Epitheloid Variant of Angiomyolipoma in a Patient with Tuberous Sclerosis.

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Abstract

Epitheloid angiomyolipoma of kidney is a type of Perivascular endothelial cell derived tumor with an aggressive behaviour with specific pathological, immunohistochemistrical and genetic characteristics. They can occur in a pure form or in assosciation with classical angiomyolipoma. It can be assosciated with tuberous sclerosis in 50% of cases. Our case is a possible case of tuberous sclerosis with epitheloid angiomyolipoma, hepatic angiomyolipoma and lymphangioleiomyomatosis with normal MRI brain and no cutaneous features. Radical nephrectomy with biopsy of hepatic lesion was performed. Histopathological examination revealed epitheloid variant with features of angiomyolipoma. It is six months post surgery and patient is doing well with no evidence of recurrence. Epitheloid angiomyolipoma is a rare malignancy with only 120 cases reported in literature.

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