Rapidly progressive IgA nephritis and sarcoidosis

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Renal sarcoidosis frequently causes granulomatous interstitial nephritis, but clinically relevant nephritis is uncommon. IgA nephropathy caused by sarcoidosis is usually associated with milder stages of renal dysfunction, and only one case of rapidly progressive IgAN has been reported to date. We present an interesting case of a patient with a rapidly progressive form of IgA nephropathy caused by sarcoidosis that was successfully treated.

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INTRODUCTION

Renal sarcoidosis frequently causes granulomatous interstitial nephritis, but clinically relevant nephritis is uncommon.[1] Renal insufficiency accounts for 0.7% to 4.3% of clinical cases of sarcoidosis reported to date.[2] Glomerular lesions in addition to membranous glomerulonephritis are rare, and IgA nephropathy is an exception, especially in a form of crescentic glomerulonephritis.[1] IgA nephropathy is usually associated with milder stages of renal dysfunction.[3] Few cases of crescentic glomerulonephritis in sarcoidosis and only one with IgA nephropathy have been reported to date.[4-6]

CASE PRESENTATION

In January 2021, a 40-year-old woman was hospitalized for acute renal failure - creatinine was 290 µmol/L and urea was 15.5 mmol/L. Diuresis was about 2000 ml, without macrohematuria. Before hospitalization, she was healthy. Within four days, her creatinine increased to 465 µmol/L. In 2018 she had a normal creatinine with 1+ protein and erythrocytes in urine. In 7/2020, her creatinine was 110 µmol/L. On 18/Nov/2020 her creatinine was 170 µmol/L and she was advised to see a nephrologist but did not due to an epidemiological situation with COVID -19.

Serum calcium was normal, as well as IgA, ACE (29 U/l), complement levels, ANCA and anti-GBM. She had microscopic hematuria with proteinuria of 4.7 g/day.

We started therapy with metilprednizolone 80 mg iv after which we noted improvement in renal function. Renal biopsy revealed IgA nephropathy M1 E0 S1 T1, 7/19 glomeruli with cellular crescents, 5 globally sclerosed glomeruli, 26% IFTA, without granulomatous interstitial nephritis (Figure 1).

Because of abnormal chest X-ray MSCT scan of the thorax and abdomen was performed and showed peribronchial fibroindurative changes in the left upper lobe of the lung that was not imbibed by the contrast, nodular thickening of the parenchyma, numerous lymph nodes in the mediastinum and both hiluses up to 15×20 mm and multiple hypodense changes up to 7 mm in size in the spleen (Figure 2 and 3). PET/CT was performed and showed only mild metabolic activity of FDG in the described infiltrate in the lungs.

Video bronchoscopy was performed and was normal, as was the material for cytology and microbiological analysis. The serum level of angiotensin-converting enzyme was 29 U/l. We started therapy according to the guidelines for rapidly progressive IgAN, which was also appropriate for sarcoidosis - bolus corticosteroids (750 mg) for three days, followed by 1 mg/kg, and cyclophosphamide according to patient weight and renal function (800 mg, IV).

The patient was discharged from the hospital with a serum creatinine of 247 µmol/l.

She was treated with cyclophosphamide for the next six months (every 4 weeks), after which

