Case based discussion for Revise Nephrology 2023

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Case 1

- 64-year-old lady with h/o HTN, hypercholesterolemia, OA and h/o left Internal Carotid artery angioplasty back.

Meds:

- Perindopril 5mg mane
- Aspirin 100mg mane
- Amlodipine 10mg mane
- Atorvastatin 20mg mane

Admitted with pre-syncope thought to be due to electrolyte disturbance

Investigations

- Na- 133 mmol/L
- K- 2.1 mmol/L
- Cl- 92 mmol/L L
- HCO3- 32 mmol/L
- Urea- 5.2 mmol/L
- Creatinine- 85 umol/L
- eGFR- 62 mL/min/1.73m2 * L
- Normal LFTs/FBC

On Examination

- Appeared euvoluemic
- HR 88/reg RR 16 BP 137/90 (no postural drop) SaO2 97% on RA Afebrile
- JVPNE
- Nil pedal oedema
- HSDNM
- Chest clear
- Abdomen soft and non-tender (Did I miss something on exam??)

Treatment

- Gets started on K replacement with improvement
- Discharged 3 days later with f/u in my clinic
- THOUGHTS??????????????

Further Investigations

- Plasma aldosterone: 1,000 pmol/L *

  Aldosterone Ref. Interval: Upright: 61 – 978,Supine:32 - 654 pmol/L
- Plasma Renin: 228 mIU/L

  Renin Ref. Interval: Upright: 4.4 – 46.1, Supine: 2.8 – 39.9 mIU/L
- Aldosterone/Renin ratio : 4 (rules out primary aldosteronism as ration should be > 20)
- *Above suggests secondary aldosteronism (causes???)*

Causes of Secondary Aldosteronism

- Diuretic therapy
- Renovascular HTN (RAS and FMD)- Patient had a CTA which diagnosed left RAS
- Rarely renin producing tumours
- Certain forms of CAH

Conditions associated with metabolic alkalosis and/or hypokalemia-

- Characterised by disturbance of Na^+ re-absorption and can be further sub-divided into two groups depending on the blood pressure:
- *Low-normal BP*

- Bartter syndrome

- Gitelman syndrome

- *High BP*

- Primary hyperaldosteronism (Commonest cause in this group)

