AMSER Rad Path Case of the Month:

Undifferentiated Pleomorphic Sarcoma

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Changing What's Possible

AMSER

Patient Presentation

Clinical history: Patient is a 56-year-old female with a past medical history significant for recurrent lipomas who presents with two nodules palpable on the posterior aspect of her neck. On interview she reports that she has had surgery to remove lipomas from this region in 2002, 2007, and 2017.

Pertinent social history: Patient is married and lives at home with her family. She denies any recreational drug use.

Pertinent physical exam findings: On exam two firm nodules are palpated on the posterior aspect of the patient's neck. The skin overlying the nodules is erythematous and the area is painful on palpation.

Pertinent Labs

- N/A

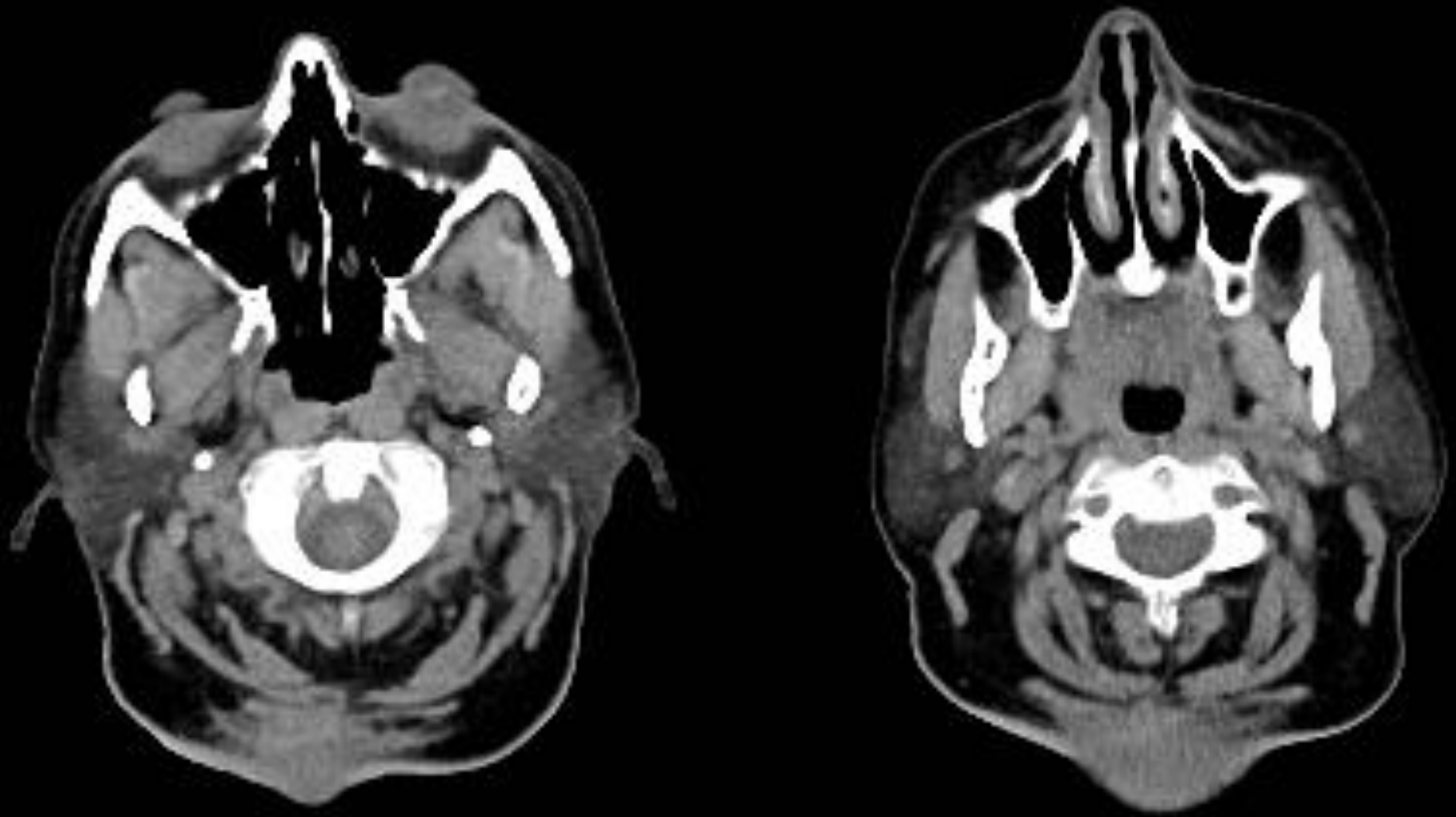

Figure 1:

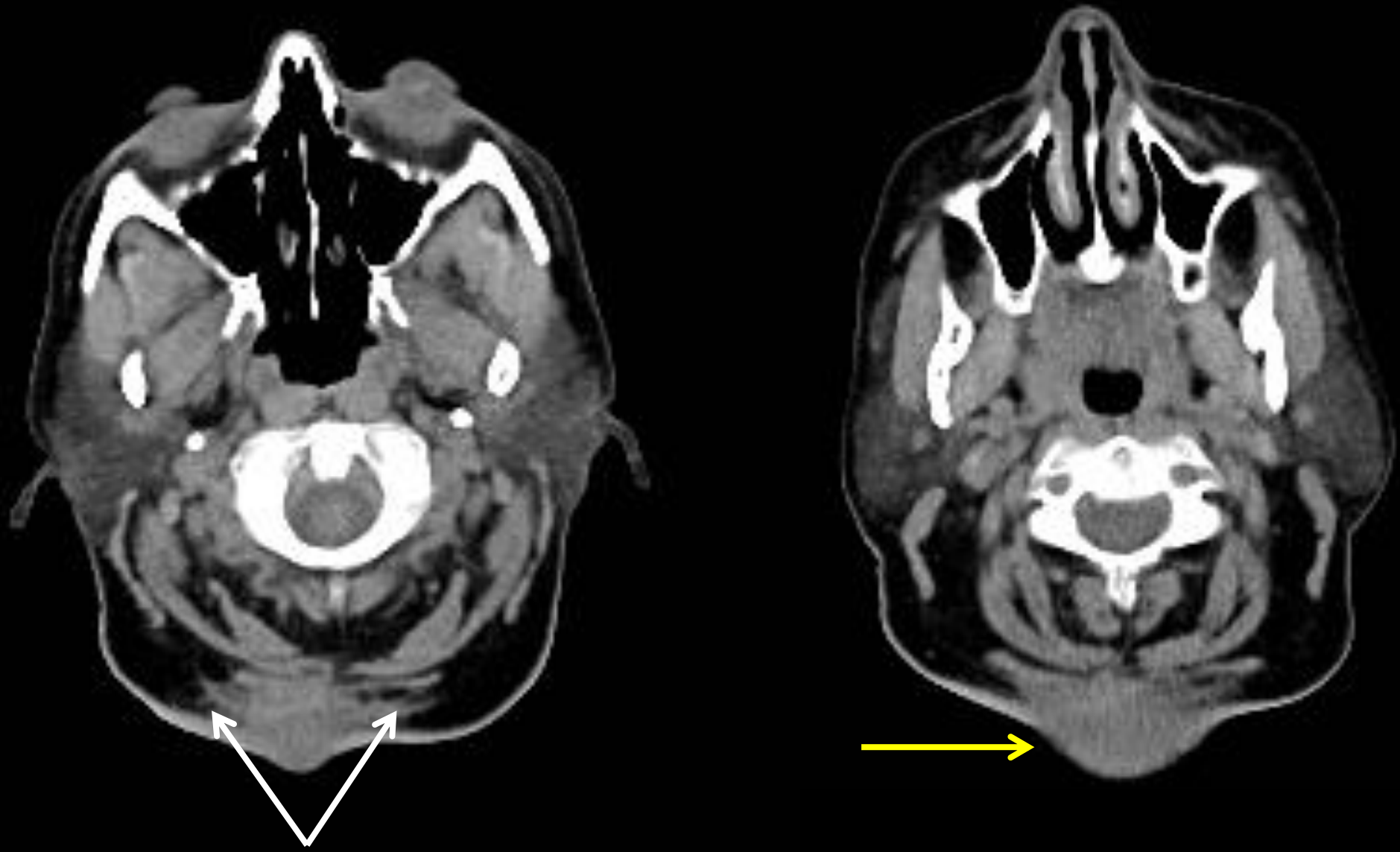

Fig 1: Axial CT images of cervical spine. Yellow arrow indicates 4 cm nodule. Fat stranding (white arrows) can be seen surrounding mass indicating inflammatory infiltration.

