AN UNCOMMON CAUSE OF END STAGE RENAL DISEASE IN A CHILD

- A case of oxalosis

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INTRODUCTION

- Oxalosis is a rare metabolic disease

- It can present in
  - Neonatal period with severe renal failure and B/L nephrocalcinosis
  - Older children as recurrent stones leading on to ESRD in adolescence
  - Older individuals as renal stone disease with poor progression to ESRD
  - Asymptomatic siblings
  - Transplanted patients

- In 2004 a 6 yr, Female ,Non consanguinous
- H/o recurrent episodes of painful micturition, recurrent hematuria
- No features of UTI
- Investigations: RFT – normal , Urine Calcium – increased, USG – B/L renal calculi
- Treatment : Alkali therapy , Diuretics
- Asymptomatic hence parents withdrew drugs

- Dec 2009
- H/o early morning facial puffiness progressed to generalised edema
- No recognisable oliguria
- Investigations:
  - Hb – 4.1%
  - Urea – 148 mg/dl
  - Serum Creatinine – 17.5 mg/dl
  - Na – 132 meq/L
  - K -6 meq/L
  - Cl-95 meq/L
  - HCO3- 12 meq/L

- Serum Ca -8.1mg/dl
- Serum Phosphorus -9.1mg/dl
- Urine analysis - Alb ++, RBC-2, urate crystals
- Spot urine – Ca – 5,Cr-21
- USG: B/L contracted kidneys, Rt renal calculi, ascites and pleural effusion

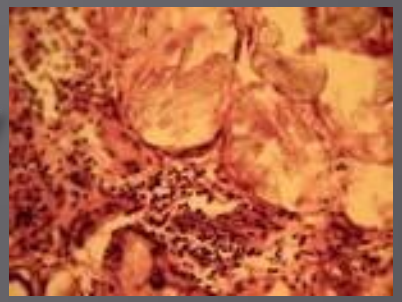
PROGRESS

- On the basis of
  - Elevated serum creatinine
  - Borderline hyperkalemia
  - Low serum bicarbonate
  - Hyperphosphatemia
  - Contracted kidneys
- Child started on hemodialysis
- After 15 HD renal functions did not improve.
- RSD rapidly progressing to ESRD – possibly oxalosis
- Nephrectomy done to confirm the diagnosis.

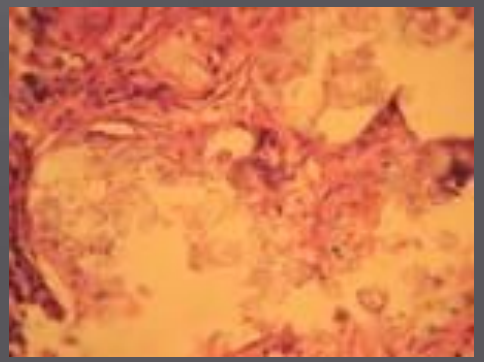
GROSS MORPHOLOGY OF THE SPECIMEN

- Rt. Nephrectomy sent; small contracted kidney with small pedicle.
- c/s: severe distortion of renal parenchyma with dilated pelvicalyceal system with numerous stones.
- Narrowed cortex.

HISTOPATHOLOGY



- Extensive crystalline material deposits in tubules, interstitium,occasional glomeruli.
- Characteristic radial striations.
- Peri glomerular fibrosis; few hyalinised glomeruli.Interstitial lymphoid aggregate.
- Biochemical analysis of deposit confirms oxalate.
- Dx: Tubulointerstitial nephritis with nephrolithiasis and renal oxalosis.



DIAGNOSIS & TREATMENT

- Prenatal –
  - DNA Analysis
- Preventive measure – Liver transplant
- Disease - Liver-Kidney transplant

TREATMENT

- Asymptomatic –
- Increased urinary volume (3-4 L/day)
- Low salt diet
- Low oxalate containing diet
- Diuretics
- Alkali therapy
- High doses of pyridoxine (vitamin B6):
- Neutralphosphates and citrate

WHY THIS PRESENTATION?

- Among RSD oxalosis has rapid onset of renal failure and worst prognosis
- 90% - calcium stones
- Lab facility to diagnose oxalosis is not standardised or easily available
- Any child with renal stones should be evaluated for oxalosis or monitored for progression
- F/H of RSD with ESRD / Death in young people - possibility of oxalosis to be considered.

THANK YOU