

Must-Know Board “Zebras”

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- Nephrology Fellowship at the BWH/MGH Joint Nephrology Program
- MPH through the Harvard School of Public Health
- Instructor of Medicine at HMS
 - Clinical Interests: Community Nephrology, Transition to Dialysis

Disclosures

- Nothing to disclose

Objectives

- To introduce cases not commonly seen in general nephrology practice but that may come up on the boards
- To expand general nephrology knowledge

Case #1

- 24 yo Kuwaiti male with CKD due to reflux nephropathy from posterior urethral valves s/p multiple procedures, requiring self-catheterization
- Valves first resected at age 15 days, then revised at 17 years
- Progressed to ESRD, started on HD in Kuwait via AVF, comes to BWH 4 months later for transplant evaluation
- Labs at BWH after missing several HD sessions while traveling: BUN 90, Cr 12.7, CO2 12, Calcium 7.8, Phosphate 6.6, Hgb 14
- Medications: Metoprolol 50mg daily, Calcium carbonate 600mg daily, Sevelamer hydrochloride 800mg qac, iron pills, Darbepoetin, Nifedipine
- HD prescription: HD 3 hours, Opti 180, K scale, 2.5 Ca, 35 HCO₃, Qb 400, Qd 800
- Minimal UF needed due to persistent 2-3L/day of urine

Case #1

Laboratory trend on dialysis at BWH:

	10/7	10/11	10/18	10/20	10/25	10/29	10/29- post HD
Potassium	4.6	4.5	5.1	4.8	5.3	4.6	3.3
BUN	90	50	76	56	62	53	15
Cr	12.7	8.9	9.7	8.2	9.8	8.4	3.2
HCO3	12	18	15	16	12	18	27
Phosphate	6.6	4.7	7.0	6.9	6.6		
Urine pH					8.0	8.0	8.0

URR 10/11- 74%

URR 10/15 with 3 hours and 15 minutes- 72%

ABG: 7.35/30/128/17

Case #1

What is the most likely reason for this patient's persistent acidosis?

- a) Inadequate dialysis due to refusal to dialyze longer than 3.25 hours
- b) RTA due to reflux nephropathy
- c) Dietary indiscretion, as evidenced by persistent hyperphosphatemia
- d) Dialysis machine did not add the bicarbonate correctly
- e) Sevelamer hydrochloride decreases bicarbonate concentration

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Other Answers

- Inadequate dialysis- Presumably the URR would be lower for this degree of acidosis, should see other signs of under-dialysis in the labs
- Dietary indiscretion- While possible, would be hard to drive bicarbonate this low with diet alone
- Dialysis machine did not add bicarbonate correctly- There are case reports of this problem, but would be expected to affect all patients on one specific machine, and post-dialysis bicarbonate would not be 28 if this were the case
- Sevelamer HCl- This can cause a mild acidosis at very high doses, but unlikely at 800mg qac as the effect is dose-dependent; $\text{HCO}_3^- < 15$ seen only in those with $> 6000\text{mg}$ per day in one study¹; fix for this would be to use sevelamer carbonate

¹Oka et al, *Cardiovasc Hematol Disord Drug Targets*, Volume 8, 2008

Reflux and the Kidney

- Reflux nephropathy develops in 30-60% of children with vesicoureteral reflux (VUR)
- Posterior urethral valves (PUV) are the most common cause
- Predominates in girls, but more severe in boys
- Causes scar formation in kidneys including caliceal deformity and thinning of the renal parenchyma
- Tubulointerstitial damage with atrophy, tubular necrosis, and interstitial mononuclear infiltrate

Reflux-Induced Distal RTA

- Distal tubular damage appears to precede glomerular disease, impairing distal acidification
- Children tend to have hypokalemia with high TTKG as well
- Often persists for years after treatment of the obstruction

DRTA Profile in Children with PUV at Time of Surgery

Patient	Arterial pH	Acidification Done	Serum HCO ₃ after acidification	Serum Potassium	Urine pH	Urine Anion Gap
1	7.39	Yes	8	5.6	5.6	Positive
2	7.38	Yes	7	5.8	5.7	Positive
3	7.22	No	12	5.9	5.8	Positive
4	7.35	Yes	11	6.2	6.0	Positive
5	7.24	No	11	5.8	5.8	Positive
6	7.37	Yes	7	3.3	6.1	Positive
7	7.33	Yes	13	4.3	6.0	Positive
8	7.36	Yes	10	3.0	5.8	Positive
9	7.26	No	10	4.4	5.9	Positive
10	7.35	Yes	6	4.8	5.6	Positive
11	7.38	Yes	11	3.3	5.8	Positive
12	7.22	No	7	4.8	5.9	Positive
13	7.26	No	13	3.1	5.6	Positive
14	7.24	No	10	3.2	5.9	Positive
15	7.36	Yes	9	3.1	6.0	Positive
16	7.41	Yes	9	2.8	6.1	Positive
17	7.38	Yes	12	3.9	5.0	Negative
18	7.36	Yes	6	4.3	5.0	Negative
19	7.38	Yes	9	4.2	5.2	Negative
20	7.34	Yes	11	4.2	5.2	Negative
21	7.36	Yes	10	4.9	5.1	Negative
22	7.42	Yes	8	3.8	5.1	Negative
23	7.21	No	13	2.9	5.9	Positive
24	7.38	Yes	14	4.6	5.1	Negative

Case Follow-Up

- Continued to be acidotic on dialysis
- Bilateral nephrectomies at the time of transplant to control acidosis

Case #2

- 77 yo male originally from the Azores with PMH of bladder cancer
- 8 months prior to admission, developed joint swelling and pain, ESR 59, prednisone started for presumptive PMR and symptoms improved
- 2 months PTA developed anasarca and rash, 5g proteinuria noted and serum albumin 1.8, CT to rule out malignancy associated with presumed membranous nephropathy revealed PE and left renal vein thrombosis
- No biopsy done due to anticoagulation needs, but prednisone increased to 60mg daily to treat membranous nephropathy, started lisinopril and warfarin
- Labs at the time revealed negative hepatitis serologies and anti-DNAse, normal C3 and C4, and SPEP without a monoclonal gammopathy and K/L ratio 1.06

Case #2

- Presents to an emergency room 2 months later with severe diarrhea and sepsis
- Found to have esophageal Candidiasis, E. Coli bacteremia and Enterococcal bacteremia
- Exam with massive anasarca, normotensive, otherwise unremarkable
- Albumin 0.8g/dL, Cr 0.5mg/dL
- Triglycerides 233mg/dL, HDL 15mg/dL, LDL 88mg/dL
- UA with 3+ blood and 3+ protein, Pr/Cr ~5g
- Urine microscopy with hyaline casts and occasional dysmorphic RBCs
- Renal u/s with 11.3 and 11.5cm kidneys
- ANCA and GBM both negative, rheumatoid factor <11, C3 68 (low), and C4 9 (low), cryoglobulins negative
- Prior to his subsequent death due to overwhelming sepsis, he was also found to have disseminated strongyloides and aspergillosis

Case #2

What is the most likely cause of the patient's renal presentation?

