Case Report

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Renal Leiomyoma Associated with Tuberous Sclerosis

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Key Words

Renal leiomyoma
Tuberous sclerosis

Abstract

The renal lesions characteristic of tuberous sclerosis are angiomyolipoma and cysts, with the former considered to be more common. Other renal tumors are rarely associated with tuberous sclerosis. Here we present a tuberous sclerosis patient with a renal leiomyoma which was detected incidentally during the investigation of fever of unknown origin.

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Introduction

Tuberous sclerosis is an inherited autosomal dominant disease characterized by mental retardation, epilepsy, adenoma sebaceum, and hamartomas of the bone, brain, eyes, lungs, heart, and kidneys. Most of the renal tumors associated with tuberous sclerosis are angiomyolipomas (renal hamartomas). We report an extremely rare case of tuberous sclerosis associated with renal leiomyoma, a rare benign tumor arising from smooth muscle in the renal capsule, renal cortical vessels, or renal pelvis.

located in the upper pole of the left kidney. The mass showed a low echogenicity on ultrasonography and a low density on computed tomography with contrast enhancement (fig. 1). Left renal arteriography revealed that the mass contained numerous vessels and microaneurysms, while T2-weighted magnetic resonance imaging showed that it was of low intensity. Partial nephrectomy was performed under the diagnosis of a benign tumor causing left hydronephrosis and pyelonephritis. The resected specimen contained a homogeneous gray-white tumor, which was 6 cm in diameter (fig. 2). Histological examination of the entire tumor revealed a benign lesion composed of spindle cells arranged in interlacing fascicles without nuclear atypia or mitotic figures. There was no fatty component and none of the vascular changes which would suggest angiomyolipoma. The histological findings, including desmin staining (fig. 3), led to a diagnosis of typical leiomyoma.

Case Report

A 23-year-old man was referred to us because a left renal mass was detected during the investigation of fever of unknown origin. The patient had a history of surgery for a brain tumor

(subepithelial giant cell astrocytoma) and displayed many of the characteristic cutaneous features of tuberous sclerosis. Physical examination was otherwise normal and laboratory parameters were within normal limits except for leukocytosis. Intravenous pyelography showed deviation of the upper calyx and slight hydronephrosis caused by a mass

Discussion

Tuberous sclerosis is an inherited autosomal dominant disease characterized by mental retardation, epilepsy, adenoma sebaceum, and hamartomas of the bone, brain, eyes, lungs, heart, and kidneys. The incidence of renal involvement in tuberous sclerosis has varied from 40 to

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Fig. 1. Computed tomography shows a homogeneous mass containing a low-density area in the left kidney.

Fig. 2. Cut surface of the tumor. The lesion is a homogeneous and gray-white mass with a diameter of 6 cm.

Fig. 3. Histological section showing bundles of smooth muscle cells. There is no fatty tissue and none of the vascular changes suggestive of angiomyolipoma. a HE. $\times 100$. b The tumor is positive for desmin staining. $\times 100$.

80% in previous reports. The characteristic renal lesions are angiomyolipoma and cysts, with the former considered to be more common. However, other renal tumors associated with tuberous sclerosis have also been reported recently. The majority of the lesions were renal cell carcinomas [1], while the others were Wilm's tumor [2], leiomyosarcoma [1], liposarcoma, and oncocytoma [1]. As far as we could determine, however, renal leiomyoma has not previously been reported in association with tuberous sclerosis.

Renal leiomyoma is a rare benign tumor that arises from smooth muscle in the renal capsule, renal cortical vessels, or renal pelvis [3]. The majority of renal leiomyomas are detected incidentally at autopsy and surgical treatment has only been reported in a few cases. Renal leiomyoma has no characteristic signs on imaging examinations, so preoperative diagnosis is very difficult. Thus, a renal leiomyoma associated with tuberous sclerosis is likely to be misdiagnosed as angiomyolipoma. With the development and increasing use of various imaging techniques, the detection and preoperative diagnosis of renal leiomyoma, associated with tuberous sclerosis is expected to increase.

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