DDX (based on imaging)

- Lipoma
- Dedifferentiated Liposarcoma
- Rhabdomyosarcoma
- Undifferentiated Pleomorphic Sarcoma
- Sarcomatoid Carcinoma

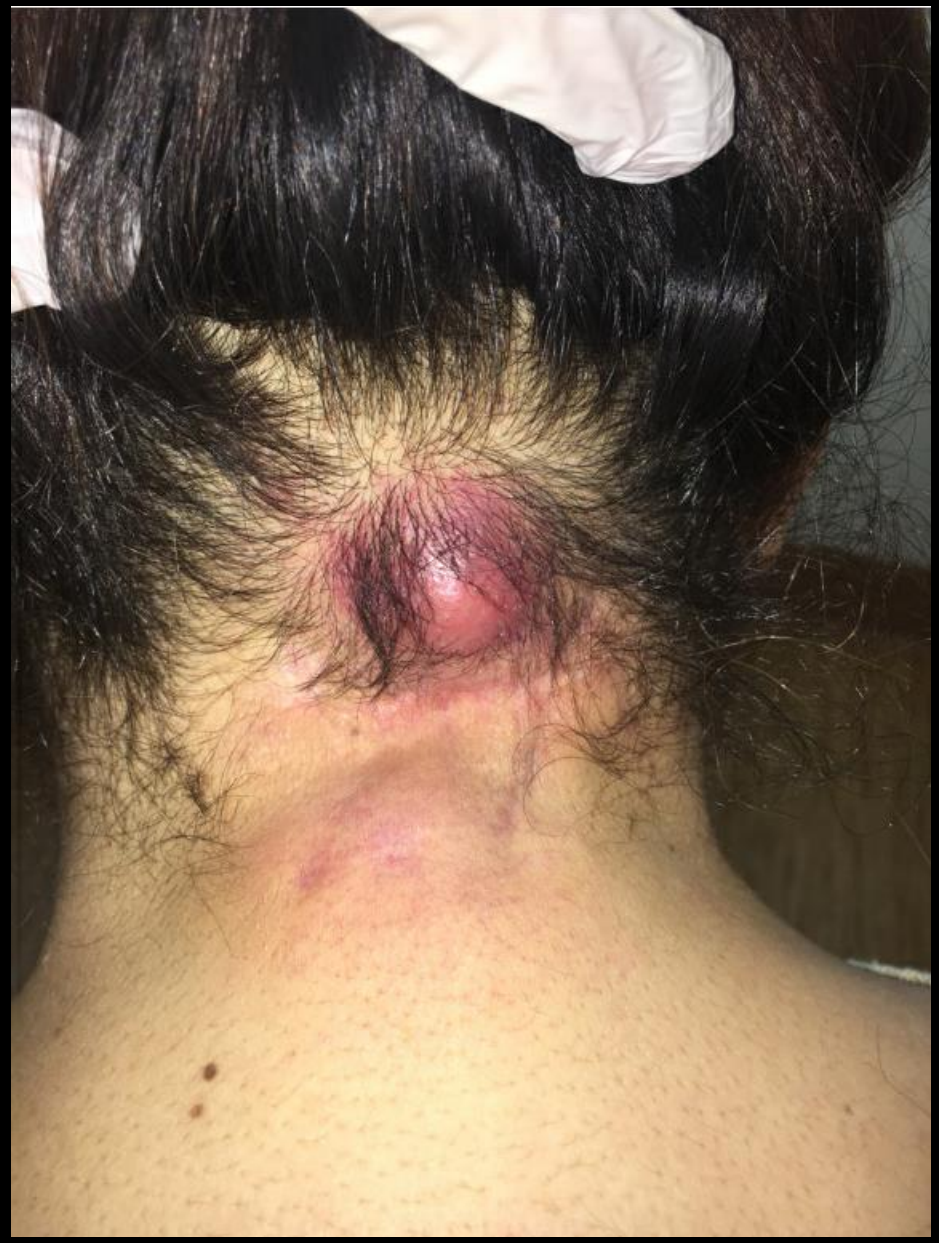

Figure 2: Clinical image of posterior neck. The nodules are firm and painful.