- a) Post-streptococcal GN from an undiagnosed strep infection
- b) AL Amyloidosis
- c) Strongyloides-associated membranous nephropathy
- d) Candidiasis in the kidney
- e) Renal Vein Thrombosis

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Other Answers

- Post-streptococcal GN- Rarely a nephrotic syndrome, usually a nephritic syndrome with rise in creatinine and hypertension, low C3 with normal C4, self-limited course
- AL Amyloidosis- Less common without abnormal light chains in the blood
- Candidiasis- Can cause microabscesses in the kidney, potentially papillary necrosis, rarely pyelonephritis, but nephrotic syndrome unlikely
- Renal Vein Thrombosis- Usually is the consequence of hypercoagulability in nephrotic syndrome, not the cause

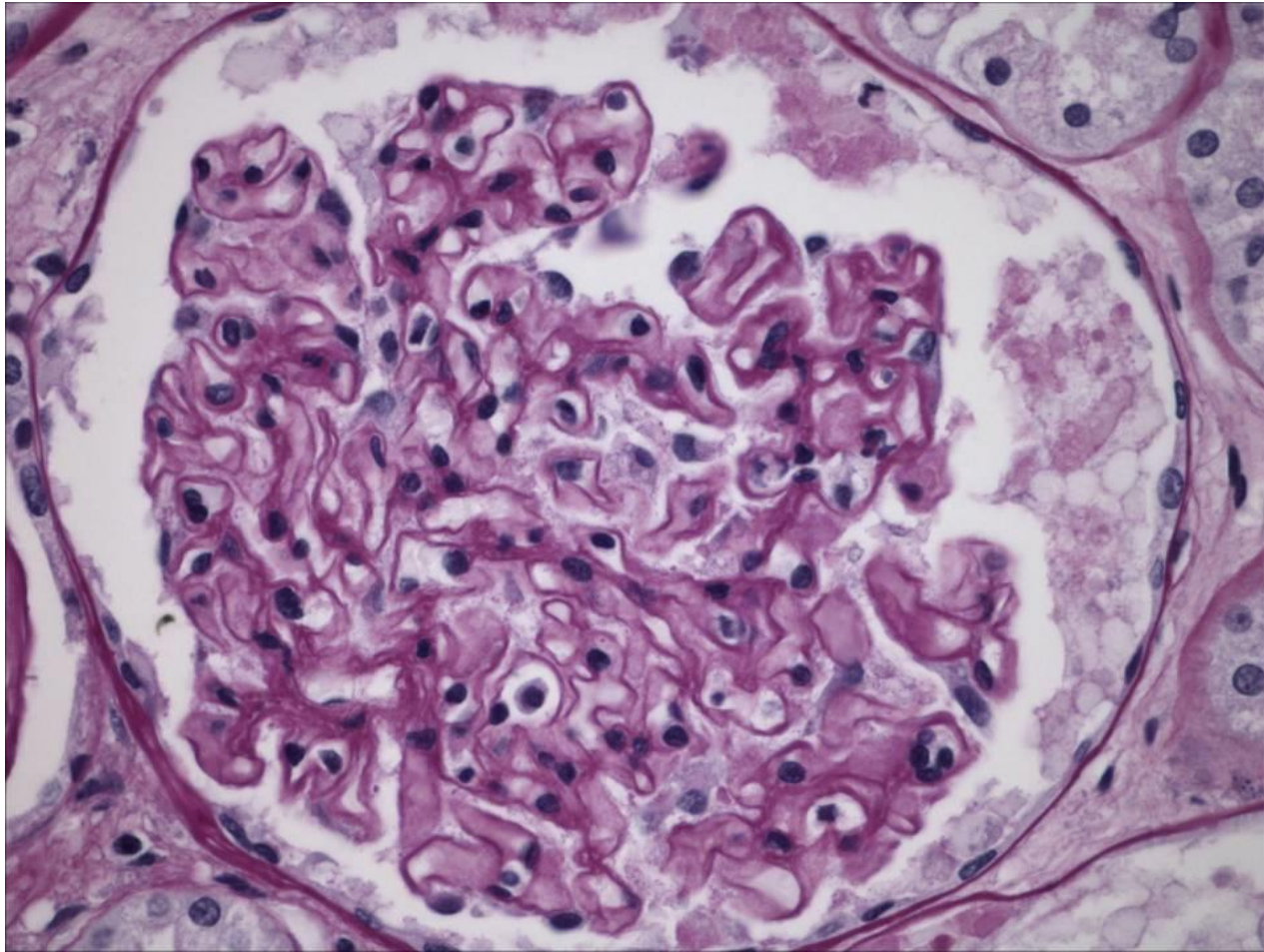
Secondary Membranous Nephropathy

- Most common causes are cancers, NSAIDs and other drugs, Lupus, Hepatitis B and C
- Also reports with some other chronic infections:
 - Schistosomiasis
 - Syphilis
 - Strongyloides
 - Malaria

Disseminated Strongyloides

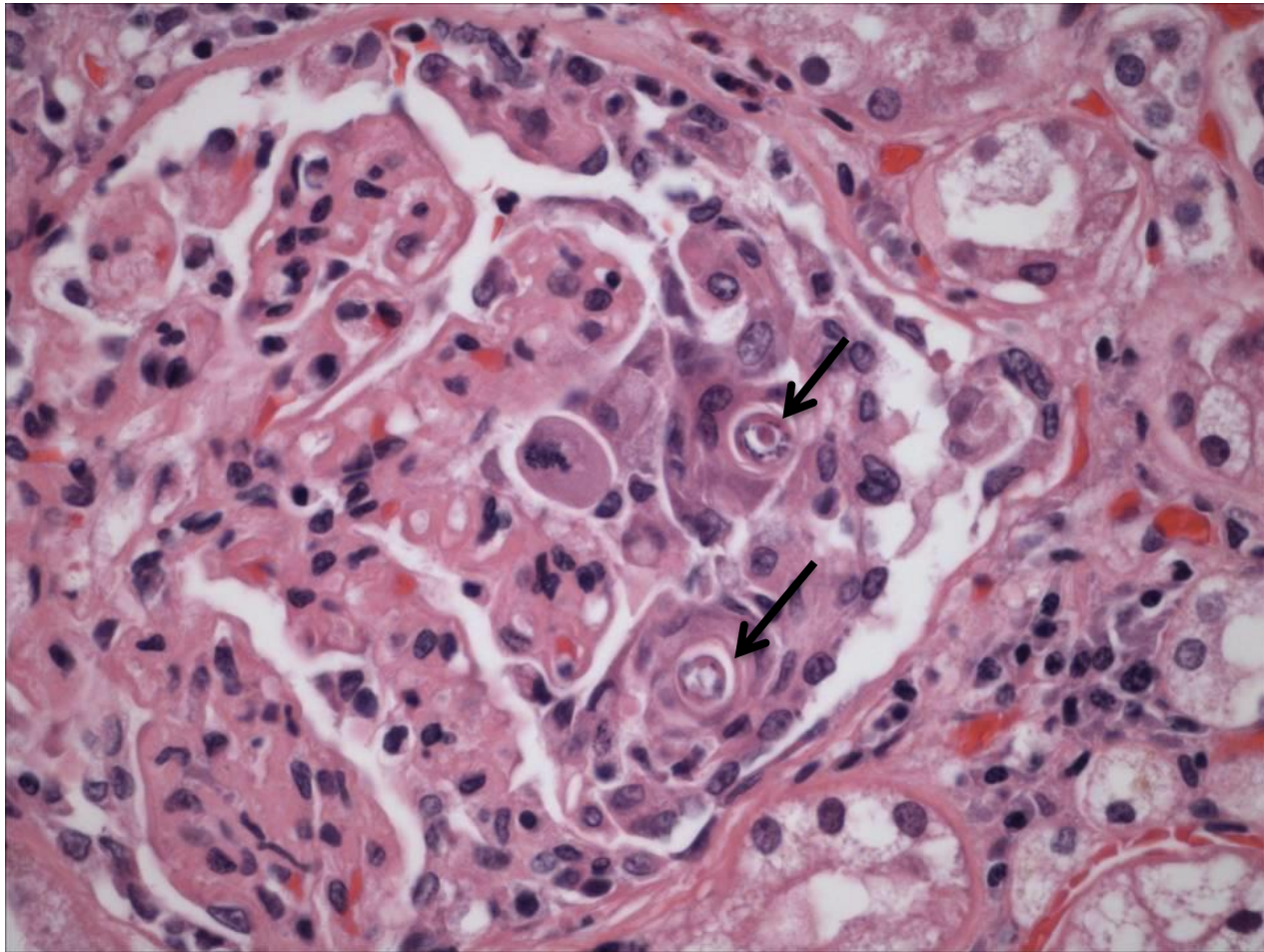
- Often in a patient with chronic strongyloides who then becomes immunosuppressed
- Common symptoms are skin rashes and itching, GI distress, meningitis, and worsening pulmonary function
- Can disseminate to almost any organ
- Dissemination to kidneys can cause minimal change disease or membranous nephropathy
- Case reports of improvement in the kidney disease with ivermectin to treat the infection

