

Low grade fibromyxoid sarcoma presenting as a pelvic mass

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Abstract

Low grade fibromyxoid sarcomas are rare lesions that typically develop in the soft tissue of the extremities and trunk. This patient presented with a somewhat unusual location in the mons pubis, and was treated by primary excision. She will require long term follow up, as local recurrences and late metastases are possible.



Figure 1. Pelvic CT demonstrating location of mass (arrow).



Figure 2. Gross appearance of lesion during excision.

Case Report

A 39-year-old healthy female was referred with a primary complaint of a firm, painless soft tissue mass in the midline of the mons pubis, and clearly outside of the abdominal cavity. The patient reported gradual enlargement over a one to two year period prior to presentation. There was no history of trauma.

A CT scan (Figure 1), demonstrated a heterogeneous mass, approximately 5 cm in diameter, with an average Hounsfield measurement of 34.47 units. A frozen section biopsy under anesthesia initially suggested an angiomyxoma, and the lesion was removed in its entirety. Gross examination demonstrated a firm, pseudoencapsulated mass (Figures 2 and 3), which on pathologic evaluation proved to be a low grade fibromyxoid sarcoma (LGFMS) comprised of predominantly spindle-appearing cells (Figures 4 and 5). No mitotic figures were identified, and the pathologic stage was pT1apNx. LGFMS may have a rather bland histologic appearance, and are sometimes difficult to distinguish from other low grade sarcomas and benign mesenchymal tumors unless the characteristic genetic translocation [t(7;16)(q34;p11) or t(11;16)(p11;p11)] is identified. Even such stains as anti-CD34 or vimentin may not be specific enough to clearly identify LGFMS. In this case, strongly positive staining with Mucin 4 transmembrane glycoprotein (MUC 4) and weak staining with epithelial membrane antigen (EMA) confirmed the diagnosis. Positive staining for MUC 4 was recently proposed as a highly specific marker for LGFMS, and a high percentage of LGFMS will stain focally with EMA as well. Despite the gross appearance of encapsulation, these lesions have a propensity for local recurrence, and late distant metastases.



Figure 3. Gross appearance of lesion (note pseudocapsule).

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After consultation with an academic sarcoma clinic, close observation without further therapy is now being undertaken.

Reference

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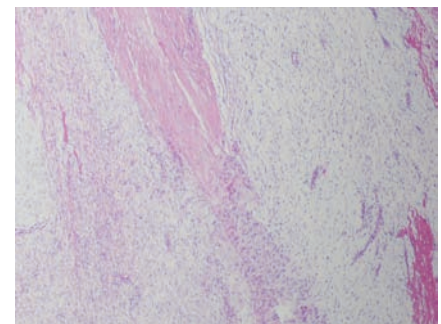


Figure 4. Histologic appearance, demonstrating predominantly spindle appearing cells (H&E stain).

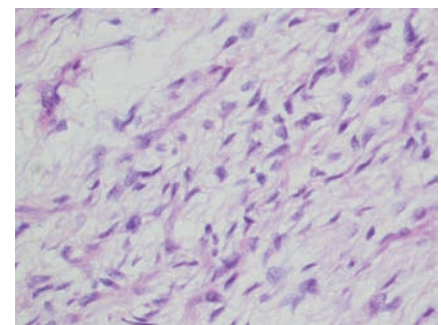


Figure 5. Higher power magnification of area shown in Figure 4.