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.

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, pseudocapsulated 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.

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

Reference