Kidney Histology



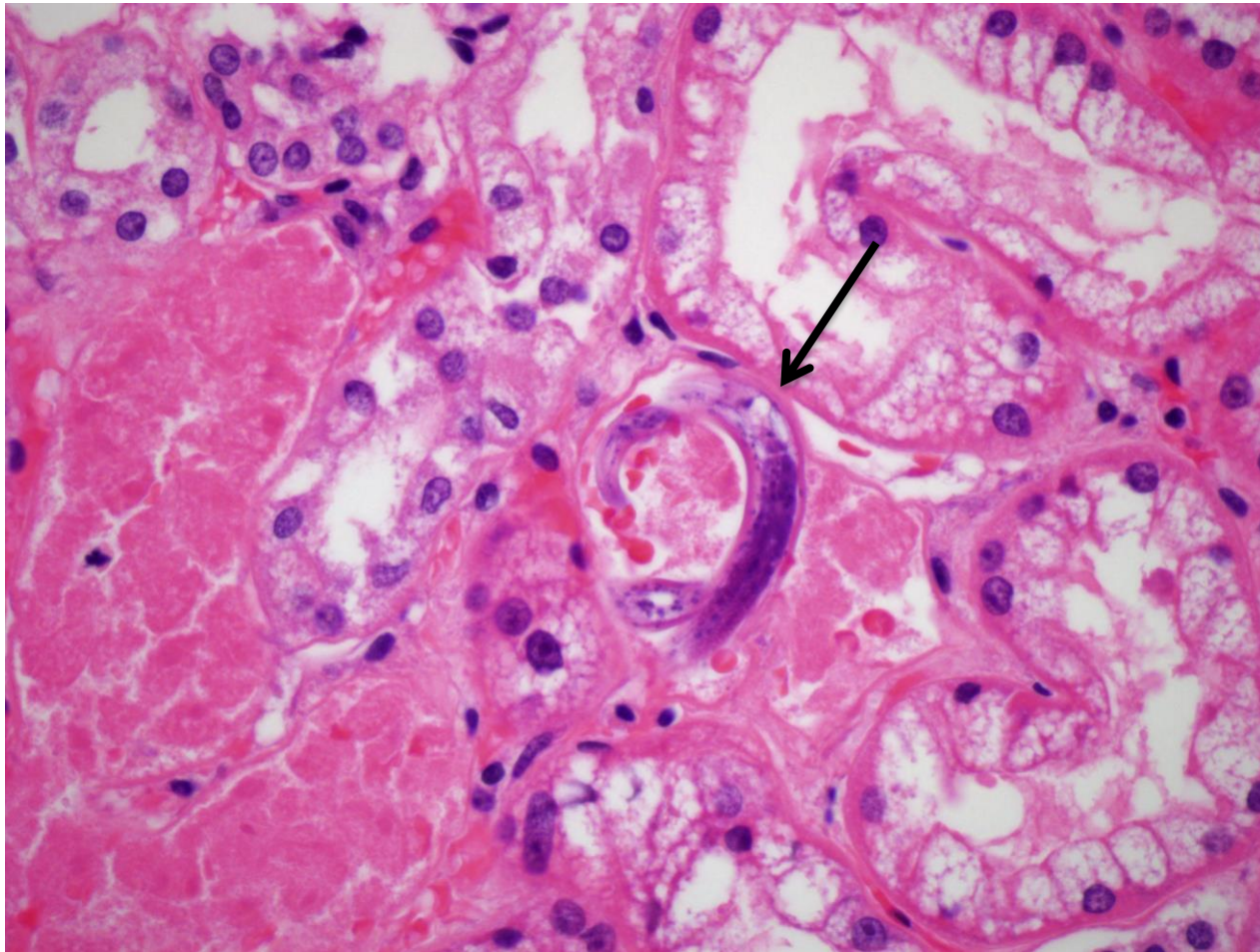
Membranous Nephropathy

Kidney Histology



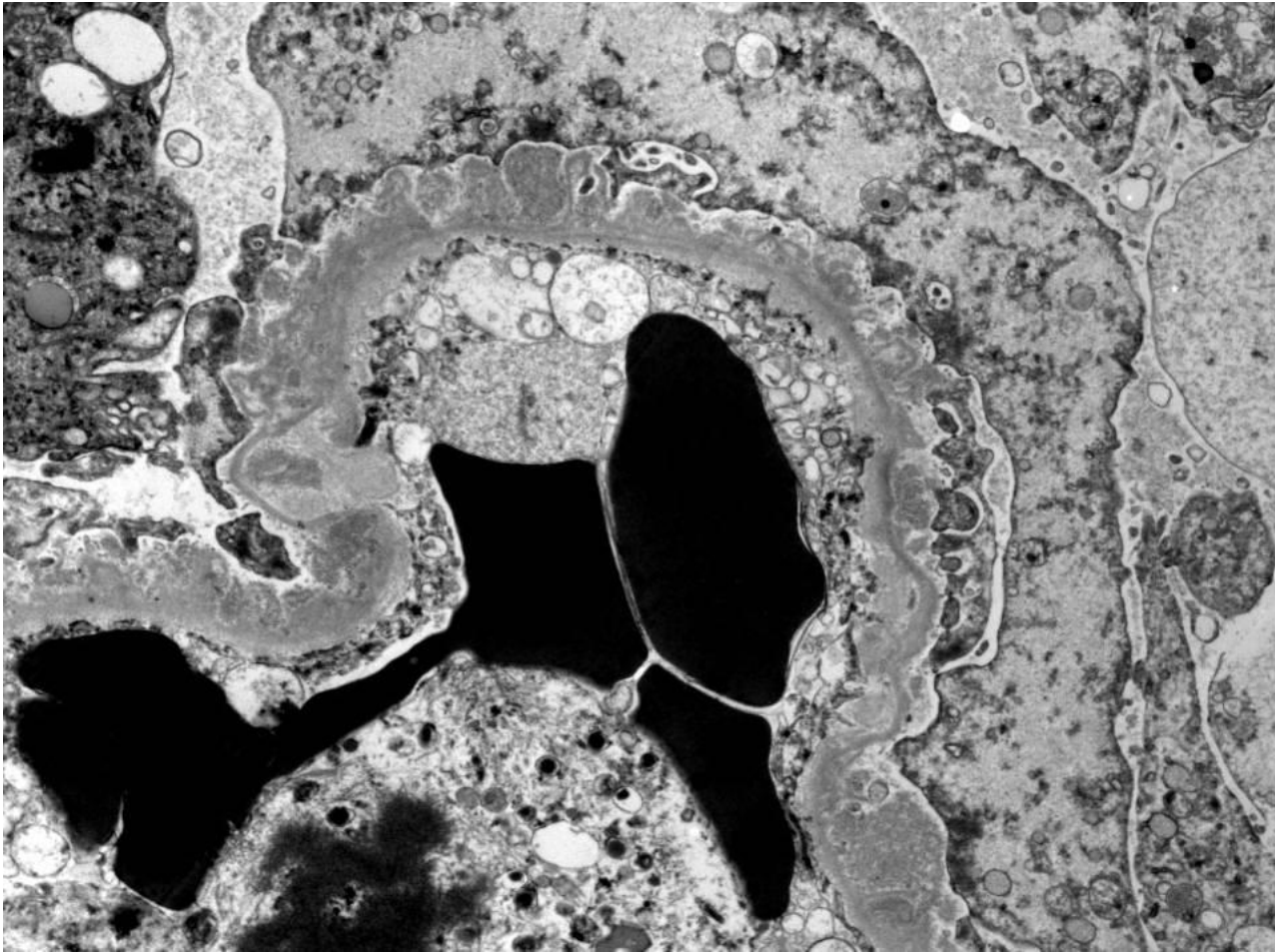
Strongyloides in the glomerulus

Kidney Histology



Strongyloides in a peritubular capillary

Kidney Histology



Subepithelial deposits

Take-Home Messages

- Patient likely exposed to strongyloides in Azores
- Parasites disseminated when started on steroids months prior to this presentation
- Worsened when steroids increased as the nephrotic syndrome developed
- Always consider testing for endemic infections before initiating or increasing immune suppression
- In theory, ivermectin may have been the better treatment here
- Patients on organ transplant lists from areas with endemic parasites should be screened pre-transplant due to expected addition of immunosuppression

Case #3

- 54 yo male with history of hypertension and hypokalemia with K as low as 2.8 for at least 7 years
- Noted to have Mg 1.1mg/dL with PCP, never checked prior, sent to nephrology clinic for evaluation and management
- PMH: HTN, hypokalemia, GERD, epicondylitis
- Meds on presentation to renal clinic: Magnesium gluconate 2000mg tid, KCl 60meq daily, Amlodipine 10mg daily, Omeprazole 20mg daily, Spironolactone 12.5mg daily
- Social history: Ex-smoker, 2 alcoholic drinks per week. Married, works full-time
- Family History: No known history of low K or Mg

Case #3

24-hour urine results PRIOR to renal visit:

Analyte	Value	Reference Range
Sodium	110 mmol	75-200 mmol/24h
Potassium	64 mmol	40-80 mmol/24h
Chloride	143 mmol	75-200 mmol/24h
Calcium	11 mg	50-400 mg/24h
Creatinine	1443 mg	800-1800 mg/24h
Protein	129 mg	0-165 mg/24h
Magnesium	Not tested	

Case #3

- Labs at nephrology clinic: Mg 1.6mg/dL, K 3.5mmol/L, HCO₃ 25mmol/L, Phosphate 2.7mg/dL, , Calcium 8.7mg/dL, Cr 0.8mg/dL
- PTH 31 pg/mL
- TTKG 9.7
- Aldosterone <1.6ng/mL
- Plasma Renin Activity <0.1ng/ml/hour
- Spot Urine Magnesium- undetectable

Case #3

What is the most likely reason for the hypomagnesemia and hypokalemia?

- a) Omeprazole
- b) Gitelman syndrome