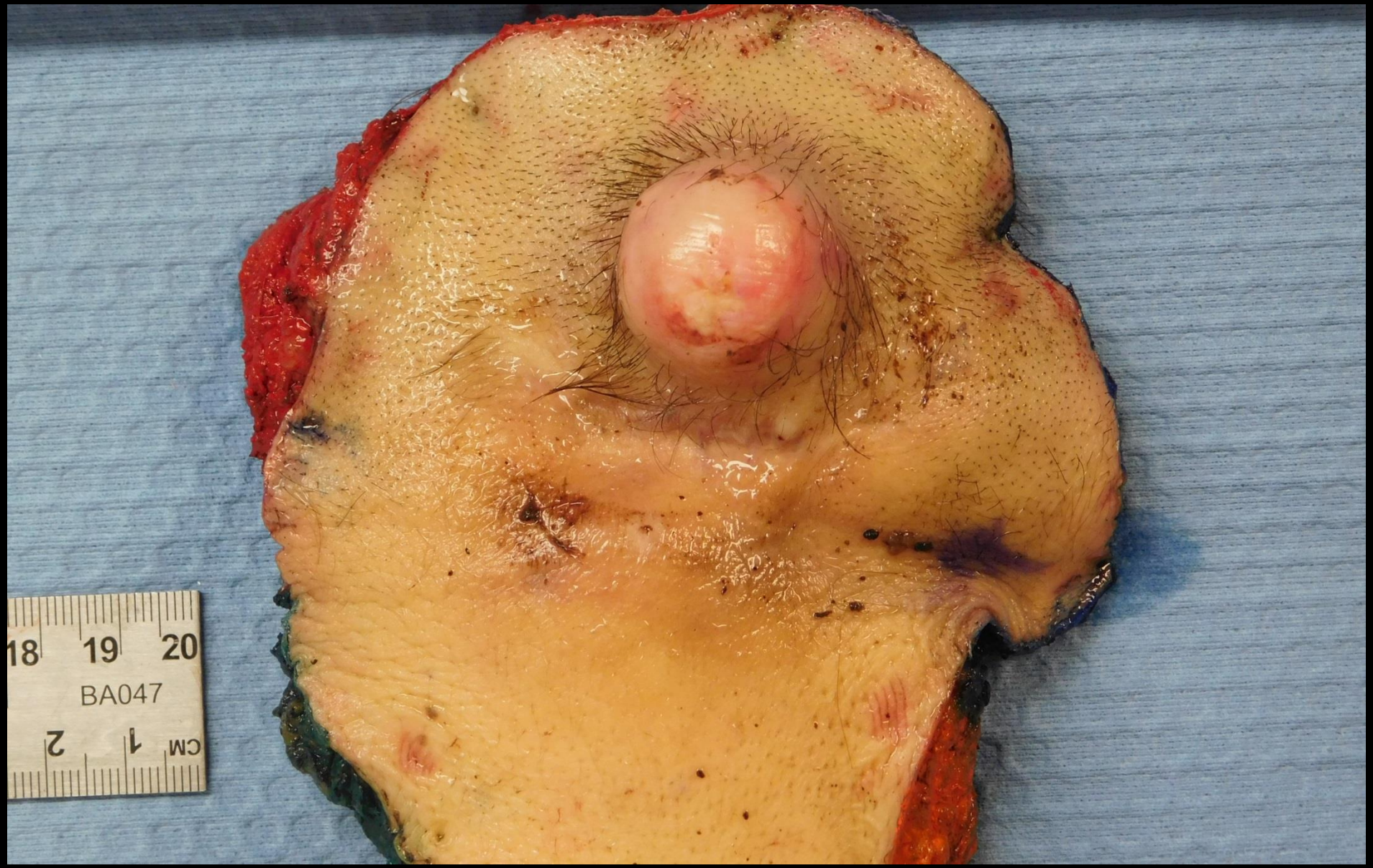

Figure 3. Gross pathology image of resected mass. Tumor appears circumscribed with a fleshy texture

