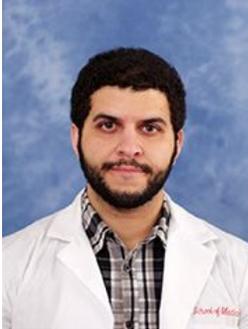


May 2016

A 25 year old female with a palpable mass in the right lower quadrant of her abdomen

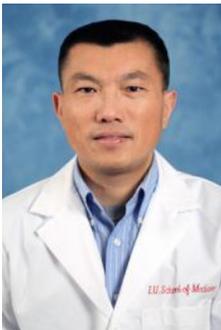
Contributed by:



**Paul Ndekwe, MD, Resident Physician, Indiana University School of
Department of Pathology and Laboratory Medicine, PGY-3.**

Medicine,

and



**Shaoxiong Chen, MD, PhD, Assistant Professor, Indiana University School of Medicine, Department of
Pathology and Laboratory Medicine.**

CLINICAL HISTORY:

The patient is a 25 year old female who presented with a palpable mass in the right lower quadrant of her abdomen. Abdominal CT imaging revealed a 6.3 x 4.3 x 4.2 cm complex enhancing mass that appeared to be arising from the right ovary. Laparoscopic exploration of the abdomen revealed that the mass was actually arising from the mesentery of the small bowel. Resection of a portion of the small bowel to include the mesenteric mass was then performed.

GROSS EXAMINATION:

A 7.4 x 5.6 x 5.4 cm gray-tan to white, granular, nodular mass was identified within the mesentery of the segment of small bowel.

MICROSCOPIC EXAMINATION:

Sections of the mass show a uniform appearing spindle cell neoplasm with cells having small ovoid or more tapering nuclei and delicate cytoplasmic processes. In areas, the tumour cells are associated with a copious myxoid matrix, but elsewhere there is abrupt transition to a more fibrous stroma. There are cellular whorls of tumour cells having with focal areas having an organoid appearance. There is strong and diffuse immunopositivity for MUC-4.

Figure 1. Bland spindle cells with delicate cytoplasmic processes are identified within alternating areas of fibrous and myxoid stroma.

