

中文題目：雙側巨大的腎血管平滑肌脂肪瘤造成出血性休克和腸沾黏阻塞

英文題目：Tuberous sclerosis-associated bilateral huge renal angiomyolipomas caused bowel obstruction and hemorrhagic shock.

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Introduction

Renal angiomyolipoma (RAML) is a rare hamartoma in general population, but it is the most common renal presentation in tuberous sclerosis complex (TSC). We presented a case of TSC with bilateral huge RAMLs which caused hemorrhagic shock and severe colon adhesion and compression.

Case Presentation

A 39 year-old female had previously been diagnosed with TSC and took Everolimus for bilateral renal RAMLs since one year ago. She presented to Kaohsiung Medical University Hospital with a chief complaint of fever, anorexia, nausea and vomiting of three-day duration. Laboratory examinations yielded leukocytosis, high C-reactive protein and renal function impairment. The abdominal computed tomography showed mild progression of bilateral multiple huge RAMLs (longest diameter: 33 cm) which compressed bowel. Angiography revealed RAMLs bleeding which was treated by transarterial embolization. Under the reason of RAMLs complications (bleeding and bowel compression), she received bilateral nephrectomy. During surgery, the operator found severe adhesion of colon by RAMLs so subtotal colectomy (from terminal ileum to sigmoid colon) and side to side ileocolostomy was performed. Histological examination indicated RAMLs composed of well-differentiated vessels, smooth muscle and adipose tissue. To date, the patient recovered well and received regular HD.

Discussion

RAML is a benign tumor with composition of smooth-muscle-like cells, adipocyte-like cells and epithelioid cells. It could grow over time with complication of hemorrhage, organ compression by mass effect, chronic renal disease, anemia and hypertension. Renal lesions are a significant source of morbidity and mortality in TSC. For acute bleeding, embolization followed by corticosteroids is recommended. We presented a rare case of TSC-associated RAMLs which were 33 cm in largest diameter and caused ileus and hemorrhagic shock.