- c) RTA
- d) Amlodipine
- e) Bartter syndrome

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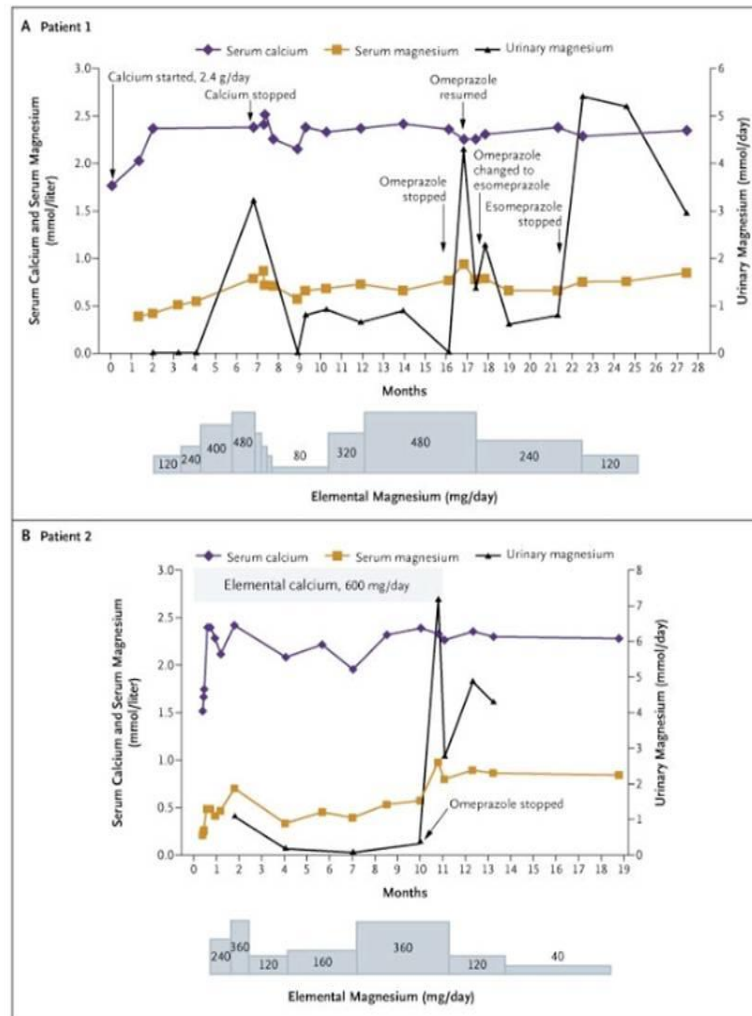
Other Answers

- Gitelman syndrome- Urine calcium is low which is consistent, but should show magnesium wasting
- RTA- Serum bicarbonate is normal, and an RTA would not be the cause of low magnesium that is not renally excreted, and is not typically a cause of severe hypomagnesemia
- Amlodipine- This is not a known cause of any of these findings
- Bartter syndrome- Should present with high urine calcium, high urine magnesium, usually presents in childhood

PPI-Induced Hypomagnesemia

- First reported in 2006¹
- Seen in patients with >1 year of PPI intake
- Seems to be related to any and all PPIs
- Not dose-related
- Often also associated with hypocalcemia and inappropriately low PTH

Magnesium and Calcium Trends on Two Patients



Potential Mechanisms- Low Magnesium

- Low urine magnesium levels suggest that low magnesium is likely due to GI loss from poor absorption
- Magnesium absorbed actively and passively in the GI tract
- Hypomagnesemia can be partially corrected by high-dose supplementation, suggesting that passive transport is not disrupted
- Thus, likely due to inhibition of active magnesium transport

Potential Mechanisms- Low K

- TTKG is often high in these patients, suggesting renal potassium wasting
- Aldosterone often low, so non-aldosterone mediated renal potassium wasting
- Normal magnesium levels are believed to inhibit the renal outer medullary potassium channel ROMK
- ROMK is an ATP-dependent K channel that will secrete reabsorbed K in the thick ascending limb
- Low serum magnesium levels may relieve this inhibition and thus increase K secretion

Potential Mechanisms- Low Ca and PTH

- Low magnesium is known to suppress PTH secretion, and parenteral magnesium administration will raise PTH levels
- Low magnesium also causes resistance of bone to PTH
- These findings most often seen with magnesium levels $<1.0\text{mg/dL}$
- Explain the serum and urinary calcium in these cases