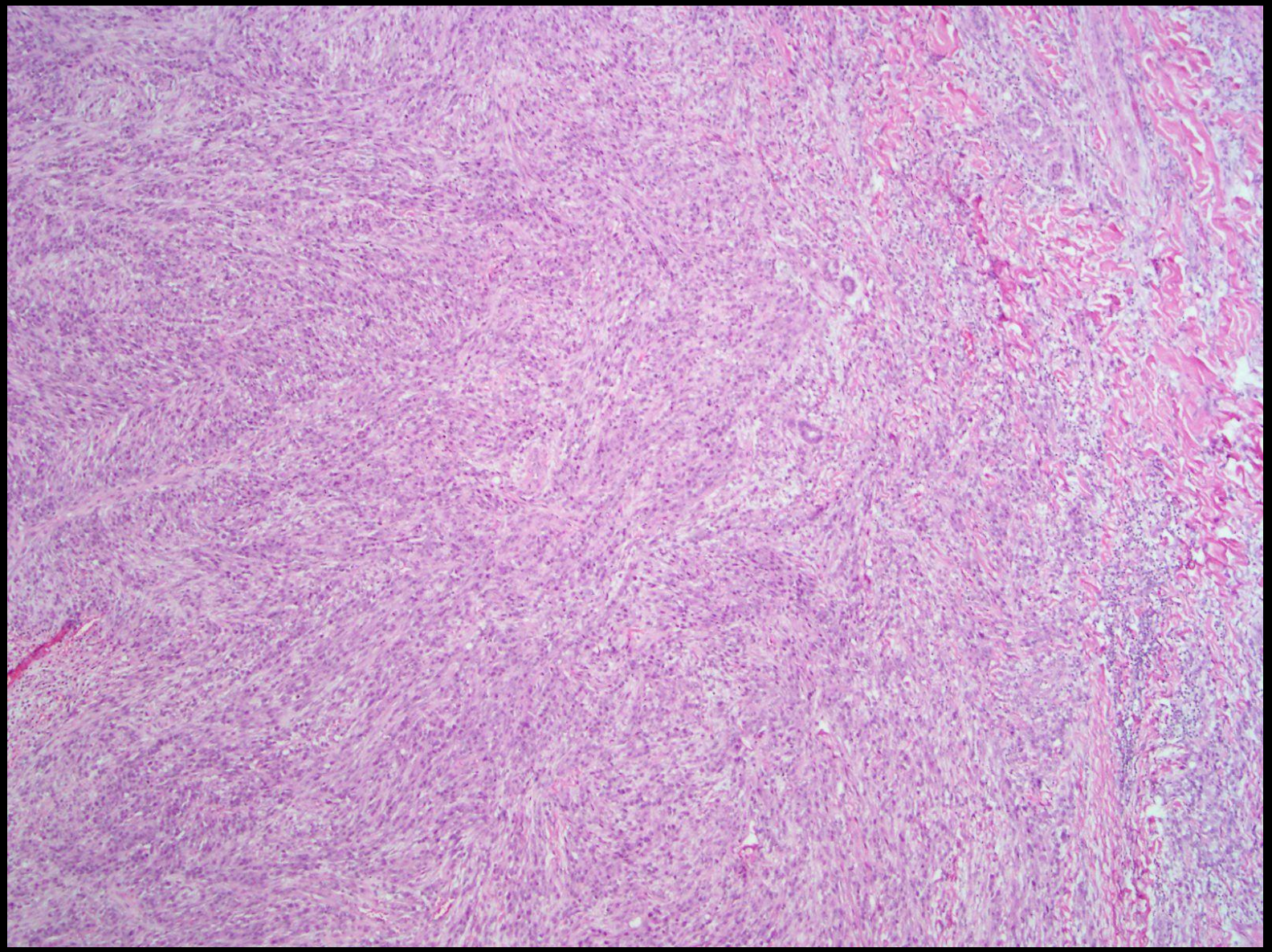

Figure 4. Microscopic pathology of resected mass. Low power view of tumor showing storiform pattern (whorls of spindle shaped cells)

