2016 USCAP-SUP J. Allan Tucker Award Winner

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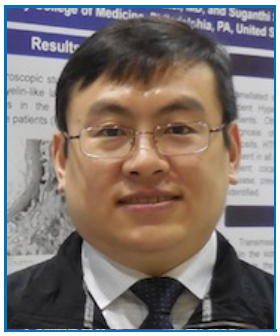

***Abstract:* Drug Induced Phospholipidosis in Kidney: Ultrastructural Features.**

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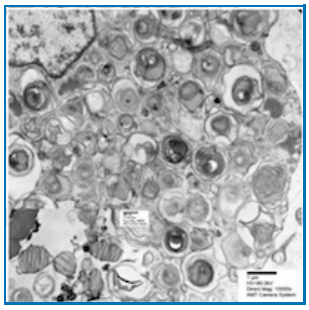
Background: Drug induced phospholipidosis (DIPL) is considered to be an acquired lysosomal storage disorder due to an adaptive response to cationic amphiphilic drugs (CADs) and toxic effects of gentamycin. DIPL is characterized by excessive accumulation of phospholipids in different organs mimicking the myelin figures (myelinoid bodies). In the kidney, Fabry disease, metabolic diseases and DIPL are known to cause myelin figures. Among the over 50 marketed CADs, only a few have relevance in the kidney. Thus, renal DIPL is rare and could lead to diagnostic difficulty, particularly due to the lack of specific staining on light microscopy. Renal DIPL identified by electron microscopy (EM) is the diagnostic hallmark.



Design: In our EM pathology database (2008-2015), three native kidney biopsies with DIPL were identified. There were 3 adults (1 male: 2 females) with an age range from 54 to 74 years. The histological sections, toluidine blue stained thick sections and electron micrographs were reviewed.

Results: Ultrastructural studies showed characteristic cytosolic inclusions consisting of myelin- like lamellated structures resembling zebra bodies or lamellar bodies in the expanded podocyte cytoplasm and Bowman's space in all three patients.

Lamellated structures were present in the tubular epithelium in one patient. Hypersensitivity-type tubulo-interstitial nephritis was present in two patients. Other significant coexisting pathologic diagnoses include recent diagnosis of SLE, membranoproliferative glomerulonephritis with C3 deposits, hypertensive and diabetic nephropathy. Multiple drug intake history was present in all patients, cocaine abuse in two of them and azithromycin in one patient; cocaine and azithromycin are CADs. No evidence of clinical Fabry disease, previous metabolic inclusion diseases or chloroquine intake was identified.

Conclusions: Ultrastructural discovery alone of DIPL has been approved to be the essential criteria in diagnosing this rare disease. In the kidney, diagnostic myelin figures are located within the lysosomes of the podocytes and the tubular epithelium. Pathologists should be aware of these inclusions, caused by pharmacological/recreational agents, in the absence of clinical Fabry disease.

Category: Techniques (including Ultrastructure)