcoma
D) Metastatic melanoma
E) Metastatic leiomyosarcoma

Answer and Discussion

B) Metastatic clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres (PEComa).

Clear cell myomelanocytic tumor (CCMT) of the falciform ligament/ligamentum teres was first described by Folpe et al in 2000, where the authors described 7 cases gathered

from 3 institutions over a 50-year period. They characteristically occurred in children and adolescents, and in females. Interestingly, the one case which behaved aggressively in that series (radiographic evidence of pulmonary metastasis) was the only case outside of that typical demographic – a 29 year old male.

This tumor belongs in the perivascular epithelioid cell family of tumors (PEComa), which includes angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear cell sugar tumor of the lung (CCST), and very rare tumors in other soft tissue and gynecologic locations. They are defined by the dual expression of both myoid markers (smooth muscle actin) and melanocytic markers (melan-A, HMB-45). They are usually S-100 negative. Microscopically, CCMTs are composed of uniform spindle cells with clear to lightly eosinophilic cytoplasm. The neoplasm is arranged in small nests and fascicles, divided by an arborizing and delicate vascular network, reminiscent of clear cell renal cell carcinoma. Foci of more epithelioid morphology can be seen. The histologic differential diagnosis would include a variety of clear cell and spindle cell tumors including melanoma, clear cell sarcoma, leiomyosarcoma, and clear cell carcinomas (including RCC and clear cell HCC). The dual expression of SMA and melanocytic markers seen in PEComa are not seen in these entities. The characteristic demographic and location (young woman, falciform ligament), clue one in to the specific diagnosis of CCMT.

The behavior of these tumors is uncertain. In 2005, Folpe et al reported 26 cases of non-AML, non-LAM PEComas, 8 of which showed local recurrence and/or distant metastasis. These included PEComas in rare gynecologic and soft tissue locations, including one CCMT. Features that were associated with high risk behavior included size >8 cm, mitotic activity >1/50 HPF, and necrosis. A meta-analysis in 2012 of 234 cases of PEComa-NOS (PEComas other than AML, LAM, and CCST occurring in their typical anatomic location) reported in the English literature showed that primary tumor size ≥5 cm and a mitotic rate ≥1/50 HPF were the only factors significantly associated with recurrence following surgical resection. In this case, the patient's primary tumor was only 5 cm but showed abundant central necrosis. No lymphovascular space invasion or significant cytologic atypia was seen, and the mitotic count was only 1/50 HPF. These features highlight the difficulty in predicting the behavior of these tumors. The patient was alive and well two years after resection, and was subsequently lost to follow-up.

References

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