Shrinking lung syndrome and tubulointerstitial nephritis – Sjögren's syndrome or systemic lupus erythematosus manifestation?

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Introduction

- Shrinking lung syndrome (SLS) is a rare complication of several autoimmune diseases with unclear pathogenesis[1].
- SLS manifests with progressive dyspnea, pleuritic chest pain, diaphragmatic elevation, dry cough, restrictive pattern in respiratory function tests and lack of objective parenchymal abnormalities[1].
- Due to a small number of patients with this syndrome, there are no available guidelines, so the treatment is based on the physician's judgment[2-4].
- So far there has not been described an association between SLS and renal damage due to tubulointerstital nephritis (TIN).

Case report

- A 33-year-old woman
- Presented with arthralgia, fever, xerophthalmia, dyspnea and right thoracic wall pain with elevation of the right hemidiaphragm
- Laboratory findings showed anemia, lymphopenia, elevated erythrocyte sedimentation rate, positive anti-nuclear, anti-Sm, anti-dsDNA, Anti-Ro/SSA and anti-La/SSB antibodies, low C3 complement level, polyclonal hypergammaglobulinemia, proteinuria (up to 0.95 gr/24 hour)
- Acid-base status revealed the presence of moderate metabolic acidosis(HCO_3^-=17mmol/l), with hypokalemia (K^+ down to 2.9 mmol/L) and alkaline urine (pH=8).
- Restrictive pattern on pulmonary function testing and few atelectatic segments without signs of pulmonary embolism or interstitial lung disease on high resolution CT scan were found
- Due to clinical symptoms and laboratory findings, primary Sjögren syndrome (SS), systemic lupus erythematosus (SLE) or overlap syndrome were suspected.
- Kidney biopsy revealed TIN with moderate activity and chronicity, primarily as SS manifestation. Interestingly, the biopsy didn't show any typical signs of lupus nephritis (Figure 1).
- Changes in skin biopsy associated with the clinical picture probably corresponded to SLE (Figure 2).
- The patient was treated with high dose glucocorticoids, azathioprine and theophylline
- Application of the therapy resulted in regression of clinical symptoms, improvement of laboratory findings, including acid-base status and proteinuria normalisation.

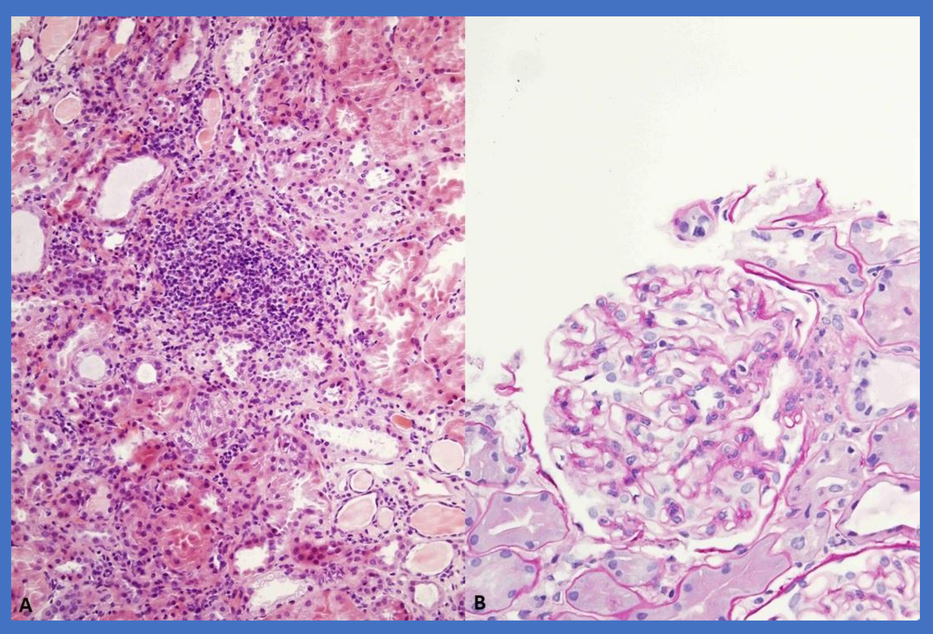

Figure 1. Kidney biopsy specimen. A) Dense mononuclear interstitial inflammation. HE stain, original magnification x200. B) Glomerulus with normal morphology. PAS stain, original magnification x400

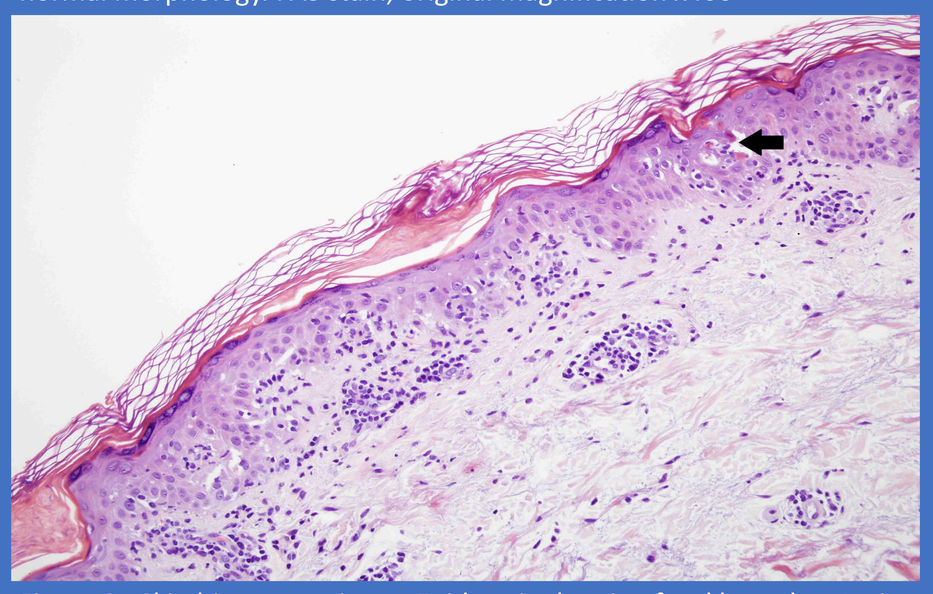

Figure 2. Skin biopsy specimen. Epidermis showing focal hyperkeratosis. There is interface change with basal vacuolisation and focal keratinocyte apoptosis (arrow). In the dermis there is perivascular mononuclear inflammation. HE stain, original magnification x200.

Conclusion

- This is to our knowledge the first case report of patient with SLS and TIN, nephrocalcinosis and renal tubular acidosis, probably related to SS.
- Concomitant presence of SLE might have influence on pathophysiological mechanisms in development of SLS.
- Simultaneous renal and pulmonary affection should encourage additional researches in pathophysiological mechanisms of these two rare disorders.

References
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