Case Report

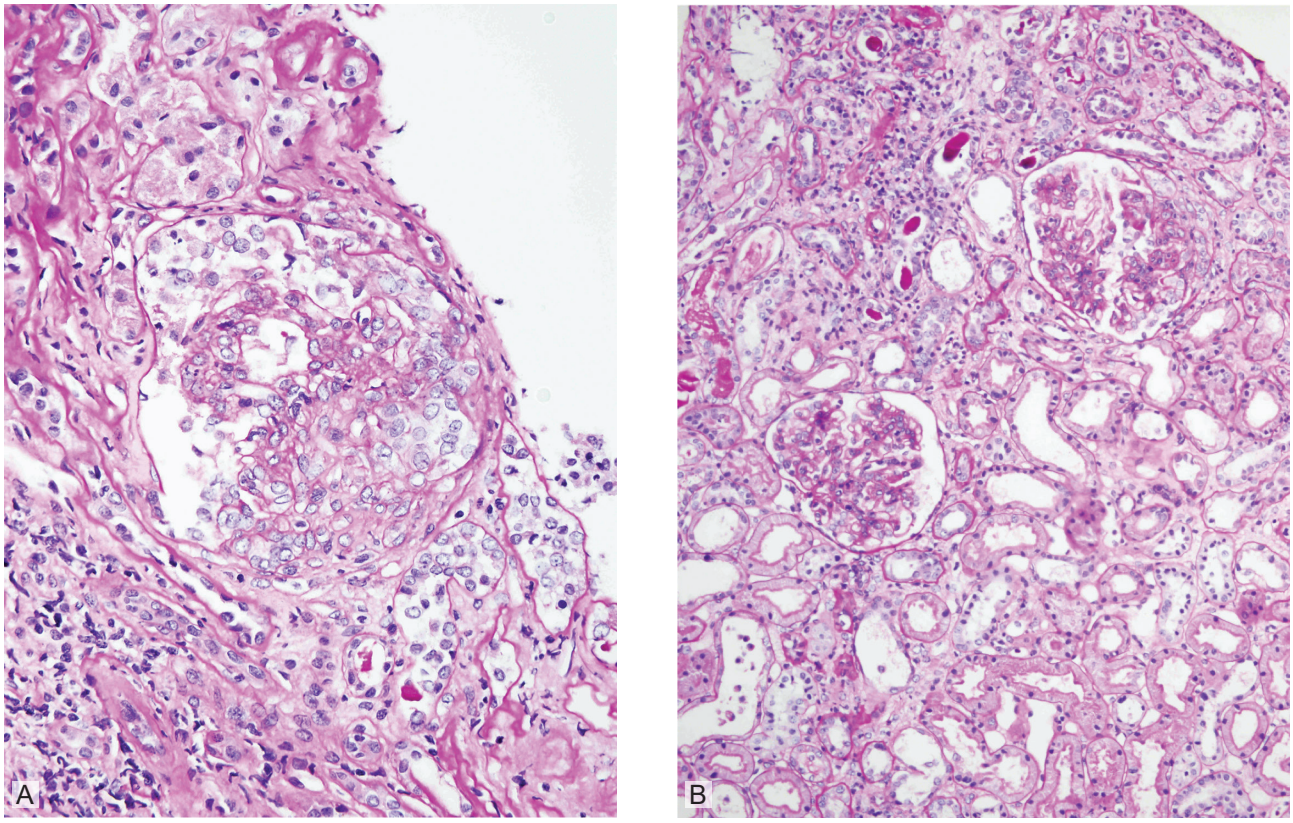

Figure 1. Kidney biopsy finding. A) Crescent, B) Area of interstitial fibrosis, normal glomerul and crescent. Stained with hematoxylin-eosin

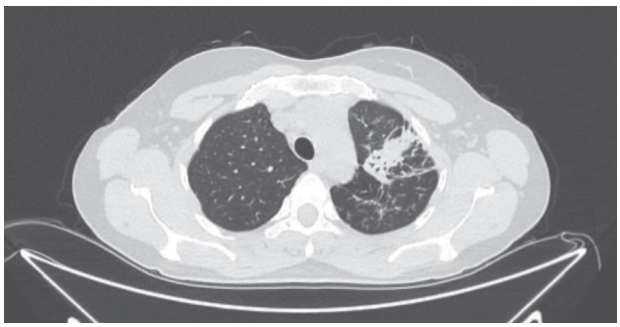

Figure 2. Peribronchial thickening in the left upper lobe of the lung

she was switched to azatioprine, with a gradual reduction in the corticosteroid dose. Her creatinine is now stable at 150 µmol/l, with proteinuria around 1.5g/24h. The control MSCT after 6 months showed regression of solid peribronchial thickening in the left upper lung lobe and mediastinal lymph nodes, and disappearance of nodular thickening of the lung parenchyma and changes in the spleen.