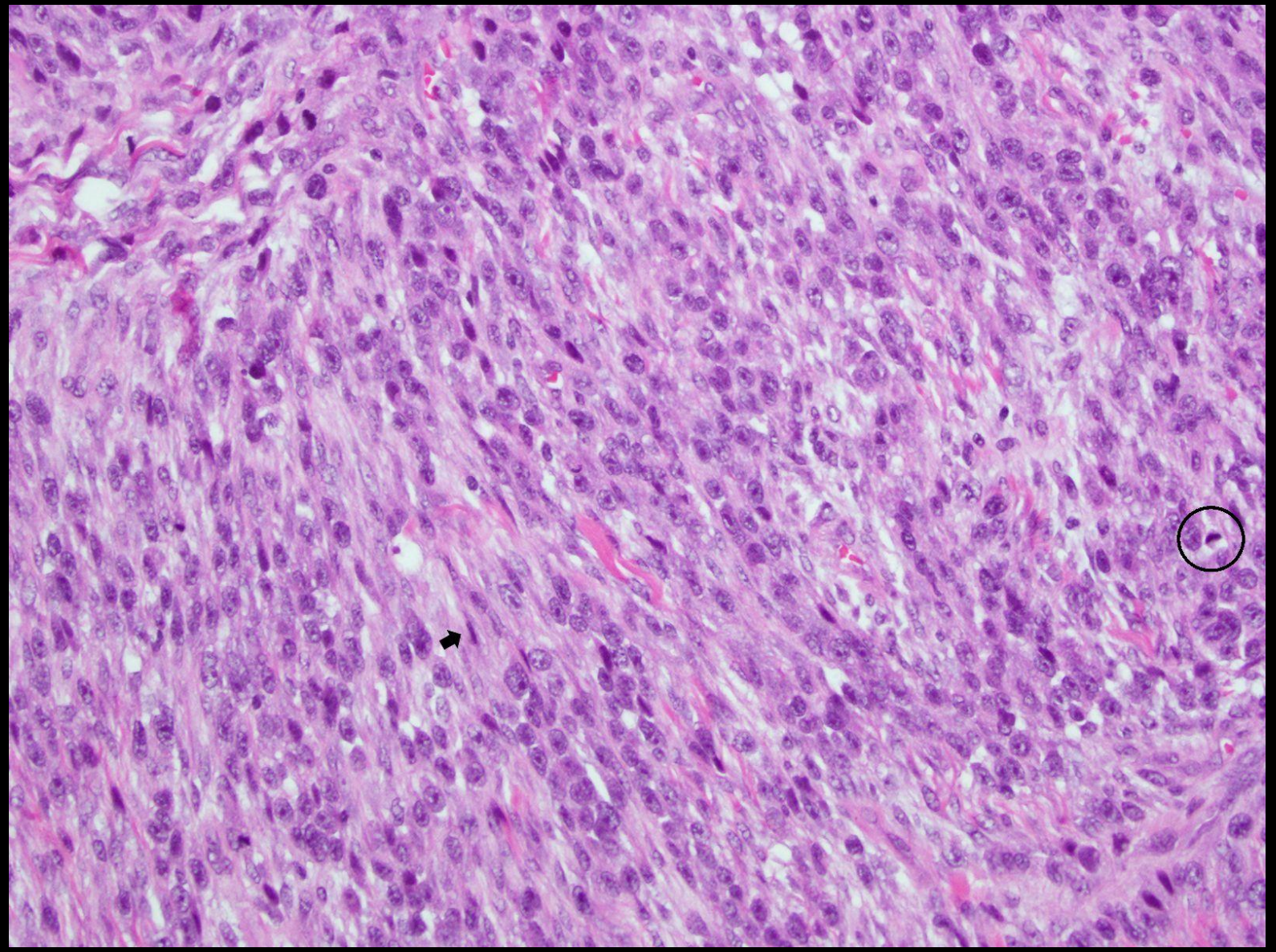

Figure 5. Microscopic pathology of resected mass. Higher power view of tumor showing spindle shaped cells, mitoses, and nuclear pleomorphism. Circle indicates mitoses and arrow highlights spindle shaped cells.

