FDG-PET SCAN AND SERUM NSE IN EARLY RESPONSE TO SIROLIMUS IN MALIGNANT PERIVASCULAR EPITHELIOID CELL TUMOR (PEComa)

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PEComas are rare tumors originally described by Liebow and Castleman. Standard chemotherapies for soft tissue sarcomas such as anthracyclines and ifosfamide have limited activity to PEComas. However, the inhibitors of mammalian target of rapamycin (mTOR), such as sirolimus have demonstrated significant activity. Although FDG-PET scan is useful for evaluation of malignant PEComas, little is known about the value in early response to mTOR inhibitor in PEComas. We here report a case of malignant PEComa of the uterus in a woman treated with oral mTOR inhibitor sirolimus. The FDG-PET scan and serum NSE levels were useful in the early evaluation of response to sirolimus.