

A case of crescentic glomerulonephritis associated with relapsing polychondritis

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ABSTRACT

Relapsing polychondritis (RP) is a rare autoimmune disease, characterized by recurrent inflammatory episodes affecting various cartilage structures. Renal involvement in RP is unusual. Our case was a 42-year old woman presented with nephritic syndrome. Diagnosis of RP was made because of recurrent chondritis of both auricles, chondritis of costovertebral joints, non-erosive inflammatory arthritis of knees, and inflammation of ocular structures. Renal biopsy findings revealed diffuse crescentic and sclerotic glomerulonephritis, pauci-immune type. The diagnosis of RP may be missed when the destructive cartilage feature is not apparent or rare presentation of disease such as ocular or renal involvement preceded other manifestations. In this case we found that the irregular prednisolone use may prevent destructive cartilage damage but not renal failure of patient. We concluded that the association of crescentic glomerulonephritis in a patient with RP was assumed to be a rare manifestation of this disease.

Implication for health policy/practice/research/medical education:

The association of crescentic glomerulonephritis in a patient with relapsing polychondritis was assumed to be a rare manifestation of this disease.

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Introduction

Relapsing polychondritis (RP) is a rare autoimmune disease, characterized by recurrent inflammatory episodes affecting the cartilage structures. It can involve all types of cartilage including that of joints, tracheobronchial tree, ear and nose. Additionally, connective tissues which are rich in proteoglycans, such as heart, eye, blood vessels and inner ear may involve (1).

This disorder is frequently associated with rheumatoid arthritis, systemic vasculitis, connective tissue diseases, and hematologic disorders, however renal involvement is unusual.

Renal involvement in RP may be a part of an associated disorder like systemic lupus erythematosus or systemic vasculitis or may be primarily due to the disease itself. Renal biopsy most frequently shows a mild mesangial expansion and cellular proliferation (2). Other findings include crescentic glomerulonephritis, glomerulosclerosis, tubular loss, IgA nephropathy and tubulointerstitial

nephritis (3). Mesangial deposits of C3, IgG or IgM are seen in immunofluorescence studies.

Renal involvement is detected in 25% of cases and is associated with poor prognosis (4).

Case Presentation

A 42-year-old woman was admitted to our ward with renal failure. She had nausea, vomiting, weakness, dyspnea and conjunctivitis for a month. In past-medical history, recurrent attacks of erythema, edema and tenderness in her both ears noted which were treated as external otitis with antibiotics and dexamethasone. In addition, she cited nasal erythema, hoarseness, sore throat, knee arthritis and carpometacarpal joints discomfort occasionally for four years. Physical examination revealed bilateral nasal congestion, shortness of breath and chest wall pain especially in costovertebral joints. The result of examination by an ophthalmologist showed bilateral conjunctivitis. Cardiovascular and neurologic examination was normal.

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Diagnosis of RP was made because of recurrent chondritis of both auricles, chondritis of costovertebral joints, non-erosive inflammatory arthritis of knees, and inflammation of ocular structures. In laboratory tests, serum FBS; 96 mg/dL, Na; 136 mEq/L, K; 5.4 mEq/L, Urea; 195 mg/dL and creatinine was 8.5 mg/dL. Accordingly serum TSH was normal, Ca ; 8.6 mg/dL, phosphorus; 4.3 mg/dL, ALT 24 IU/L, AST 20 IU/L, CRP ++. Accordingly, WBC count was 10300/ μ L with normal differentiation, PLT count was 243000/ μ L. ESR 90 mm/h, urine RBC 15-20/hpf, urine WBC 4-6/hpf, urine protein 2+, urine sedimentation showed RBC cast. Serum and urine protein electrophoresis was normal. In serology HCV Ab, HIV Ab, HBS Ag, ANA, ANCA C and P, dsDNA was all negative. In DNA analysis HLA-B51 and HLA-B5 were positive. Ultrasound study revealed normal kidney size with high rose echogenicity of cortex. Renal biopsy has been performed with 16-gauge core needle. Histopathologic examination of the specimen showed 24 globally sclerotic glomeruli out of 42. The remaining glomeruli showed crescent formation that 14 were cellular to fibrocellular with 7 of them showing capsular rupture and also four fibrous crescents (Figure 1). The specimen also showed tubular atrophy and interstitial fibrosis in 70% of the tissue surface. Immunofluorescence study showed no immune deposits.