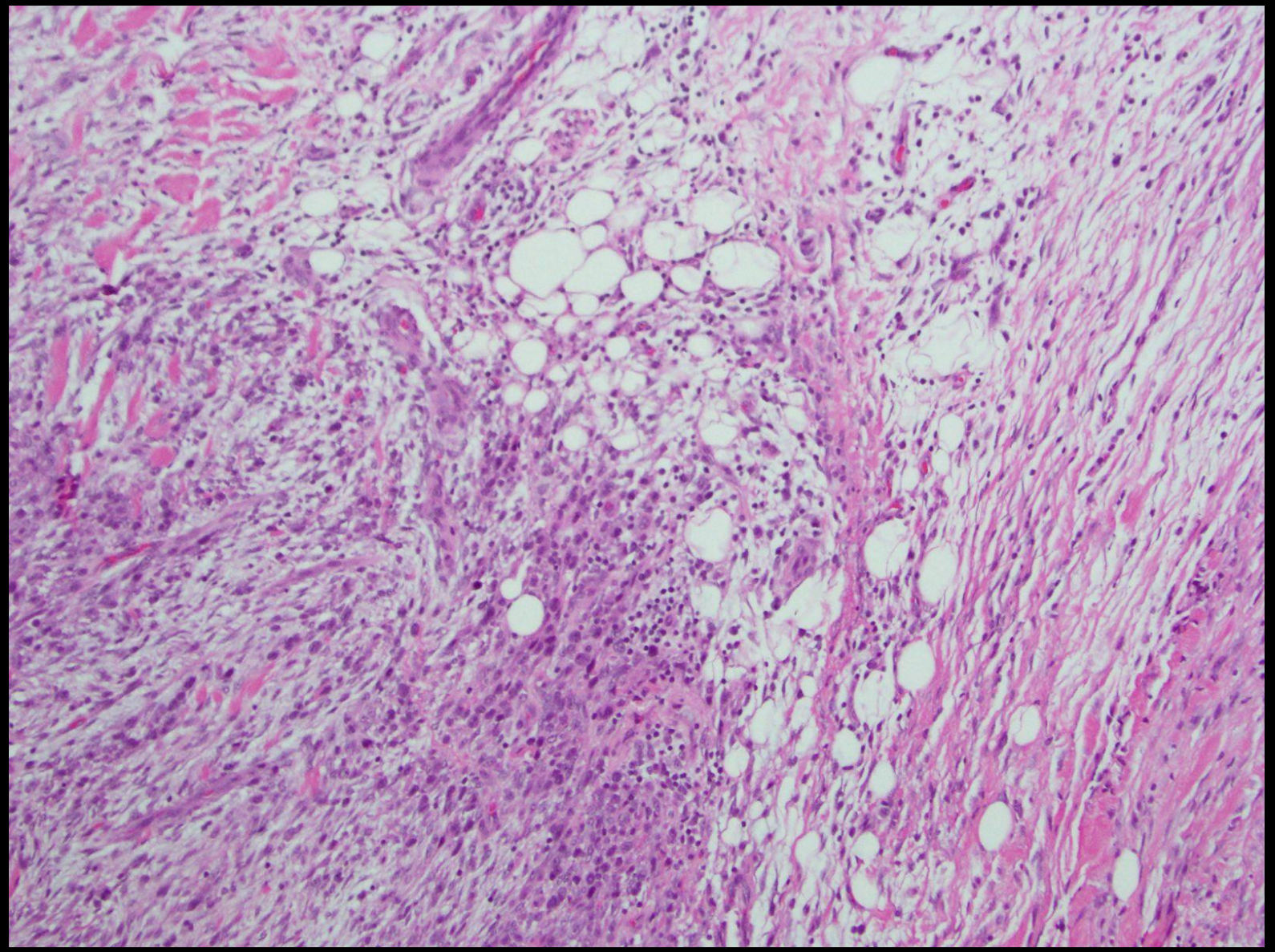

Figure 7. Microscopic pathology of resected mass. Tumor is invasive of local adipose tissue.