Case #4

- 67 yo female with GIST s/p imatinib treatment, HTN, DM on insulin
- Hospitalized for low back pain resistant to naproxen, serum Na 114mmol/L
- Presumed SIADH due to NSAIDs and pain, fluid restricted, NSAIDs stopped, Na went up to 124mmol/L on d/c
- Readmitted 3 weeks later for weakness
- Admission chemistries: Na 120, K 2.9, HCO₃ 27, Cl 87, BUN 7, Cr 0.67, Glucose 79
- LFTs: Total protein 4.7, Albumin 2.2, T bili 19.5, D bili 13.6, ALT 151, AST 133, Alk Phos 1026, coags WNL
- Exam: BP 74/50 (improved to 120/77 with IVF), HR 80, dry mucous membranes, no LE edema, clear lungs, normal mentation
- Current medications: Famotidine 20mg daily, KCl 20meq daily, Glargine insulin, MVI daily, Phos-NaK 250mg bid, Ursodiol 300mg tid, Oxycodone prn

Case #4

Date	5-Apr	20-Jun	20-Aug	23-Sep	25-Sep	8-Oct
Parameter	GIST diagnosed	After stopping Imatinib	Outpatient	Hospitalized for back pain	After fluid Restriction	Current hospitalization
Serum sodium	137	130	126	114	124	120
Serum osmolarity	NA	295	NA	287	NA	291
Serum uric acid	NA	2.5	NA	NA	NA	NA
Serum potassium	3.9	3.4	3.5	2.9	3.6	2.9
Serum chloride	99	100	97	88	91	87
Serum creatinine	0.8	1.1	1.2	0.65	0.7	0.82
Urine sodium	NA	69	NA	97	NA	146 *after 1.5 L normal saline
Urine osmolarity	NA	411	NA	500	NA	612 *after 1.5L normal saline
Urine potassium	22	NA	NA	61	NA	48.3*after 1.5 L normal saline

Case #4

What is the reason for this patient's hyponatremia?

- a) Combination of liver disease and dehydration affecting intravascular volume
- b) SIADH due to the oxycodone and back pain
- c) Hyperglycemia causing sodium dilution
- d) Pseudohyponatremia
- e) Primary polydipsia

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Other Answers

- Normal serum osmolarity with a low serum sodium rules out causes other than pseudohyponatremia
- Volume depletion and liver disease- Urine sodium should be low in both of these cases
- SIADH- serum osmolarity should be low
- Hyperglycemia- Despite diabetes history, blood sugar is normal so would not cause sodium dilution, also often causes HIGH serum osmolarity
- Primary polydipsia- Urine osmolarity would be low

Pseudohyponatremia

- Artificially low plasma sodium
- Normal serum osmolarity
- Potential causes:
 - Hyperlipidemia
 - Hyperproteinemia
 - Irrigation solutions (glycine, sorbitol)
 - Procedures (TURP, hysteroscopy, laparoscopy)

Pseudohyponatremia

Normal serum

- Water 93%
- Fat and proteins 7%
- Sodium concentration in physiologically important serum water 154 mEq/L
($154 \div 0.93 = 142$)
- Measured Serum Sodium: 142 mEq/L

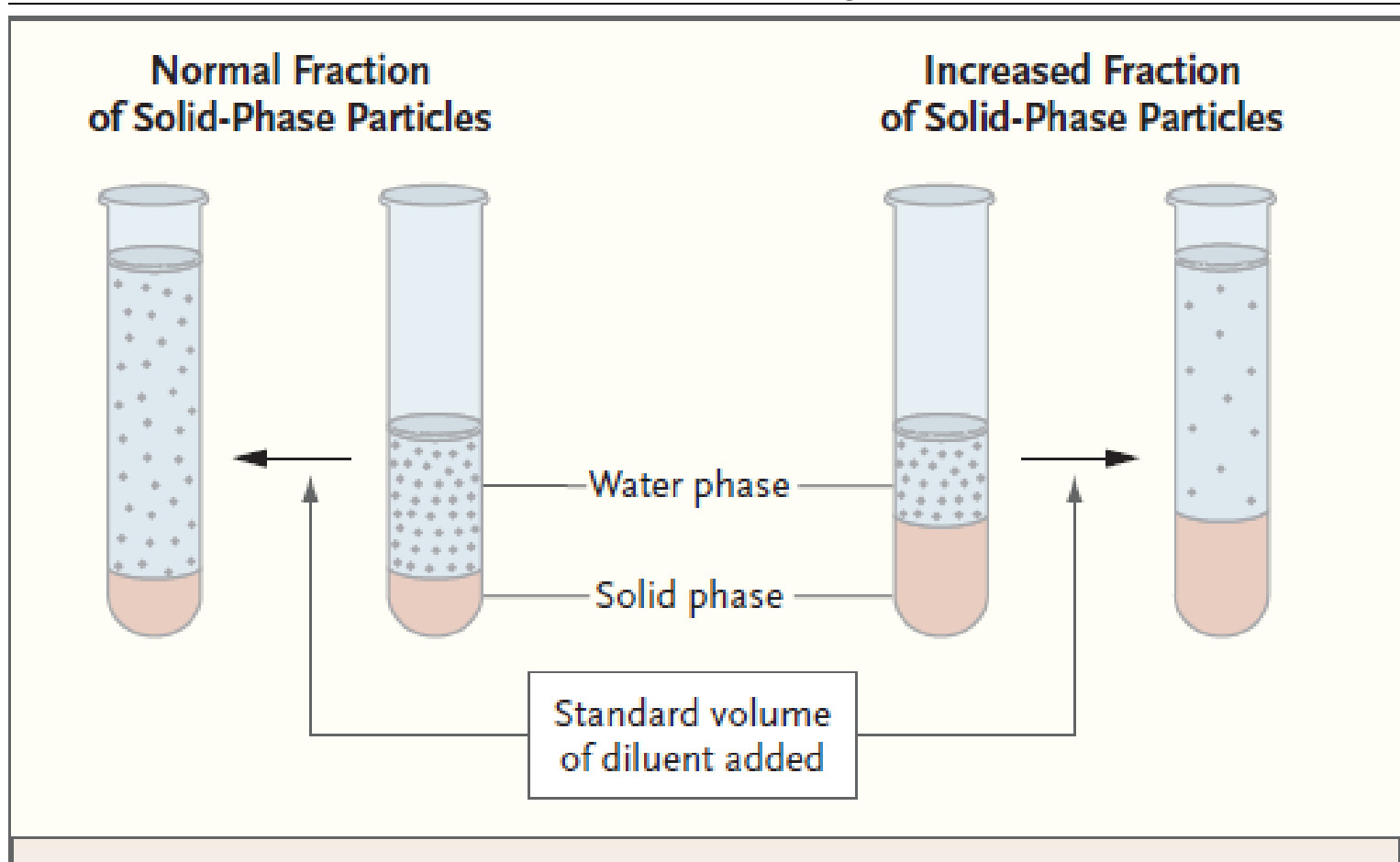
Abnormal serum

- Water 80%
- Fat and proteins 20%
- Sodium concentration in physiologically important serum water 154 mEq/L
($154 \times 0.80 = 123$)
- Measured Serum Sodium: 123 mEq/L

Measurement of Sodium

- Indirect with dilution
 - Flame photometry
 - Indirect potentiometry
- Direct
 - Direct potentiometry

Pseudohyponatremia- Indirect sodium measurement by dilution



Direct Potentiometry by Ion-Selective Electrode

- Measures electrical potential across a sodium-selective membrane immersed in an unknown serum sample
- Electrical potential is a function of the sodium activity in the sample
- Applied to undiluted sample for direct reading
- Can also be applied to a diluted sample for indirect potentiometry

What about this patient?