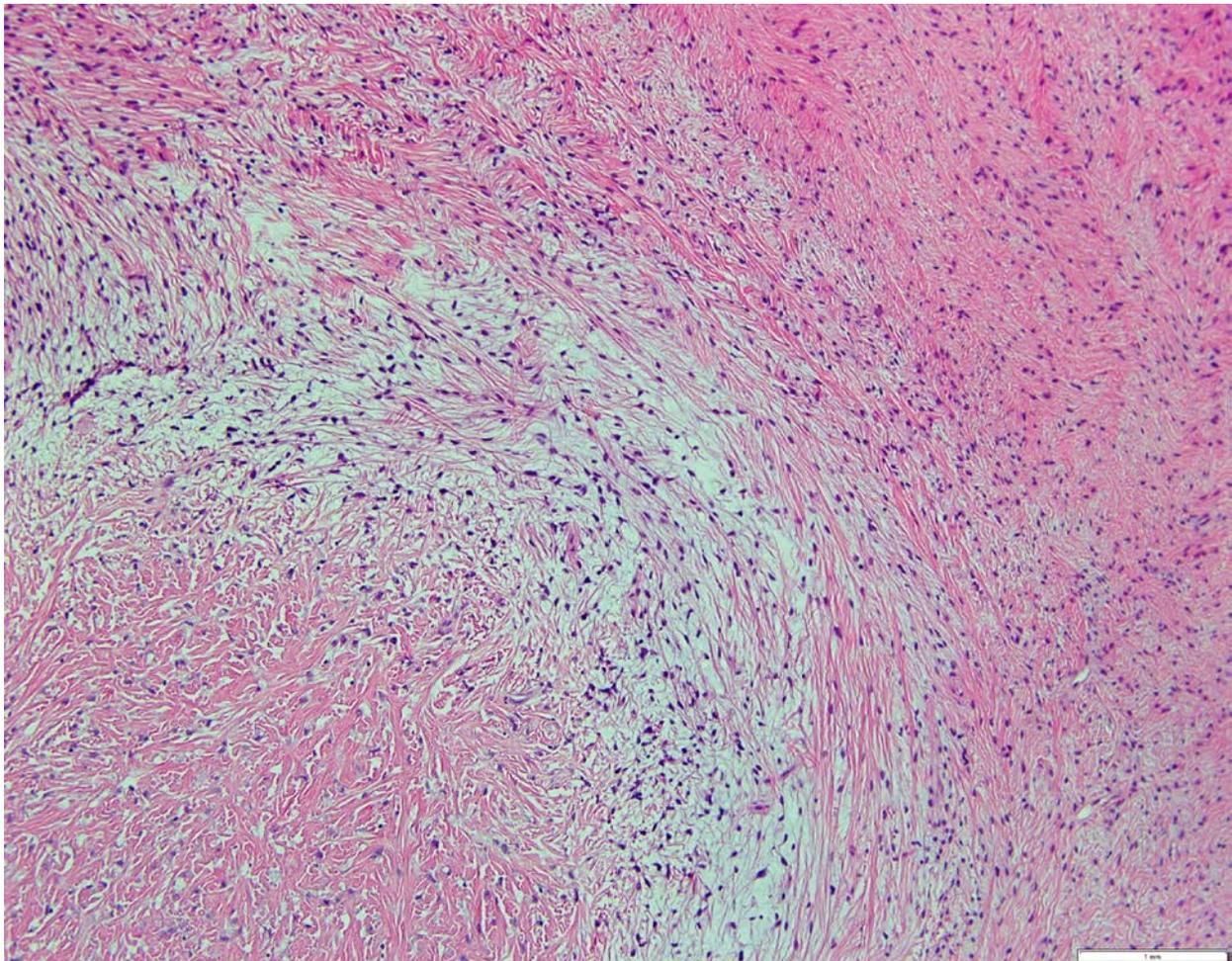


Figure 2. Focal area demonstrating a whorled growth pattern. Also note the alternating areas of fibrous and myxoid stroma.

