

# Thin Glomerular Basement Membrane Disease

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## Abstract

### A Case of Wegener's Granulomatosis with Thin Glomerular Basement Membrane Disease

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Wegener's granulomatosis is a necrotizing, granulomatous vasculitis that involves multiple organs including the upper and lower respiratory tract and the kidney. The kidney initially exhibits focal necrotizing glomerulonephritis, which progresses to crescentic glomerulonephritis in Wegener's granulomatosis. We experienced a case of Wegener's granulomatosis which was associated with a thin glomerular basement membrane disease. The patient suffered from nasal stuffiness, recurrent serous otitis media, and tinnitus. Despite antibiotic therapy and ventral tube insertion, symptoms did not improve and hearing difficulty was aggravated. Ulcerative, necrotizing granulomatous inflammations with multinucleated giant cells were seen on nasal biopsy. She had recurrent microscopic hematuria and the renal biopsy findings by light and immunofluorescent microscopy did not reveal any abnormalities but diffuse thinning of the glomerular basement membrane (226nm) was observed by electronmicroscopy.

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With the above clinical findings and biopsy results, we diagnosed Wegener's granulomatosis with thin glomerular basement membrane disease. Thin glomerular basement membrane disease, also called benign recurrent hematuria, is characterized by diffuse thinning of the glomerular basement membrane and hematuria.

Weekly low-dose methotrexate together with prednisone was used as treatment regimen because nonglomerular microscopic hematuria may be the first sign of cyclophosphamide-induced renal toxicity. With the above combination therapy, she felt well-being sense and her hearing difficulty was also much improved. She has been treated as an outpatient with glucocorticoid.

Key Words : Wegener's granulomatosis, Thin glomerular basement membrane disease

Wegener's granulomatosis, : , 58  
 :  
 1). 70~80% : 2  
 ,  
 (focal segmental glomerulonephritis) ,  
 , 가 ,  
 가 .  
 2). , 90% ,  
 (antineutrophilic cytoplasmic antibody, ANCA)가 , 5  
 3,4). 가 :  
 Thin glomerular basement membrane disease : 10  
 , : 36.5 ,  
 5). 130/80mmHg, 68 / , 20 /  
 ,  
 methotrexate glucocorticoid 가 가  
 1

가 . tuning fork  
 :  
 11.2g/dL, 32.9%, 6,000/mm<sup>3</sup>,  
 304,000/mm<sup>3</sup>  
 55mm/hr(Westergren ),  
 BUN 9.7mg/dL,

0.8mg/dL, 7.0g/dL,  
 3.9g/dL, AST 29 IU/L, ALT 19 IU/L  
 , ANCA , -PR-3 , -MPO  
 , -RNP , -Sm , -Ro/La  
 CRP 1.70mg/dL  
 IgG/A/M 1,160mg/dL,  
 187mg/dL, 124mg/dL, C3/C4 75.6/39mg/dL

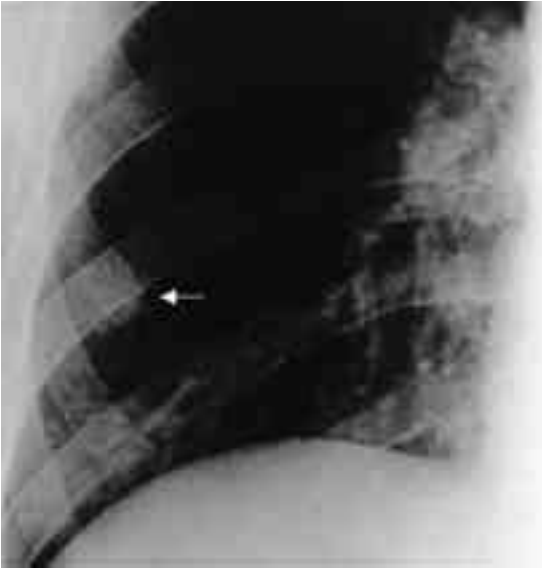


Fig. 1. Chest PA reveals ill-defined nodular density in RLL.