- Lipid panel:

Measurement	Value	Reference Range
Total cholesterol	1883 mg/dL	140-200 mg/dL
Triglycerides	945 mg/dL	30-149 mg/dL
HDL	30 mg/dL	40-59 mg/dL
Direct LDL	1760 mg/dL	0-129 mg/dL
Calculated VLDL	93 mg/dL	0-30 mg/dL

- Lipid Electrophoresis: Band of unknown significance between LDL and VLDL, likely Lipoprotein X

Lipoprotein X

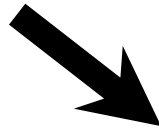
- Reflux of unesterified cholesterol and phospholipids into the circulation
- Usually in setting of cholestasis
- These particles are not soluble in water so will increase the solid fraction of plasma
- Can cause laboratory abnormalities but do not seem to have pathologic consequences

Case Patient

Imatinib

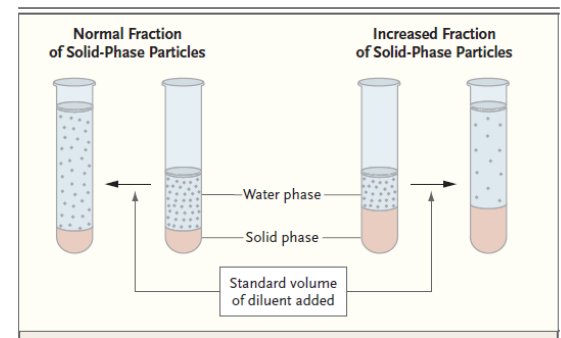
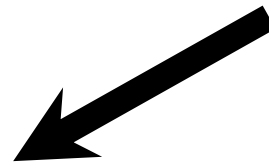


Cholestasis



Lipoprotein X

Pseudohyponatremia



Case #5

- 68 yo male with baseline Cr 1.1, presents to renal clinic for Cr up to 1.5
- Renal u/s with nephrocalcinosis (CT without calcium deposition 3 years earlier)
- Past Medical History: GERD, Small Lymphocytic Lymphoma, Melanoma
- Meds: ASA, Calcium, Vitamin D, MVI, Fish oil, Omeprazole
- Family History: mostly consistent with vascular disease

Case #5- 24h Urine

Stone Risk Factors / Cystine Screening: Negative (12/03/2015)

DATE	SAMPLE ID	Vol 24	SS CaOx	Ca 24	Ox 24	Clc 24	SS CaP	pH	SS UA	UA 24
01/19/16	S16477663	3.10	5.94	347	46 *	1049	1.38	6.394	0.21	0.706
11/30/15	S15504669	1.62	6.40	367	23	1084	2.20	5.991	0.78	0.629
11/29/15	S15504668	1.69	6.60	521	21	1041	1.13	5.521	1.53	0.589
REFERENCE RANGE		0.5 - 4L	6 - 10	male <250 female <200	20 - 40	male >450 female >550	0.5 - 2	5.3 - 6.2	0 - 1	male <0.800 female <0.750

Dietary Factors

DATE	SAMPLE ID	Na 24	K 24	Mg 24	P 24	Nh4 24	Cl 24	Sul 24	UUN 24	PCR
01/19/16	S16477663	234	86	138	1.065	22	216	41	12.27	1.2
11/30/15	S15504669	190	77	133	1.147	15	176	41	12.81	1.3
11/29/15	S15504668	189	87	140	1.079	19	203	47	13.81	1.4
REFERENCE RANGE		50 - 150	20 - 100	30 - 120	0.6 - 1.2	15 - 60	70 - 250	20 - 80	6 - 14	0.6 - 1.4

Case #5- Labs

- Na 142, K 4.7, Cl 101, CO2 29, BUN 26, Cr 1.60, Glu 94
- Ca 10.5, Phos 3.7
- 25 Vitamin D 51 ng/ml (20-80)
- 1,25 Vitamin D 122 pg/ml (18-64)

What is the most likely primary cause of the nephrocalcinosis?

- a) Lymphoma
- b) Sarcoid
- c) Low fluid intake
- d) High sodium intake
- e) Genetic factors

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Hypercalcemia and Malignancies

Malignancies associated with hypercalcemia

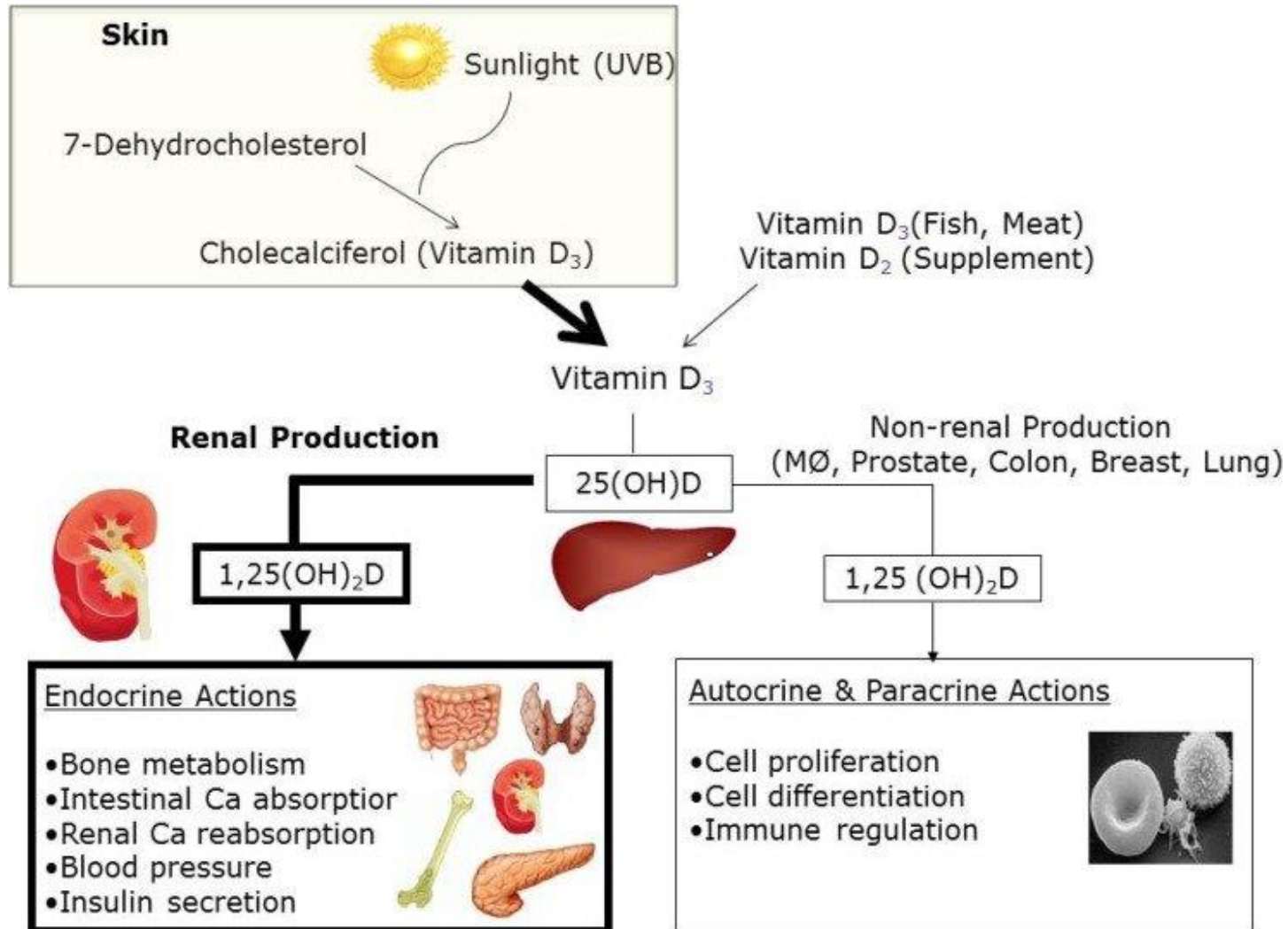
Humoral hypercalcemia (PTHrP):
Squamous cell carcinomas
Renal carcinomas
Bladder carcinoma
Breast carcinoma
Ovarian carcinoma
Prostate carcinoma
Colorectal carcinoma
Non-Hodgkin lymphoma
CML
Leukemia
Lymphoma
Osteolytic metastases:
Breast carcinoma
Multiple myeloma
Lymphoma
Leukemia
1,25-dihydroxyvitamin D:
Lymphoma (non-Hodgkin, Hodgkin, lymphomatosis/granulomatosis)
Ovarian dysgerminomas
Ectopic PTH secretion:
Ovarian carcinoma
Lung carcinomas
Neuroectodermal tumor
Thyroid papillary carcinoma
Rhabdomyosarcoma
Pancreatic carcinoma