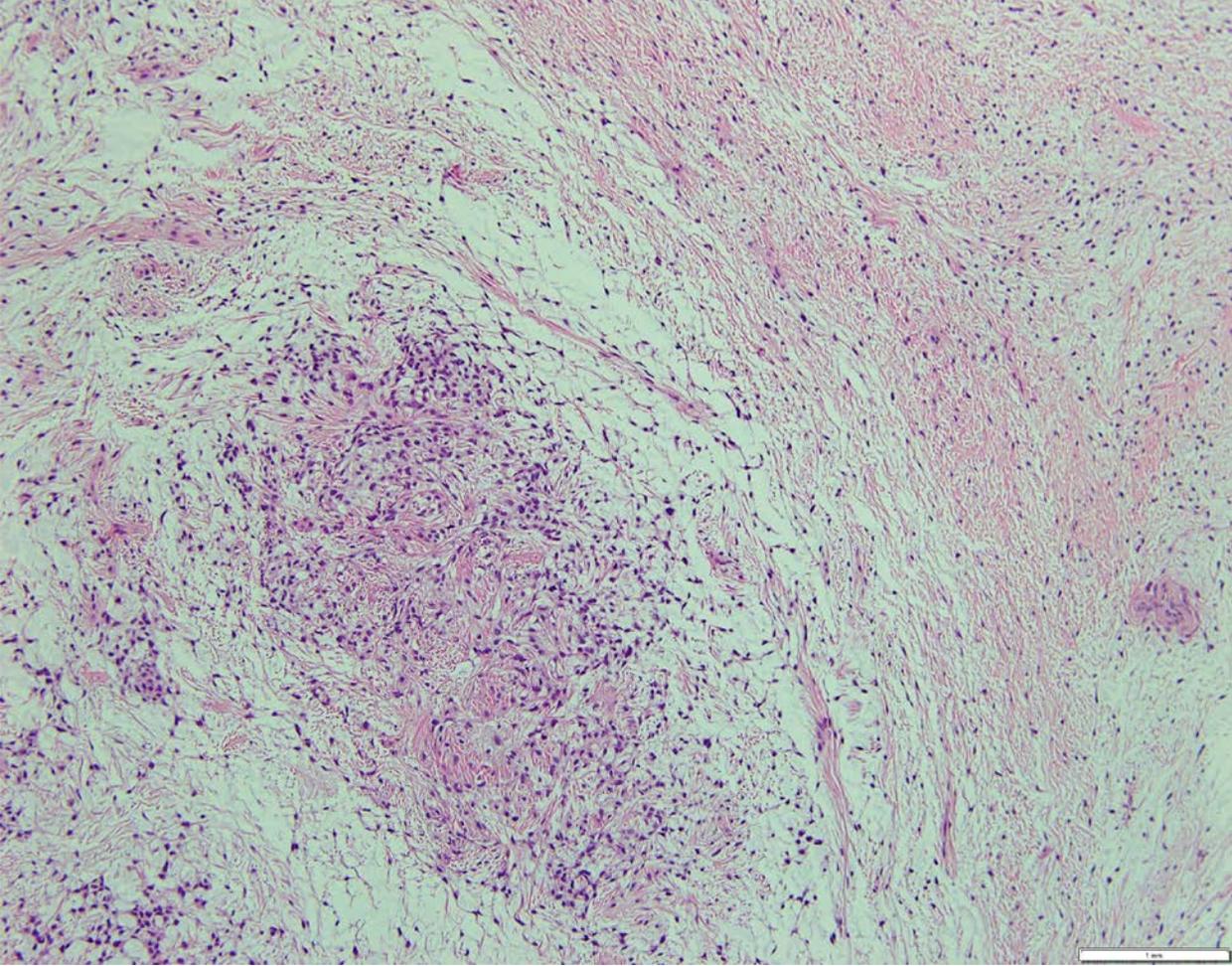
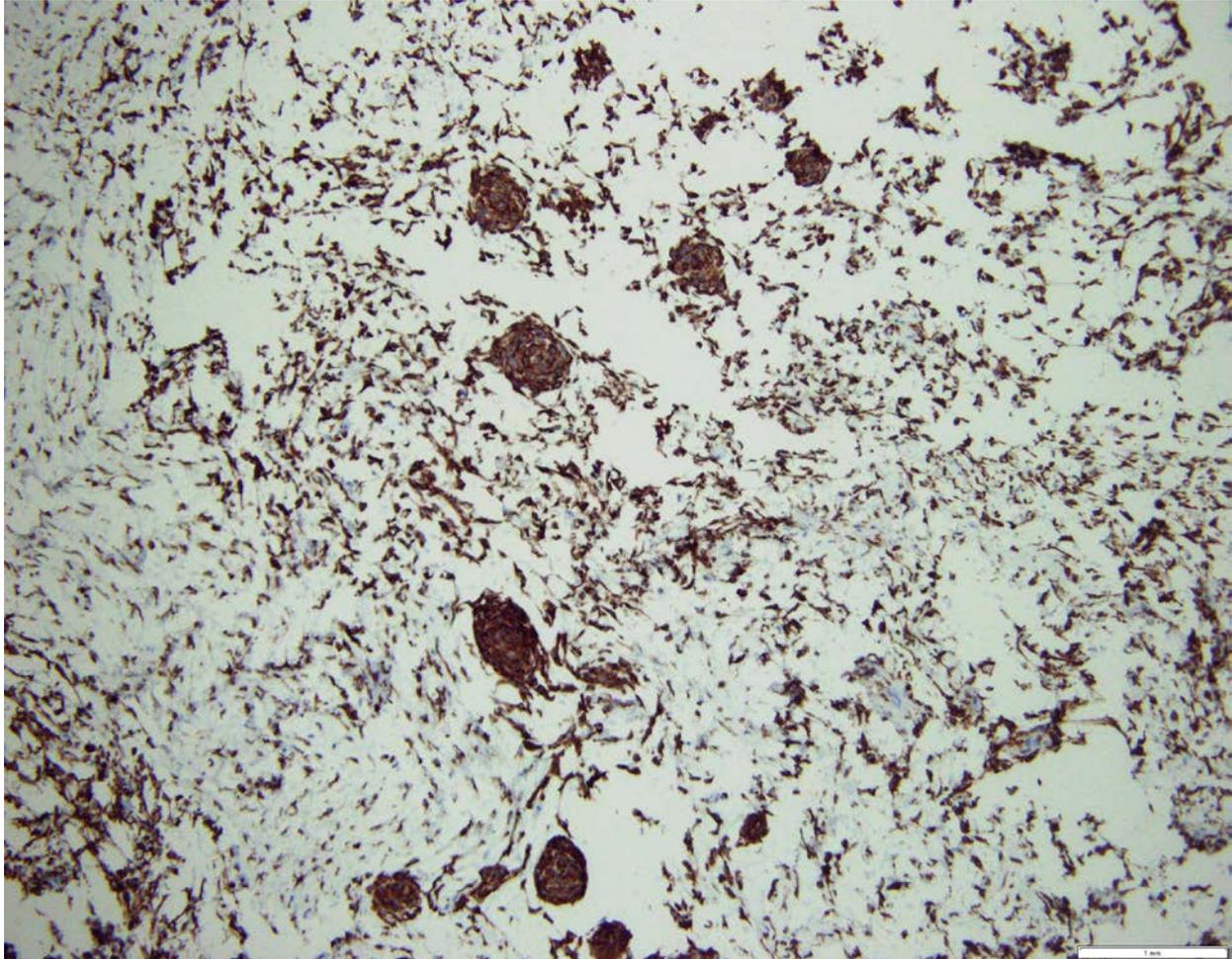