Considering all the findings diffuse crescentic and sclerotic glomerulonephritis, pauci-immune type was diagnosed in the background of RP setting.

Discussion

RP is a rare disorder and the etiology remains unknown. It is often associated with autoimmune disorders. Approximately 25%-35% of patients have other

autoimmune diseases too. It can involve all types of cartilage including that of joints, tracheobronchial tree, ear and nose and connective tissues rich in proteoglycans, such as in heart, eye, blood vessels and inner ear.

Renal disease presented in a minority of RP cases. Various kidney pathologies can occur in RP cases, including immunoglobulin (Ig) A nephropathy, tubulointerstitial nephritis, and glomerulonephritis (5,6). Espinoza and colleagues report a case with crescent GN and EM and IF showed immune complex (7) but our patient was pauci-immune. Our patient had not destructive lesions such as saddle nose or ear damage. The most important feature of our patients was renal involvement as diffuse crescentic and sclerotic glomerulonephritis before other destructive effects on cartilages organs. If the renal involvement is crescentic glomerulonephritis, ESRD may be more likely to occur than destruction of cartilage structures. That can be rare presentation of RP. The manifestations of chondritis in ear, knee and costovertebral joints and ocular signs were insidious and not disabling the patient. We find that the irregular corticosteroids use in our patient may prevent destructive cartilage damage but not renal failure. In addition absence of immune deposits on immunofluorescence study may be an effect of prednisolone administration. In our patient, the Behcet's disease criterion was not fulfilled while the pathergy test was negative too. No other rheumatologic diseases were detected. Before the biopsy, methylprednisolone and mycophenolate mofetil were started for the patient, but the treatment was discontinued because the renal biopsy showed advanced tubular and glomerular sclerosis changes. The patient sometimes had symptoms of nasal congestion and red eye and she has also attacks chest pain Which was

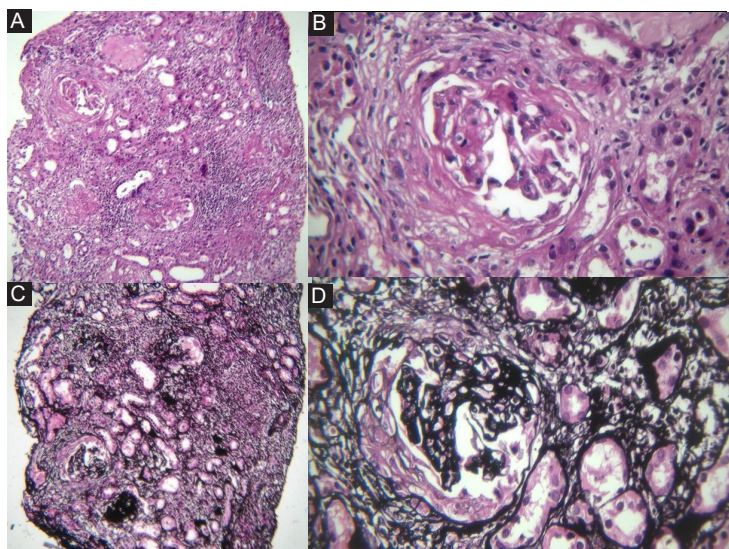


Figure 1. Light microscopy of renal biopsy. (A) $\times 40$ H&E; Partly crescentic and also sclerotic glomeruli and chronic tubulointerstitial changes. (B) $\times 10$ H&E; A crescentic and segmentally sclerotic glomerulus. (C) $\times 100$ Silver stain; severe tubular atrophy and interstitial fibrosis and crescentic glomeruli associated with segmentally and globally sclerotic changes. (D) $\times 100$ silver stain crescentic and segmentally sclerotic glomerulus.

controlled with a low dose of prednisolone and continued to remain on renal replacement therapy with regular hemodialysis.

Conclusion

The association of pauci-immune crescentic glomerulonephritis in a patient with RP was assumed to be a rare manifestation of this disease.

Authors' contribution

Handle of the patient by BB and AS. Primary draft by BB. Pathology report by MA. All authors read and signed the final paper.

Conflicts of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

Ethical considerations

Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors. The patient has given her informed consent regarding this case report.

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