PTHrP: parathyroid hormone-related protein; CML: chronic myeloid leukemia; PTH: parathyroid hormone.

1,25 Vitamin D and Lymphoma

- Normally 1-alpha hydroxylase from kidney converts 25-vitamin D to 1,25 vitamin D
- In this case there is regulation by PTH and FGF23
- In some lymphomas, malignant lymphocytes or macrophages cause PTH-independent conversion to 1,25 vitamin D

Vitamin D Metabolism



Other Sources of Extra-Renal 1,25 Vitamin D

- Locations of 1-alpha hydroxylase enzyme:
 - Renal proximal tubules
 - GI tract
 - Skin
 - Vasculature
 - Mammary epithelial cells
 - Osteoblasts and Osteoclasts

Granulomatous diseases and 1,25 vitamin D

- Most common are sarcoidosis and tuberculosis but also seen with other conditions including many fungal infections
- In granulomatous conditions, activated macrophages in lung and lymph nodes produce 1-alpha hydroxylase

Case follow-up

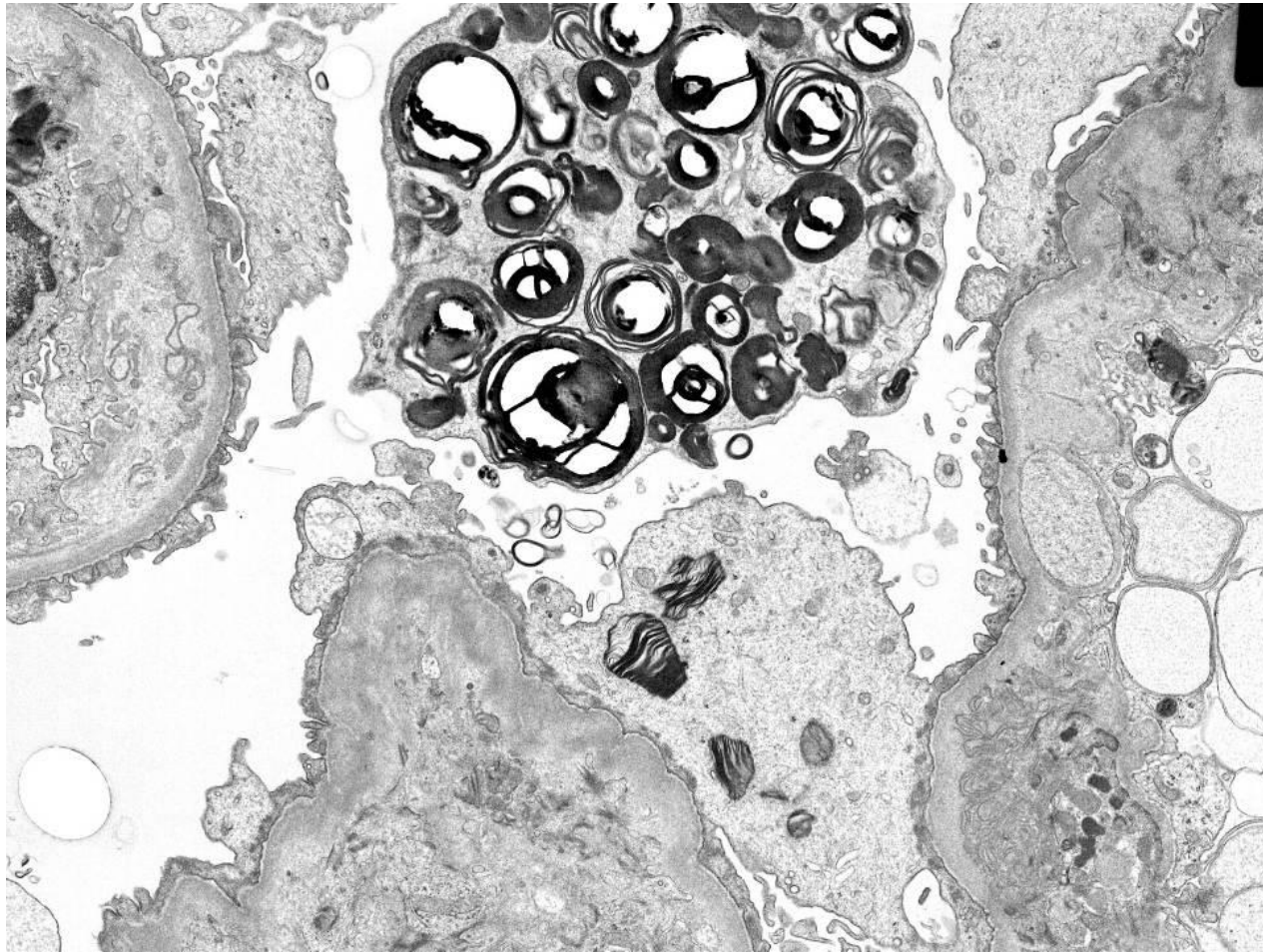
- Patient started on ibrutinib for lymphoma
- Creatinine baseline down to 1.1-1.3
- 1,25 vitamin D level decreased to 32pg/ml

Case Follow-Up

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DATE	SAMPLE ID	Vol 24	SS CaOx	Ca 24	Ox 24	Cit 24	SS CaP	pH	SS UA	UA 24
09/13/18	S25675499	3.26	1.20	73	38	526 *	0.30	6.518	0.14	0.649
08/22/17	S25350497	2.98	1.17	78	29	510 *	0.32	6.358	0.21	0.639
06/10/16	S19237991	1.77	10.14	326	46 *	1355	2.61	6.280	0.38	0.588
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Final “Must-Know” Zebra



What disease presents with these
“zebra bodies”?