- Liddle syndrome

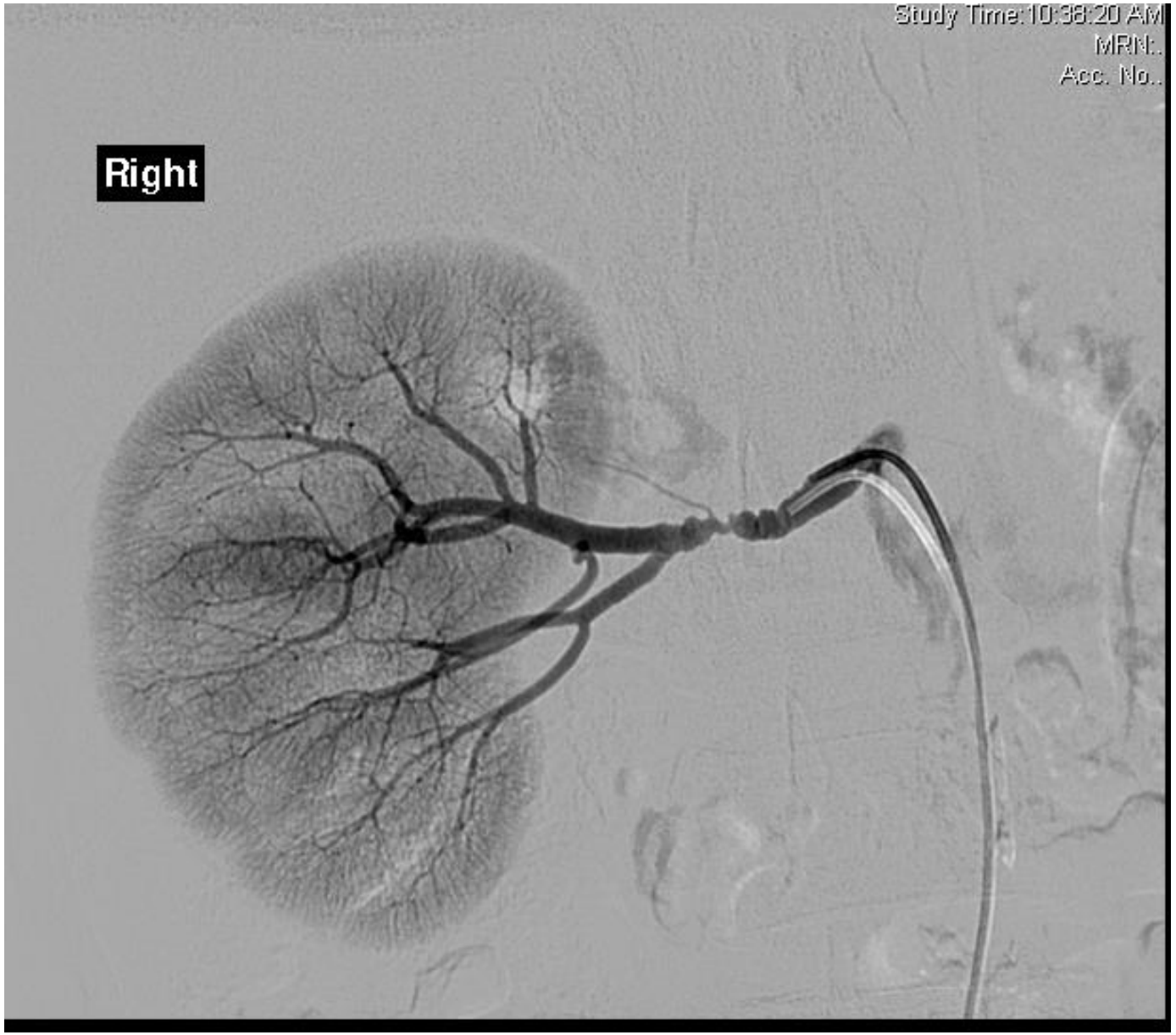
- Apparent mineralocorticoid excess

-Excessive Liquorice ingestion

- Familial Hyperaldosteronism (including Glucocorticoid –remediable hyperaldosteronism)
- *RAS by causing secondary hyperaldosteronism*

FMD of renal artery "string of beads" appearance

Middle or distal portion of renal artery



Case no. 2

- 54 yo gentleman feeling unwell for a week referred by GP for AKI
- Background- HTN (on perindopril 5 mg), ex-smoker and occ social drinker
- ED has done bloods- Creatinine is 385 umol/L with urea 25.2 mmol/L

❑Questions to ask the patient ?

❑What else would you like to know in investigations and why e.g. do you need to know the LFTs? FBC? Calcium?

❑What is the one thing your nephrology consultant would expect you to do yourself and NOT ask the nurse?