Final Dx:

Undifferentiated Pleomorphic Sarcoma

Case Discussion

Background

Pleomorphic undifferentiated sarcomas are fibrohistiocytic soft-tissue tumors. Soft tissue tumors are mesenchymal in origin and can be broken up into several categories including:

- tumors of adipose tissue
- tumors of fibrous tissue
- fibrohistiocytic tumors
- tumors of skeletal muscle
- tumors of smooth muscle
- vascular tumors
- peripheral nerve tumors
- tumors of uncertain histogenesis

Case Discussion

Background Continued

Fibrohistiocytic tumors are made up of fibroblasts and phagocytic cells which resemble histiocytes. However, these cells are not macrophages but are primitive mesenchymal cells, fibroblasts, and myofibroblasts. Examples of this type of tumor include fibrous histiocytomas, giant cell tumors of soft tissue, and pleomorphic undifferentiated sarcoma (PUS).

Case Discussion

Epidemiology

Pleomorphic undifferentiated sarcomas are malignant tumors which have traditionally been considered the most common soft tissue sarcoma, making up 15-20% of all soft tissue sarcomas. However, some tumors which have previously been considered PUSs are being reclassified, many as myogenic sarcoma variants, leading this tumor type to be less common than previously thought. PUSs usually occur in adults with a mean age of presentation of 59 years old and are more common in men (M:F 1.2:1). They may occur in the extremities (most common), retroperitoneum, head, neck, and breast. Risk factors include previous radiotherapy and Li-Fraumeni syndrome.

Case Discussion

Radiologic Characteristics

Pleomorphic undifferentiated sarcomas may be seen as soft tissue masses on plain radiographs and have density similar to muscle on CT. They are found within or around muscle and may present with calcifications. PUSs are also identifiable on imaging by their large size, with retroperitoneal tumors often reaching 20 cm. The tumors will enhance with contrast and calcifications may be seen in 15%. On imaging primary differential considerations would include other soft tissue tumors or metastases.

Case Discussion

Pathologic Characteristics

Grossly pleomorphic undifferentiated sarcomas appear somewhat circumscribed and have a fleshy surface with areas of hemorrhage. Microscopically these tumors characteristically demonstrate a storiform pattern of spindle shaped cells. Important cellular features include nuclear pleomorphism, numerous mitoses, and commonly necrosis. As the features of PUS are non-specific immunohistochemical staining is useful in ruling out similar appearing malignancies. Pleomorphic sarcoma may be further classified as PS with giant cells or PS with inflammation. Although similar in appearance to liposarcomas, they may be distinguished by the absence of lipoblasts.

Case Discussion

Grading

The grading of soft tissue sarcomas is done using the FNCLCC grading system which uses differentiation (1-3), mitotic activity (1-3), and extent of necrosis (0-2) as criteria. Differentiation is given a 3 if the tissue is undifferentiated or poorly differentiated. Mitotic activity is given a 3 if there are 20 or more mitoses/10 HPF. Necrosis is given a 2 if greater than 50% of the tumor is necrotic.

- Grade 1: 2-3
- Grade 2: 4-5
- Grade 3: 6-8

Based on this grading system this patient had a histologic grade of 3 due to a high mitotic rate and poor differentiation.

Case Discussion

Treatment and Prognosis

Pleomorphic undifferentiated sarcomas are aggressive tumors which frequently metastasize. Their prognosis depends on location, with tumors in the extremities having a ~75% 5-year survival rate, while that number drops to 25% in the head and neck. Treatment includes a wide surgical resection and adjuvant radiotherapy for high grade lesions.

References:

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