Figure 3. Diffuse MUC-4 Positivity within the tumor cells.



FINAL DIAGNOSIS

Low grade fibromyxoid sarcoma (LGFMS)

DISCUSSION

Low grade fibromyxoid sarcoma is, microscopically speaking, a relatively benign appearing spindle cell neoplasm characterized by the presence of bland appearing fibroblastic spindle cells often configured in a whorled pattern with alternating interspersed collagenous (fibrous) and myxoid areas [1- 7]. Aside from a whorled configuration, the spindle cells can also be seen arranged in a storiform or linear pattern [4]. The neoplastic spindle cells are usually small, with eosinophilic cytoplasm, round to oval shaped nuclei, and inconspicuous to absent nucleoli [6]. Cellularity within these tumors is usually low to moderate, with increased cellularity occasionally identified focally and around vasculature [4]. Necrosis

is usually absent [6]. Rare mitoses and minimal vascularity are typical of these tumors [4, 6]. However, prominent capillary networks may be seen in the more myxoid areas of the tumor [4]. Moderate focal nuclear pleomorphism may be present, but is usually absent [4]. The borders of the tumor can be well defined or somewhat infiltrative in spite of the gross impression [4]. In metastases there may be increased cellularity, focally increased nuclear pleomorphism, and somewhat increased mitotic activity [4]. Given its typically benign appearance and histological heterogeneity, establishing the diagnosis can be quite difficult [3].

Low grade fibromyxoid sarcomas typically show diffuse positive immunohistochemical staining with vimentin and MUC4. Occasional weak focal positive staining with other immunohistochemical stains such as smooth muscle actin, desmin, keratin, S100, CD34, CD31 and EMA may be seen [4, 6].

The histologic differential diagnosis for LGFMS includes essentially any benign appearing spindle cell proliferation with myxoid areas [6]. Desmoid fibromatosis, myxoid neurofibroma, low-grade myxofibrosarcoma, myxoid solitary fibrous tumor, perineurioma, and various myxomas are just a few examples of entities that may be included in the differential diagnosis [4]. Differentiating LGFMS from these other lesions usually relies on having a specimen that is histologically representative, and on the immunohistochemical profile of the lesion.

Additionally, it is believed that a high number of low grade fibromyxoid sarcomas have a t(7;16)(q32-34;p11) chromosomal translocation, leading to the creation of the FUS-CREB3L2 gene [8]. Other cases of LGFMS not demonstrating this translocation may have a t(11;16)(p11;p11) translocation, resulting in a FUS-CREB3L1 gene fusion [8]. Therefore, a diagnosis of LGFMS can be confirmed by utilizing FISH to detect the FUS gene rearrangement [9].

Grossly, these tumors are typically well circumscribed and heterogeneously fibrous to myxoid in appearance. Occasionally there may be evidence of infiltration beyond the circumscribed borders [4].

Clinically, LGFMS occurs mainly in young adults [4]. However, it can occur in older adults, teens and children [4]. It is a rare entity, and accounts for less than 1% of all soft tissue sarcomas [3]. These tumors usually arise from the deep soft tissues of the thigh, trunk, and shoulder area [4]. Cases of LGFMS have also been noted to arise from the subcutaneous soft tissue at various sites [4,5]. Low grade fibromyxoid sarcomas are typically slow growing [6]. Unlike many other low grade sarcomas, they have a relatively high potential for metastasizing, sometimes decades after the initial diagnosis [1-3, 6]. Due to the risk of metastasis, accurate diagnosis and careful clinical follow-up are needed [3]. The treatment for LGFMS is typically wide local resection of both primary tumors and metastases. In some situations chemotherapy or radiotherapy may be used [3].

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