Incidental detection of retroperitoneal lymphangioleiomyomatosis (LAM) - CT and MRI findings with relevance to the urologist

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A 48 year old lady presented with blunt abdominal trauma following a road traffic accident. She had no hematuria or other injuries. Vitals were normal. Examination was significant for left loin tenderness. Ultrasound was suspicious for retroperitoneal fluid. Computed tomography revealed normal abdominal viscera but multiple diffuse low attenuating cystic retroperitoneal masses showing peripheral enhancement (Figure-1). Lung segments showed multiple thin walled parenchymal cysts bilaterally (Figure-2, arrows). T2-weighted MRI revealed conglomerated cystic masses in the paraaortic and parailliac regions (Figure-3, arrows) suggesting multiple lymphangiomyomas.

A diagnosis of lymphangioleiomyomatosis (LAM) was made. She was managed conservatively and was well at 6 months.

Lymphangioleiomyomatosis is a rare hamartomatous lesion affecting women of reproductive age who manifest with respiratory symptoms secondary to multiple pulmonary cysts (1). Abdominal manifestations include angiomyolipomas (20–54%), lymphangioleiomyoma (20%) and lymphadenopathy (40%) (1). The disease is slowly progressive and results in respiratory failure and death in 31–62% cases at 8-10 years (1). The importance of this entity to the urologist is its association with angio myolipoma and tuberous sclerosis. Though clinical pulmonary manifestations are rare (2-3%)
in tuberous sclerosis, morphological changes similar to LAM are seen in a much larger percentage [2].

REFERENCES


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