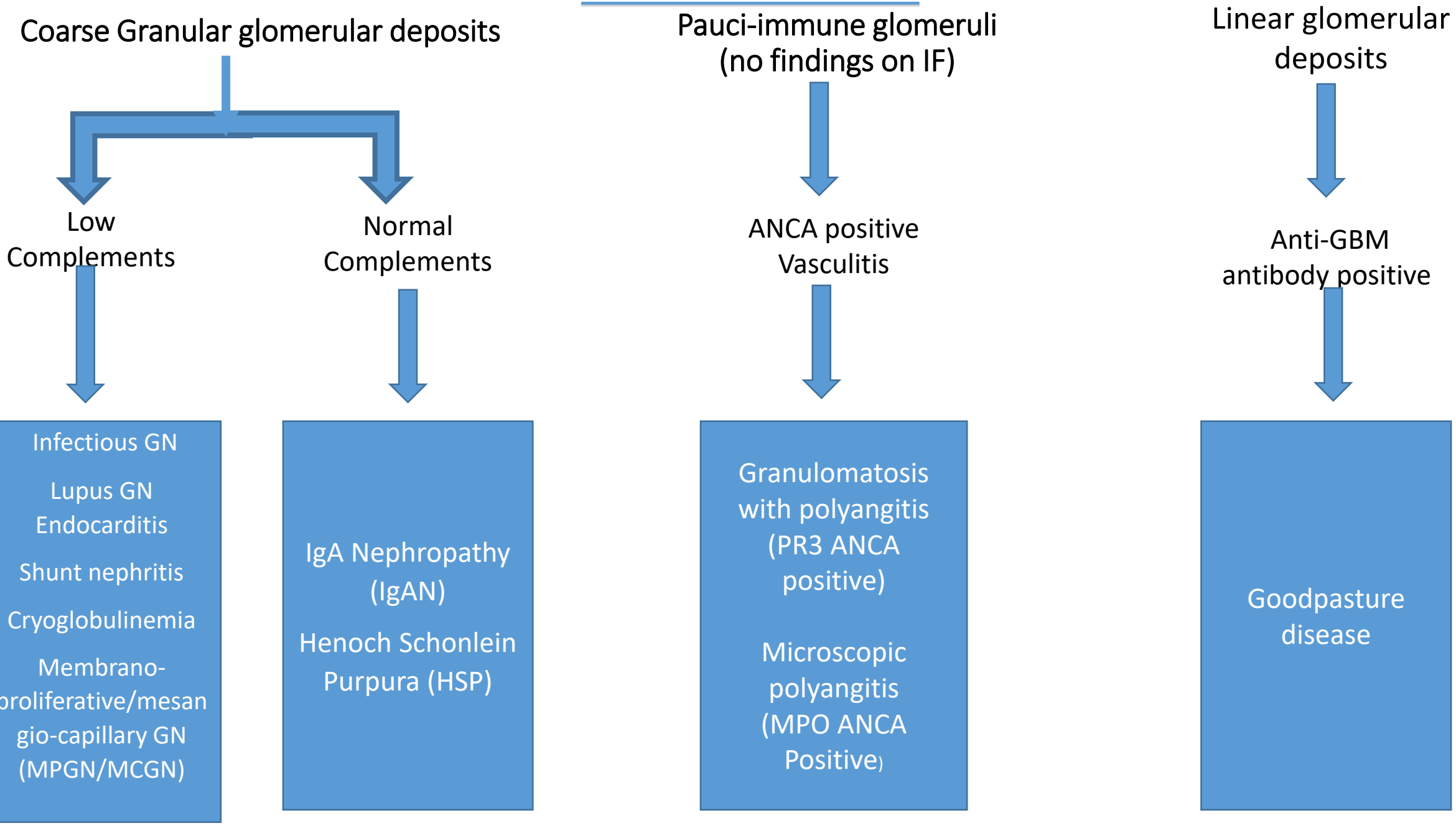
More info

- Denies diarrhoea/vomiting/prolonged exercise
- Has not taken any new medications like antibiotics (r/o interstitial nephritis)
- Denies renal stones, herbal meds, NSAIDs, drug abuse, family history
- Has been on perindopril for last six years and no dose change
- LFTs good, Hb 110, Platelets- 160, CMP normal
- Urine dipstick- blood ++++ AND protein ++

USG KUB- Normal-sized kidneys

NOW WHAT?

GN CLASSIFICATION



Case no. 3

- 27 yo male was admitted under respiratory with CAP. Found to have BP 200/110 mm Hg
- Admitted to having been started by GP on amlodipine 6 years back, discontinued after few weeks and BP never rechecked
- Urea /Creatinine/LFT/FBC normal
- K- 3.3 mmol/L and HCO3- 30 mmol/L

NEXT LINE?

Investigations

- Aldosterone 547 pmol/L
- Renin <2 mIU/L

What next?

What questions do we need to revisit in history?

Diagnosis of Primary Aldosteronism

- Initial test: Plasma aldosterone concentration/plasma renin activity or concentration (PAC/PRA or PRC) ratio >70 suggestive of primary aldosteronism
- REMEMBER the PAC/PRA ratio is **denominator** dependent
  - *Interfering drug for PAC/PRA: aldosterone antagonists and beta blockers are the only ones to really worry about…*
  - *Undetectable PRA in someone taking ACEI or ARB is a strong indicator for primary aldosteronism as normally ACEI or ARB causes high PRA*
- Confirmation of diagnosis: Oral sodium loading OR saline infusion test

Imaging in Primary Aldosteronism

- Adrenal computed tomography (CT) to determine subtype (adenoma versus hyperplasia versus adrenal carcinoma)
  - unilateral large (>4 cm) adrenal mass suggests carcinoma
  - abnormality in both glands suggests adrenal hyperplasia
- Limitations of CT: From 203 patients evaluated with CT and Adrenal Vein Sampling (AVS)
  - CT was accurate in only 53 % of patients
  - Based upon CT, 22 %would have been incorrectly excluded from adrenalectomy and 25 % might have had unnecessary or inappropriate surgery
- **Need adrenal vein sampling (AVS) to clearly differentiate between unilateral adenoma VS BL adrenal hyperplasia causing PA**

Role for adrenal venous sampling in primary aldosteronism. Young WF, Stanson AW, Thompson GB, Grant CS, Farley DR, van Heerden JA , Surgery. 2004;136(6):1227.

