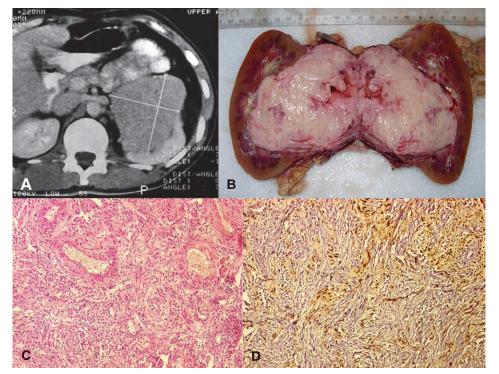
Pictorial Urology

Leiomyomatous Angiomyolipoma of Kidney

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A 24-year-old man was diagnosed with a mass in the left kidney on abdominal computed tomography (Figure A). Nephrectomy specimen showed a smooth-surfaced tumor, sized $9 \times 6 \times 6$ cm, attached to the hilar area, which was homogenous, well-circumscribed, grayish white, and solid, compressing the renal parenchyma, but not invading it (Figure B). Microscopic examination showed spindle cells arranged in whorls, rich in blood vessels (Figure C). On immunohistochemistry, the tumor cells expressed smooth muscle actin, vimentin, desmin, and HMB-45 (Figure D). A diagnosis of leiomyomatous angiomyolipoma was made.

Leiomyomatous angiomyolipoma is a benign mesenchymal tumor which is typically composed of a mixture of vessels, smooth muscle, and fat. In addition, it contains clear or pale epithelioid cells, currently known as the perivascular epithelioid cells,⁽¹⁾ which show positivity for HMB-45 in addition to expected smooth muscle markers. On computed tomography, leiomyomatous angiomyolipoma shows a characteristic appearance because of abundant adipose tissue, the lack of which pose a problem. Microscopically, leiomyomatous angiomyolipomas with predominant spindle cells of smooth muscle type may look like leiomyomas, leiomyosarcoma, or gastrointestinal stromal tumors.⁽²⁾ The presence of perivascular epithelioid cells in tumoral tissue, which shows reactivity to melanocytic markers such as HMB-45 in addition to smooth muscle markers, clinches the diagnosis.⁽¹⁾

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