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EPSTEIN-BARR VIRUS-ASSOCIATED SMOOTH MUSCLE TUMOR IN A LEIOMYOSARCOMA COHORT

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Introduction: Epstein-Barr Virus-Associated Smooth Muscle Tumor (EBVSMT) is a rare tumor with higher rate of occurrence in unusual locations, such as soft tissues and retroperitoneum in patients often in the setting of immunodeficiency. Characteristically, they show mild nuclear atypia with low mitotic activity and accompanying lymphocytes. In this study, we evaluated a cohort of leiomyosarcomas for the presence of EBV.

Material&Methods: Ninety-three leiomyosarcoma cases occurring various locations were reviewed and 3-4 mm diameter tissue microarrays were formed. Following manufacturer's instructions, ISH for EBER (Leica Bond Ready to Use ISH) is performed on the microarray slides using Leica Bond Autostainer. Diffuse nuclear staining was regarded as positive.

Results: Seventy-five cases (80%) were females and rest were males. Mean age was 55 ± 13 (27-91). Uterus (55) was the most common location followed by intraabdominal/retroperitoneum (18), extremities (7), trunk (5) and head&neck (5), scrotum (2) and bone (1). Only 2 non-uterine cases were positive for EBER, one of which occurred within pancreas, and the other was located at chest. None of the patients had known immunosuppression or accompanied lymphocytes. The first case was 56-yearold female with a 19x8x8 cm pancreatic mass, metastatic to lung and spleen at presentation. She also had a 5 cm, exophytic right renal mass, diagnosed as conventional renal cell carcinoma at presentation, and a 2 cm left kidney mass diagnosed as type 1 papillary renal cell carcinoma 2 years later. She received chemotherapy and underwent several metastatectomies from lung and liver after 3 years and 6 years, respectively. She was lost to follow up with enlarging pulmonary metastatic nodules 7 years after the presentation. Morphologically, multinodular tumor was composed of long fascicules of smooth muscle cells with low grade features. Focal areas of higher nuclear grade, epithelioid cells, necrosis and vascular invasion were present. Mitosis was 1-2/10 hpf. Neoplastic cells were positive for SMA, desmin, CD34, and EMA while S100 and c-kit were negative. The second case was 55-year-old female with 7x5x5 cm soft tissue mass located at chest wall beneath the right breast infiltrating right anterior parts of 9-10th ribs. Patient underwent into marginal surgical excision and post-surgical chemotherapy, and lost to follow up after 13 months. Tumor was vaguely nodular and composed of fascicules formed by pleomorphic spindle cells with a mitotic activity of 20/10 hpf. Bone and pleural invasion, focal areas of necrosis were also present. SMA showed diffuse and strong positivity and panCK was negative.

Conclusion: EBV does not seem to play a role in the etiology uterine leiomyosarcomas. EBVSMT can also occur in immunocompetent patients; however, may be associated with other malignancies. EBER

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testing may be helpful in diagnosing smooth muscle tumors occurring in unusual locations. Morphological correlates are weak.

Anahtar Kelimeler: EBER, smooth muscle tumor, Ebstein-Barr Virus