- a) Fabry's Disease
- b) Immunotactoid Glomerulopathy
- c) Amyloidosis
- d) Alport's Syndrome
- e) Balkan Nephropathy

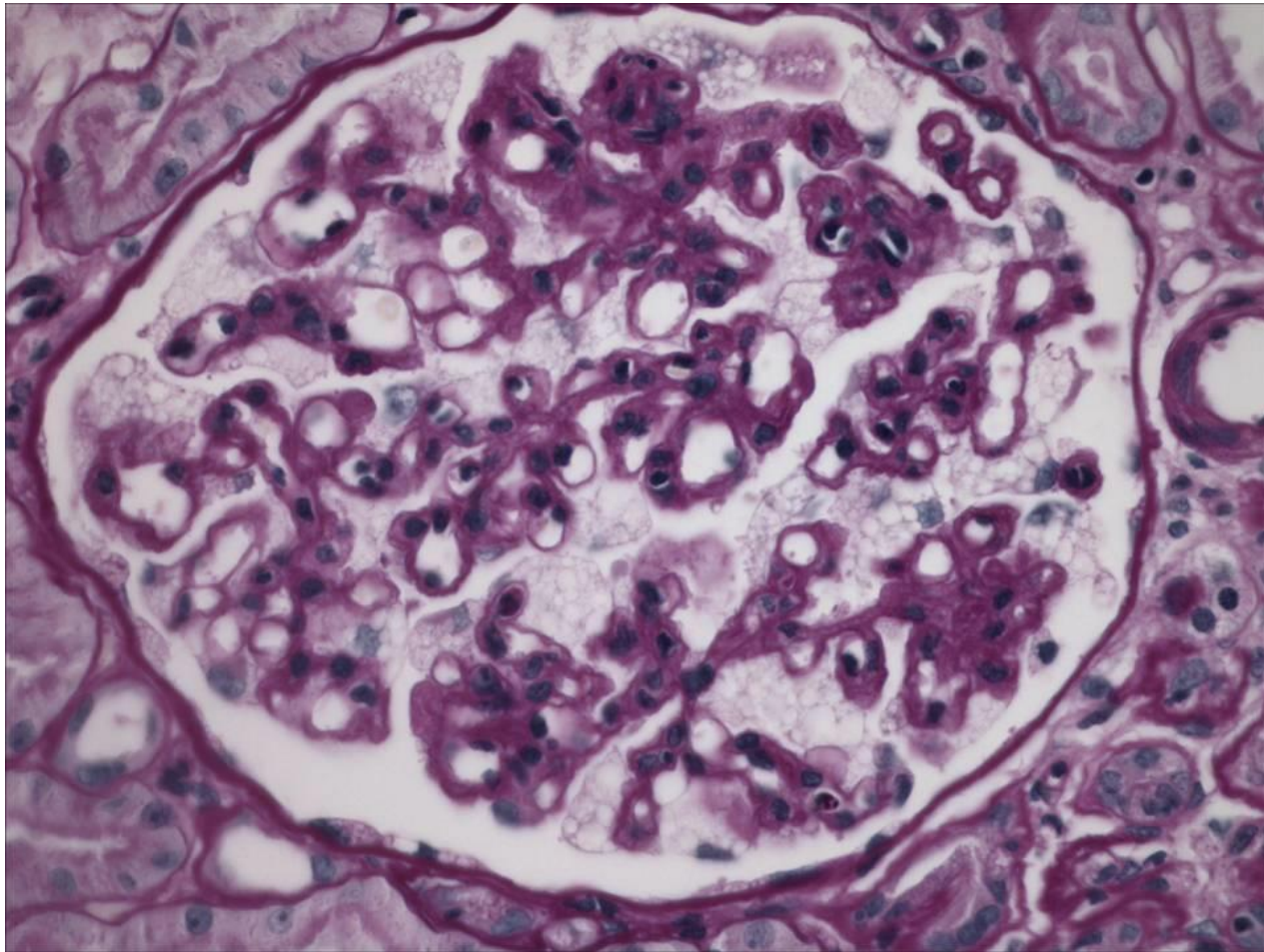
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“Zebra Bodies”

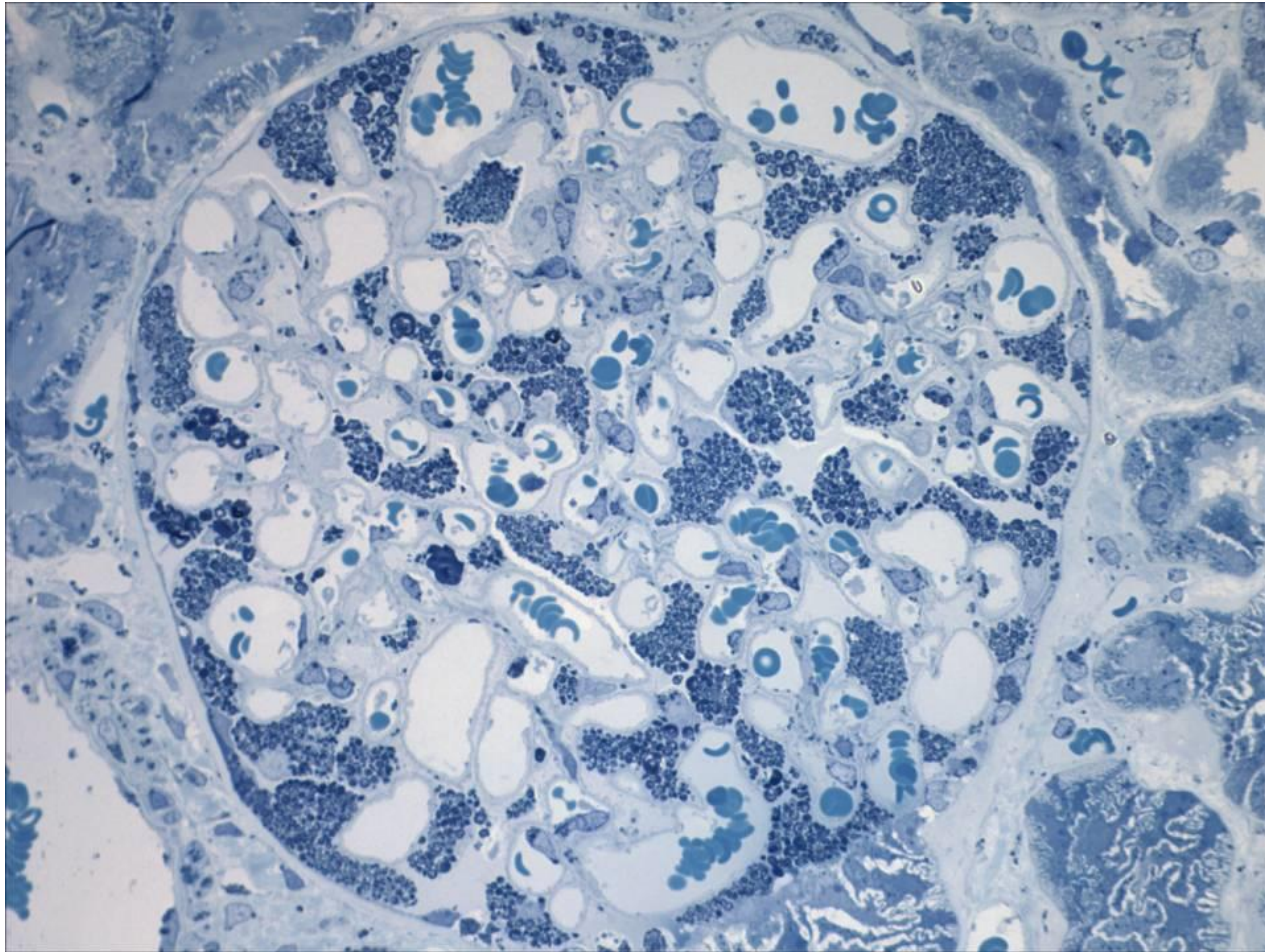
- Lysosomes filled with trihexosylceramide due to defect in lysosomal alpha-galactosidase

Other Fabry's Histology



Vacuolated visceral epithelial cells

Other Fabry's Histology



The vacuoles stain with toluidine blue in the semi-thin sections taken for electron microscopy

Thank You and Good Luck!

Supplemental References

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Disclosures

- Nothing to disclose