Angiomyolipoma in the Transplanted Kidney Presenting with Hematuria- Rare Case Report

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ABSTRACT

Renal angiomyolipomas (AML) are a type of benign renal neoplasm composed of vascular, smooth muscle, and fat elements, usually exhibiting characteristic radiographic appearances. Although benign, they carry the risk of spontaneous hemorrhage, which can potentially be fatal. AML has an incidence of 0.3% to 3%, hence reports of this condition are relatively scarce; even more so is the unique presentation of AML in the transplanted kidney. Predictably, there have been few reports of this uncommon condition in the medical literature. Herein, we report a very rare case of an isolated AML in a transplanted kidney that developed more than a decade after uneventful renal transplantation.

A 56-year-old female had a kidney transplant in early 2010, but subsequently developed chronic graft rejection. She presented with haematuria, and upon further assessment, a fairly large SOL in the kidney allograft was revealed by imaging including abdominal ultrasound, a 99mTc-DMSA scan, and a CT scan. She had graft nephrectomy in early 2022 and is still on haemodialysis.

Keywords: Transplanted kidney, Angiomyolipoma, Post-transplant tumour, 99mTc-DMSA scan.

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INTRODUCTION

Renal angiomyolipomas (AML) are a type of benign renal neoplasm that originates from the mesenchymal tissue, composed of vascular, smooth muscle, and fat elements (1). It was first described by Morgan et al. in 1951 (2). AML has an incidence of 0.3% to 3% (3), hence reports of this condition are relatively scarce. The majority of AMLs are sporadic and isolated (80%) and are typically identified in adults in the 4th to 5th decade of life (mean age of presentation: 43 years), with a female predilection (F: M of 2-4:1) (4, 5). AML might be associated with tuberous sclerosis, on the other hand, can be multiple, bilateral, and affect mostly younger patients (6). Although benign, they carry the risk of spontaneous hemorrhage, which can potentially be fatal. The first clinical presentation is often rupture and massive internal hemorrhage (7).

CASE REPORT

A 56-year-old female with chronic renal failure (CRF) had history of a kidney transplant in March, 2010 at a tertiary care hospital outside Bangladesh. Her post-transplant period was uneventful for a long period until she developed chronic graft rejection, as evidenced by progressively worsening eGFR levels. Consequently, she was put on maintenance hemodialysis in January 2021. On February 2022, she presented with hematuria which warranted further investigations. As such, an abdominal ultrasound scan, and later a CT scan were requested. The ultrasound scan revealed a fairly large, oval shaped, uniformly echogenic soft tissue lesion in the left iliac fossa region involving the infero-medial aspect of the transplant kidney measuring 5.8 X 4.0X 4.5 cm. Rest of the transplant kidney showed slightly raised cortical echogenicity. No pelvi-calyceal dilatation, features of hydronephrosis or calculus was noted. Color Doppler showed hypovascularity of the lesion.

Later, non-contrast and contrast-enhanced abdominal CT scan were performed which confirmed the ultrasound findings. It revealed a large soft tissue mass (71 x 60 x 55 mm) in the postero-infero-medial aspect of the transplant kidney of mixed attenuation (+15 to -91 HU CT attenuation), implying predominantly fatty tissue.

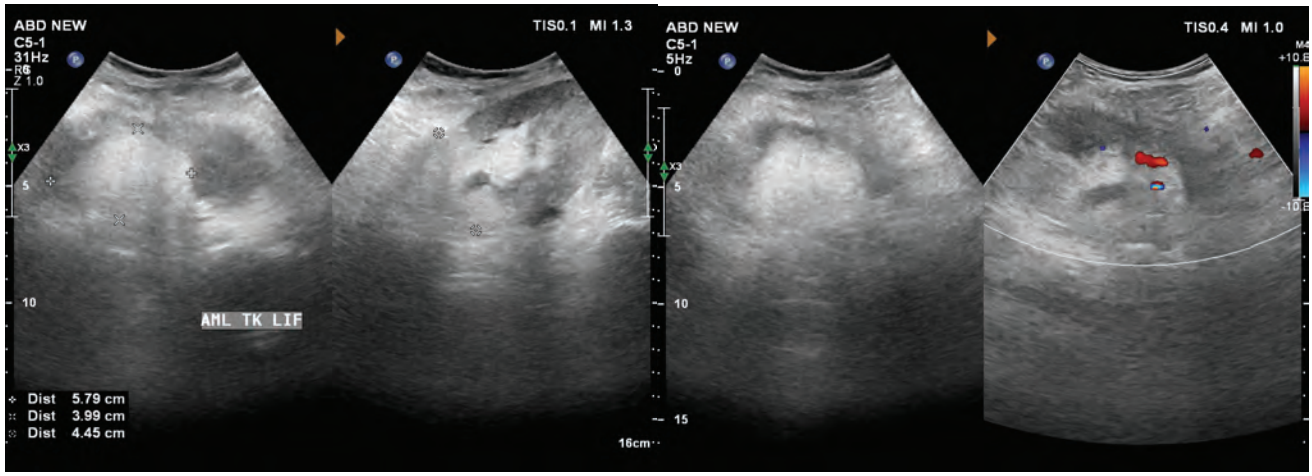

Figure 1: Ultrasound images of the transplant kidney showing an oval, echogenic, soft tissue lesion (left) with colour Doppler showing hypovascularity (right).