Case 4

- A 32 yo female referred by GP with symptomatic hypokalemia
- Background: Sjogren's syndrome without any significant organ involvement and followed up by Immunologist
- O/A hemodynamically stable with normal system examination
- **K: 2.0 mmol/L (3.3 to 5 mmol/L)**
- **HCO3: 11 mmol/L (22 to 32 mmol/L)**
- **Cl : 120 mmol/L (ref 95 to 110 mmol/L)**
- **Normal Anion gap/Na/urea/creatinine/LFTs/FBC**

What do we do next????

Urine dipstick to look for ?

How will the urine anion gap help to differentiate from metabolic acidosis/hypokalaemia due to chronic diarrhoea?

Renal Tubular Acidosis (RTA)

- Disorders of the tubule characterized by a normal anion gap (hyperchloremic) metabolic acidosis despite a relatively well-preserved GFR with either hypokalaemia (types 1 and 2) or hyperkalaemia (type 4)

- Defect in distal tubule H+ leads to distal (type1) RTA while defect in HCO3 reabsorption in proximal tubule leads to proximal (type2)RTA

- Type 4 RTA (commonest RTA) is characterised by decreased production of aldosterone or diminished responsiveness of the cortical duct to aldosterone

RTA….Low K+ in both types 1 and 2

Distal RTA (Type 1)

- Inability to secrete H+
- Urine pH >5.5 (no H+ in urine)
- Proximal tubules reabsorb all alkali including citrate which normally keeps Ca in urine soluble and so….renal stones
- **No** Fanconi syndrome
- *Sjogren's syndrome, SLE, PBC, autoimmune hepatitis,*
- Treat with alkali and K+ replacement

Proximal RTA (type2)

- Inability to reabsorb HCO3
- Urine pH < 5.5 (distal tubules secrete the excess H+ as in any acidosis)
- No renal stones
- **Fanconi syndrome** glycosuria,phosphaturia, uricaciduria and aminoaciduria
- *Myeloma, Wilson disease, drugs- tenofovir, acetazolamide*
- Same treatment…..needs bigger doses of alkali though…..

Type 4 RTA….hyper and NOT hypokalemia

- Decreased production or diminished responsiveness to aldosterone
- Associated with DM (commonest), NSAIDs, ACE-I , calcineurin inhibitors (cyclosporine and tacrolimus), K sparing diuretics and high dose heparin
- In those **not** hypertensive or volume overloaded, synthetic mineralocorticoid such as fludrocortisone may help
- In patients with hypertension or fluid overload, a thiazide or loop diuretic may help by increasing distal delivery of Na and consequently increase urinary secretion of H+ and K+