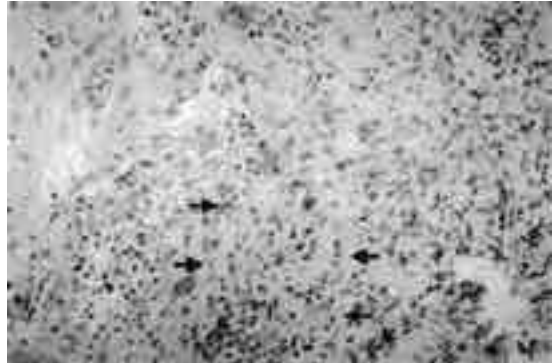


Fig. 2. A necrotic granuloma with neutrophilic microabscess is seen in biopsy obtained from the nasal cavity(H-E stain, x200).

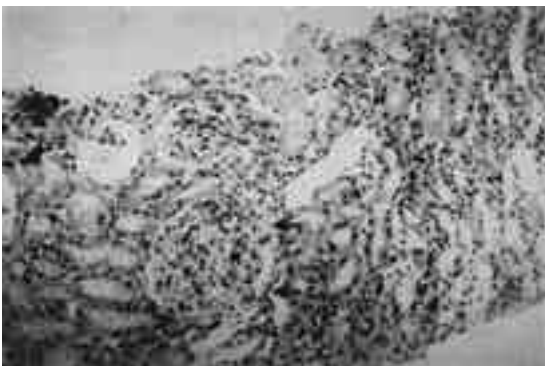


Fig. 3. Intact renal cortex without glomerular or tubulointerstitial change (PAS, x100).



Fig. 4. An electron micrograph of a glomerulus shows diffuse thinning(about 200 nm) of glomerular capillary basement membrane (x5,200).

, 24 78.0mg, ANCA가  
 676.7mg, 74.12ml/min . 1982 Davies <sup>8)</sup> , C-ANCA가  
 : 가 90%  
 ( 1). 1985 Van der Woude <sup>4)</sup>

. IgG ANCA 29-  
 , kDa serine proteinase-3 , 1988  
 , Falk , P-ANCA  
 : oxidase , myeloper-  
 ( 2) , <sup>3)</sup> 5%

. H-E PAS , 30~60mm/hr  
 ( 3),

가 가 가  
 가  
 ( 4).  
 : , 73%

prednisolone 60mg/day methotrexate 가  
 15mg/week . 가  
 , pred- 가  
 nisolone 30mg/day ,

1931 Klinger<sup>6)</sup>가 가  
 1939 50%  
 Wegener가 ,  
<sup>7)</sup>. 1973 85%  
 Fauci <sup>2)</sup> cyclophosphamide glucocorticoid ,

2/3

1/3

lamina densa

lamina

<sup>2)</sup>

densa

가

18%

가

40nm

375 ± 75nm,

325 ±

가 265nm

가

가 200nm

가

가

<sup>2)</sup>

75%

가

, Henoch-Schonlein

<sup>2)</sup>

, IgA

가

가

(crescent)

<sup>9)</sup>

가

<sup>13)</sup>

가

가

<sup>1)</sup>

H-E

PAS

90%  
phamide

2  
glucocorticoid

, cyclophos-  
75 ~ 93%

가  
cyclophosphamide

1986

Coleman

48

(idiopathic

<sup>2)</sup>

, Alport  
hematuria)

가

가

<sup>10)</sup>

cyclophosphamide

가

weekly low-dose methotrexate

<sup>5)</sup>

1973 Rogers

glucocorticoid

cyclophosphamide

familial benign essential hematuria

가

<sup>15)</sup>

가<sup>1)</sup> 1985

Dische

cyclophosphamide

<sup>5)</sup>

가

cyclophosphamide

Tiebosch

<sup>12)</sup>

methotrexate

glucocorticoid

가

6

58

가

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