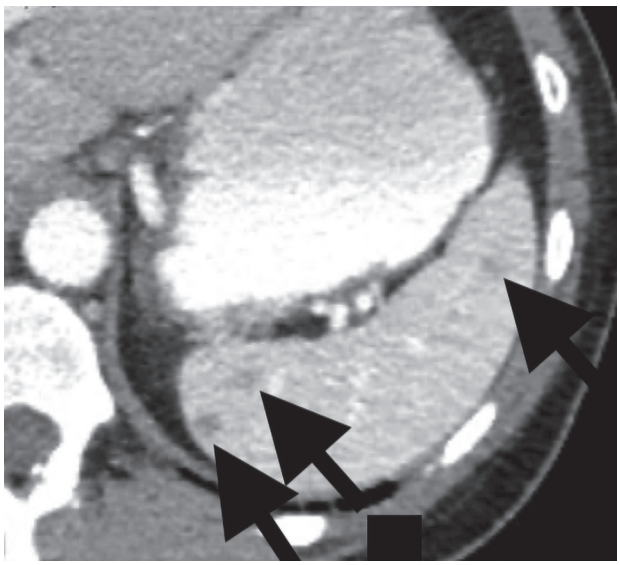

Figure 3. Hypodense changes in the spleen

DISCUSSION

IgA nephropathy is often associated with respiratory tract infections, and sarcoidosis may be associated with elevated serum IgA levels. It is not clear whether IgAN was present in our patient before sarcoidosis, but most likely an exacerbation of sarcoidosis caused rapidly progressive IgAN. Searching the literature, we found only one case

of a rapidly progressive form of IgAN with sarcoidosis, but because this article is 35 years old, unfortunately only an abstract was available.[6] Mahfoudhi and colleagues presented 12 patients with sarcoidosis and acute renal failure, and none of them were associated with IgAN.[7] Mahevas and colleagues presented the largest group of 47 patients with sarcoidosis and renal involvement.[2] Only two patients had mesangial IgA deposits, but the stage of renal failure is not reported in these patients. On the other hand, Löffler presented 27 patients with sarcoidosis and renal involvement, 7 of whom had IgA nephropathy.[3] None of them had cellular crescents and it was associated with milder stages of renal function impairment, in contrast to granulomatous interstitial nephritis, which was associated with severe renal failure.

CONCLUSION

In conclusion, although very rare, rapidly progressive IgA nephropathy is a possible consequence of sarcoidosis and cause of renal failure.

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Informed consent was obtained from the participant included in the study.

REFERENCES

1. Mannes K, Thomas PS. Sarcoidosis: rarely a single system disorder. Breathe (Sheff). 2020;16:200207
2. Mahévas M, Lescure FX, Boffa JJ et al. Renal sarcoidosis: clinical, laboratory, and histologic presentation and outcome in 47 patients. Medicine (Baltimore). 2009;88:98-106.
3. Löffler C, Löffler U, Tuleweit A, Waldherr R, Uppenkamp M, Bergner R. Renal sarcoidosis: epidemiological and follow-up data in a cohort of 27 patients. Sarcoidosis Vasc Diffuse Lung Dis. 2015;31:306-15.
4. Auinger M, Irsigler K, Breiteneder S, Ulrich W. Normocalcaemic hepatorenal sarcoidosis with crescentic glomerulonephritis. Nephrol Dial Transplant. 1997;12:1474-7.
5. Maroz N, Field H. Necrotizing crescentic glomerulonephritis related to sarcoidosis: a case report. J Med Case Rep. 2015;9:282.
6. Vanhille P, Beaudoin D, Mougenot B, et al. Glomérulonéphrite rapidement progressive à dépôts mésangiaux d'IgA au cours d'une sarcoïdose [Rapidly progressing glomerulonephritis with mesangial IgA deposits in sarcoidosis]. Nephrologie. 1986;7:207-9. French.
7. Mahfoudhi M, Mamlouk H, Turki S, Kheder A. Systemic sarcoidosis complicated of acute renal failure: about 12 cases. Pan Afr Med J. 2015;22:75

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