Urine anion gap

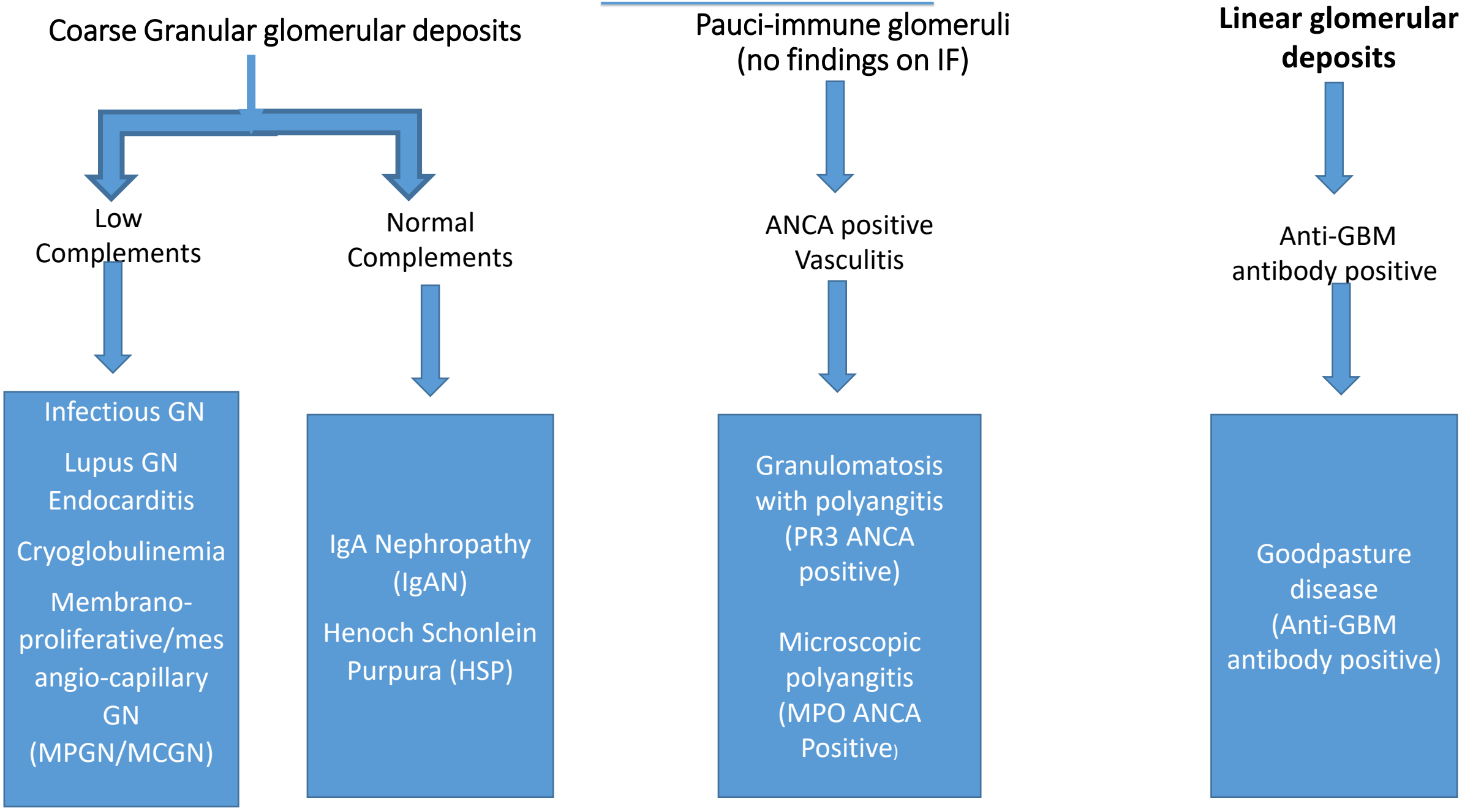
- UAG (in mEq/L or mmol/L) = Urine (Na + K - Cl)
- UAG is a measure of urinary NH4 (as ammonia exists as NH4Cl)
- Normally positive (20 to 90)
- In metabolic acidosis due to chronic diarrhoea, urine will try to excrete the extra H
- As NH3 is the major urinary buffer, for H the amount of NH4Cl in urine will increase causing negative UAG
- In distal acidosis as the defect is inability to excrete H, the urinary NH4Cl will not increase and so UAG stays positive

Case no. 5

- A 52-year-old woman presents with rising blood urea and creatinine over a 4-week period. Her urine has dysmorphic RBCs and the serum complement levels are low. Which one of the following causes is unlikely to be the cause of this presentation?

a. Infective endocarditis
b. Cryoglobulinemia
c. Microscopic polyangiitis
d. Systemic lupus erythematosus
e. Cholesterol atheroembolic disease