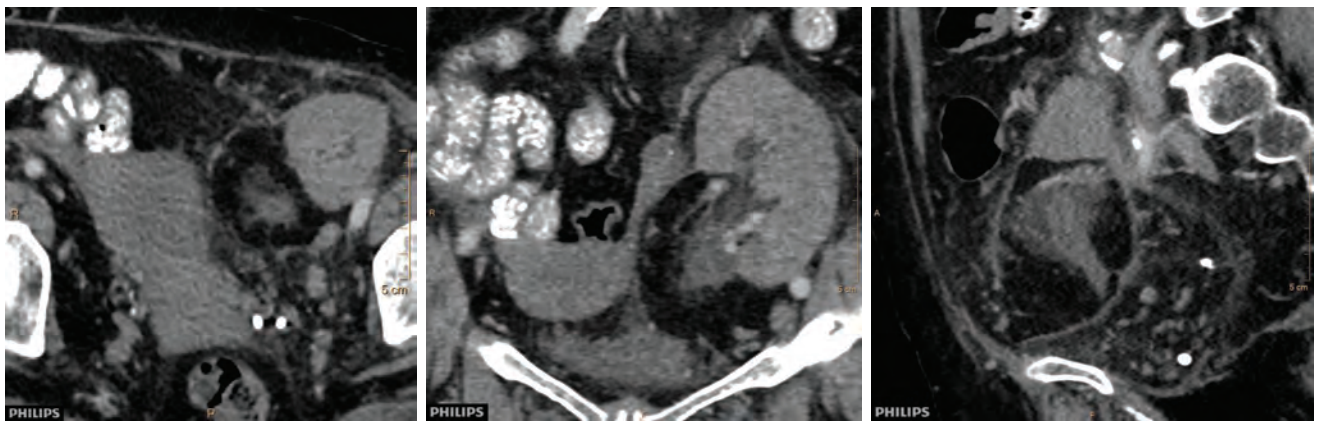

Figure. 2: CT images of the lesion in the axial, cornal and sagittal plane respectively (clockwise) showing a large soft tissue mass in the transplant kidney.

Post-contrast images showed mild enhancement at medial aspect of the lesion (up to 30 HU) except fatty tissue. In addition, the transplant kidney showed mild enhancement in early and portal phases (up to 60 HU); however, no excretion or hydronephrosis was seen. Both native kidneys were atrophic and showed mild enhancement in arterial phase; no excretion was noted there as well. A week later, a 99mTc-DMSA scan was performed which showed decreased radiotracer uptake in the transplant kidney, as well as a photopenic area in its infero-medial aspect.

Eventually, she underwent graft nephrectomy in March, 2022 and her hematuria was resolved. Histopathology revealed a predominantly fatty tissue mass with mildly fibrotic stroma and thick walled blood vessels. Post-operative period was uneventful, and she continues to be on maintenance hemodialysis.

DISCUSSION

Angiomyolipoma is a benign tumor that may occur in the kidney sporadically or as a part of genetic disorders such as tuberous sclerosis or lymphangioleiomyomatosis (1). The incidence of AML in the kidney allograft is very rare, and most of the reports of AML in kidney allografts are of donor origin.

Post-transplant tumors can originate from 3 sources: the kidney transplant donor, recurrence of previous tumor, and de novo development (8). We believe this patient developed de novo AML since there was no evidence of tumor in the received allograft. The risk factors for development of tumors after kidney transplantation are not clear. However, sun exposure, long-term immunosuppressive therapy, concomitant viral infection, advanced age, and longer pre-transplant dialysis periods have been described as risk factors of malignancies after

kidney transplantation (9).

Although tumors are treated with total allograft removal (10), partial nephrectomy has been applied based on the tumor size and location (11). In the case we report, the transplanted kidney was deemed non-functioning; hence, the decision for graft nephrectomy was made. AML imposes a risk of spontaneous hemorrhage, and may give rise to such drastic events as shock due to severe hemorrhage from rupture described as "Wunderlich Syndrome". Surgical excision or radiographic embolization remains the mainstay in management.

CONCLUSION

Despite the fact that renal transplantation has reduced mortality from end-stage renal disease (ESRD), complications still arise frequently. Even after undergoing successful kidney transplantation, there are many factors at play that predispose patients to additional medical challenges: AML being one of them, albeit very rare. Physicians should remain vigilant lest the rare ones should escape their watchful eyes and wreak havoc.

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