"Tram-line" Calcifications in Granulomatosis with Polyangiitis

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INTRODUCTION

Nephrocalcinosis consists of deposition of calcium in the renal parenchyma. Renal cortical calcification is a rare entity in comparison to calcium deposits in the medulla and is seen only in a handful of pathologies with corresponding characteristic patterns on imaging. Thin linear calcifications may get deposited in the peripheral renal cortex suggestive of cortical necrosis due to a vascular insult (vasculitis), and rarely due to glomerulonephritis. This pattern of calcification has also been referred to as the "tramline" or "railroad track" sign.

Keywords: Granulomatosis with polyangiitis, Renal calcification, Tram line, Renal failure

CASE REPORT

A 36-year-old male with end-stage renal disease secondary to GPA presented to the hospital for renal retransplant evaluation. He was diagnosed with GPA at the age of 12 and had undergone renal transplant twice in the past, with recurrent failures.

Computerized tomography (CT) scans of the abdomen and pelvis were performed which showed extensive vascular and cortical rim calcifications in the native kidneys [Figure 1a and b]. This was thought to be secondary to chronic renal disease (glomerulonephritis) and polyangiitis. This was accompanied by vascular calcification throughout the abdomen and pelvis.

Ultrasound of the abdomen [Figure 1c] revealed rim calcified atrophic native kidneys with some cysts (likely acquired kidney cystic disease). Also seen were several scattered areas of calcifications in the renal hilum corresponding to vascular calcifications, better seen on CT.
Renal calcification is typically classified into two groups, namely (i) nephrolithiasis where there are calcified deposits seen in the collecting system of the kidneys and (ii) nephrocalcinosis involving the renal parenchyma.

Randall’s plaque is microscopic calcium deposits in the interstitial tissue of the renal papilla. These plaques are thought to serve as a nidus for the formation of urinary stones. Large plaques are characteristic for the formation of idiopathic calcium oxalate stones.

The most common causes of cortical calcification are renal cortical necrosis and chronic glomerulonephritis, \cite{5-4} Other causes may include trauma, primary or secondary oxalosis, Alport syndrome, chronic pyelonephritis, and nephrotoxic drugs such as amphotericin B. Alport syndrome is a genetic disorder characterized by glomerulonephritis, end-stage kidney disease, and hearing loss. This condition typically demonstrates cortical calcifications with sparing in the medullary pyramids.

Cortical calcifications typically suggest necrosis of underlying parenchyma as opposed to medullary calcifications where the underlying renal parenchyma is normal.

On CT imaging, three distinct patterns can be appreciated although none is specific to any etiologies: (i) Most commonly, a thin peripheral rim or band of calcification often with extension into the septal cortex is seen; (ii) two thin calcified tracks running in parallel (tram-line) lines. These may be often non-continuous indicating the relatively more patchy distribution of the cortical necrosis; and (iii) a third and very rare type is multiple punctate calcifications distributed randomly in the renal cortex representing necrotic calcified cortical glomeruli and tubules. \cite{5}

GPA (previously known as Wegener granulomatosis) is an idiopathic necrotizing vasculitis characterized by inflammation and necrosis of small- to medium-sized blood vessels. Although the pathogenesis of GPA is not clear, autoimmune response is thought to be the possible etiology in the development of the disease. Antineutrophil cytoplasmic antibodies (ANCAs) are serum autoantibodies directed against proteins present in neutrophils and are the serological markers for small vessel vasculitis. \cite{6}

GPA usually affects the upper and lower respiratory tract, but a generalized form of the disease can affect extrapulmonary organs such as the kidney, skin, or nervous system. \cite{7}

Most common clinical manifestation of the GPA is in the upper airway. Other organs that are rarely involved are the eyes, skin, joints, nervous system, and kidneys. Renal involvement with GPA results in abnormal renal function secondary to glomerulonephritis and results in red cell casts in urine. Renal failure may occur in cases with renal involvement if left untreated and this eventually results in a rapid fatal course. Renal pathology usually demonstrates focal segmental necrotizing glomerulonephritis and rarely necrotizing granulomas.

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Ultrasound typically shows non-specific increased echogenicity of the renal parenchyma without specific Doppler abnormalities and rarely one or more infiltrative mass-like lesions.\(^3\) Dense peripheral calcification in the kidneys may limit visualization of the atrophic renal cortex due to the posterior acoustic shadowing.

There are no definitive diagnostic criteria for GPA. Diagnosis is based on a combination of clinical manifestations, positive ANCA serology, and histological evidence of necrotizing vasculitis, necrotizing glomerulonephritis, or granulomatous inflammation on biopsy of the organ involved. The presence of extensive vascular calcifications and renal cortical (tram line) calcification in the absence of other etiologies of renal and vascular calcifications should raise suspicion for GPA.

**CONCLUSION**

Renal calcification can involve the renal parenchyma as well as the collecting ducts (calculi). Cortical deposits are less common and can be associated with the pathology depending on the characteristic patterns. Tram-line pattern of calcification in association with renal failure can be seen with GPA. This is often associated with extensive vascular calcifications throughout the body. Diagnosis can usually be confirmed with serology.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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