GN CLASSIFICATION



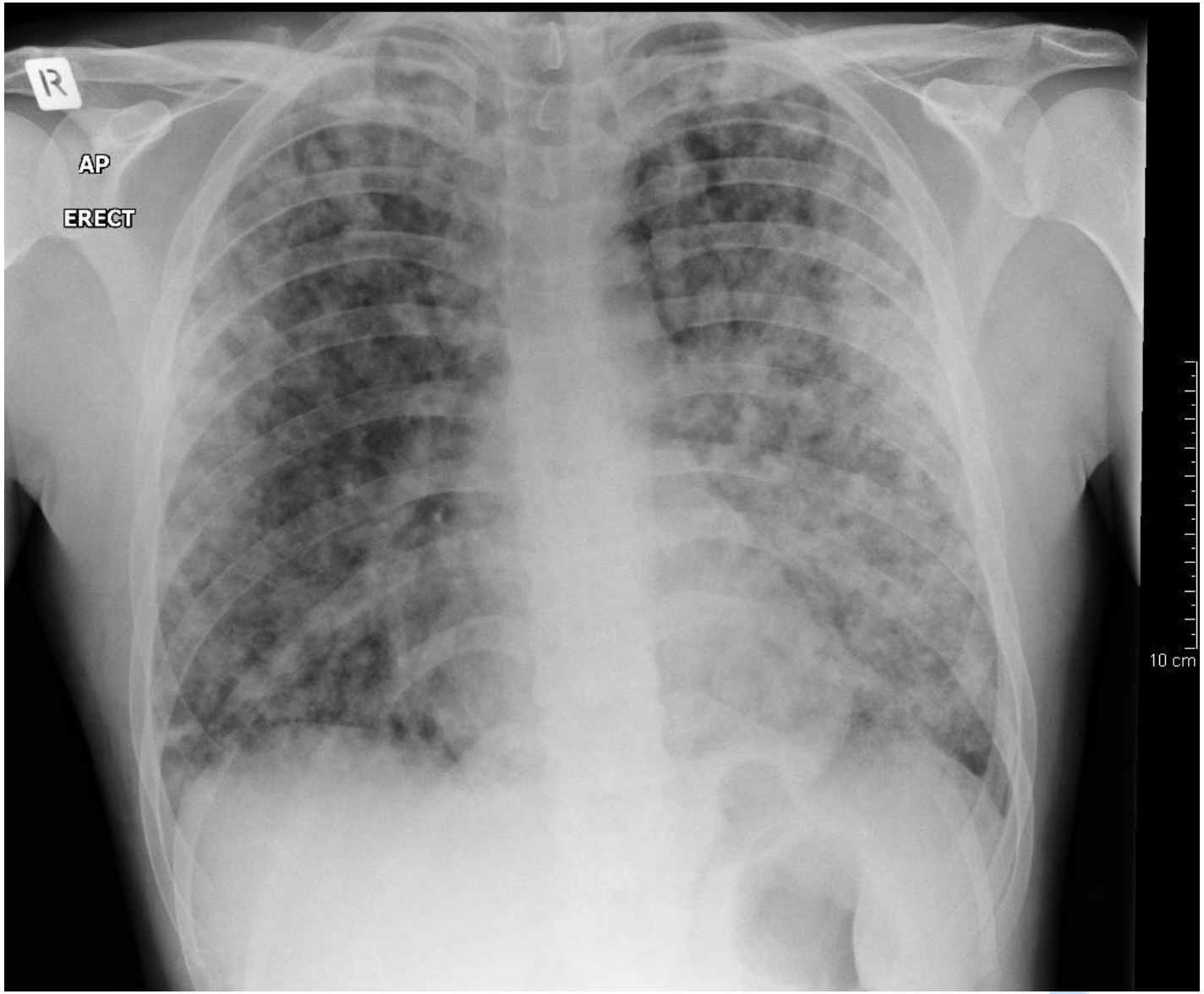
Answer is E: Atheroembolic disease

- Embolism of small usually distal arteries by cholesterol crystals/small pieces of atheromatous material originating from an atherosclerotic plaque
- May follow arteriography (commonest), cardiac catheterization or vascular surgery
- Effects skin (livedo reticularis, blue toes),GI (abdo pain, diarrhea, bleed), CNS (stroke, TIA, confusion),eyes(Hollenhorst plaques) and kidney (AKI with bland urine)
- May have transient hypocomplementemia and eosinophilia
- **Unlike contrast nephropathy presents few weeks to months after the inciting event and associated with a poor prognosis**

Case no. 6

- A 44 years old man with no previous medical history is admitted with lethargy, nausea and increasing SOB over two weeks. His BP is 160/92m Hg with fine inspiratory crackles in his chest. Investigations reveal a creatinine of 152umol/L with Hb of 108. Urine dipstick shows 3+ blood and 3+ protein. His CXR is on next slide and the respiratory team suggests DLCO which is raised. What is the likely diagnosis?

a. Bronchiectasis
b. Pulmonaray edema
c. Post streptococcal GN
d. Goodpasture syndrome
e. Membranous nephropathy


R
AP
ERECT
10 cm

Answer is D.

- Goodpasture's Disease is due to antibodies against NC1 domain of the alpha-3 chain of type IV collagen found in the glomerular and alveolar basement membranes
- Patients are positive for anti-GBM antibodies in their plasma
- Renal biopsy showing characteristic immunofluorescence finding of IgG deposition in a linear pattern along the GBM
- Typically presents with RPGN often accompanied by pulmonary haemorrhage and hence **DLCO raised**
- 10% to 40% of anti-GBM antibody-positive patients may also be positive for MPO ANCA and these patients usually have more aggressive disease