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P441 -Darn if it's dry, darn if it's wet!

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A young lady with background of cystic fibrosis was referred to the general nephrology clinic following finding of increased urinary protein creatinine ratio with preserved excretory function. She was also found to be hypoalbuminic. Her CF diagnosis was in her early teens and she had at least two infective exacerbations annually. She was also on maintenance antibiotics including nebulized aminoglycosides.

At presentation, she had evidence of peripheral oedema. Urine PCR was greater than 3g with normal level of serum creatinine. Albumin was low end of normal range. Acute screen including ANCA, myeloma screen, and lupus markers were negative with normal sized kidneys with no obstruction on ultrasound. A biopsy was planned as an outpatient procedure.

Unfortunately, she missed the appointment twice and presented acutely to her local hospital with significant nephrotic syndrome. She was subsequently transferred to the tertiary unit for a renal biopsy and diuresis. This was delayed due to concurrent infective exacerbation of CF. Formal anticoagulation was also delayed pending biopsy.

Biopsy was undertaken and the Congo red stain was strongly positive and immunohistochemistry confirmed AA amyloidosis. She was subsequently referred to the National Amyloid Centre for further assessments. The diagnosis was in keeping with chronic inflammatory state she was in due to her underlying CF and the recommendation from the National Centre was to keep her CF under control.

The respiratory team was consulted with regards to controlling her CF. Two conundrums arose in her case: anticoagulation was further delayed due as she required bronchial tree dilatation embolised and overdiuresis would increased thickening of her mucous, causing further exacerbation of her CF. Indeed, the latter would become a problem in the following months and she was repeatedly readmitted with fluid overload, electrolytes imbalances which was partly contributed by poor nutritional state, and infective exacerbation of CF. She continued to be nephrotic despite adequate dose of diuretics and angiotensin system blockade. Careful fluid balance could only able to be achieved whilst inpatient which resulted in lengthy admissions for each episode. This had impacted on patient’s mental health.

Her last admission was due to fluid overload and she underwent a second embolisation of the bronchial arteries. Unfortunately, she developed further complication in the form of spontaneous pneumothorax which was drained. Due to her poor functional baseline, she was unable to recover from this event and has since passed away. This was the first case seen by the local respiratory team in a patient with CF and renal AA amyloidosis and indeed for the renal team.