

# Henoch-Schönlein

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## A Case of Wegener's Granulomatosis with Delayed Diagnosis due to Clinical Features of Henoch-Schönlein Purpura

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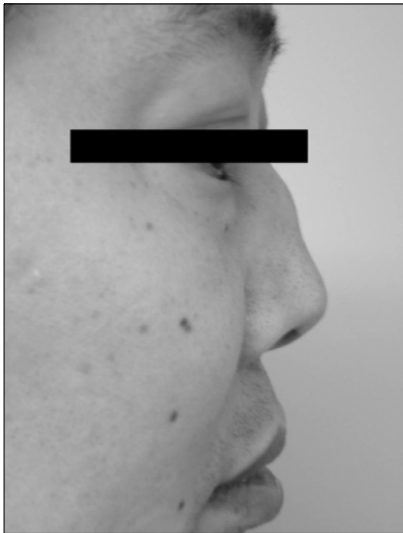
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Wegener's granulomatosis is a systemic vasculitis of the medium and small arteries, as well as of the venules, arterioles, and occasionally large arteries, and primarily involves the upper and lower respiratory tracts and the kidneys. Renal symptoms of Wegener's granulomatosis are indistinguishable from those of vasculitis such as Henoch-Schönlein purpura and microscopic polyangiitis. This case, though initially diagnosed as Henoch-Schönlein purpura, was confirmed as Wegener's granulomatosis from a lung biopsy fifteen years after the initial diagnosis. We report this case with a review of the literature. (*Tuberc Respir Dis* 2007;63:531-536)

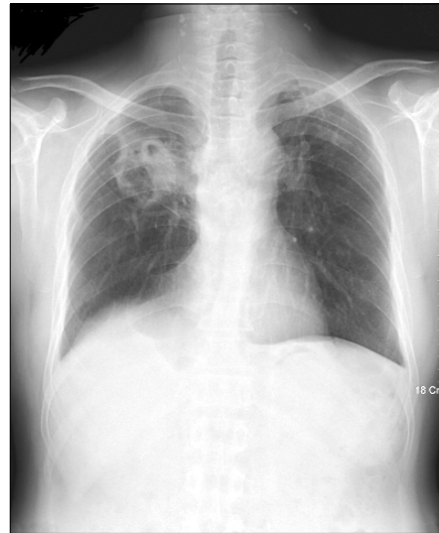
**Key Words:** Wegener's granulomatosis, Henoch-Schönlein purpura

서 론	증 례
<p>환자, 46 주소: 현병력: 3</p> <p>가 가</p> <p>Henoch-Schönlein 가<sup>2</sup>.</p> <p>15 Henoch-Schönlein 46</p>	<p>환자: ○○, 46 주소: 현병력: 3</p> <p>가 가</p> <p>과거력 및 가족력: 21</p> <p>2</p> <p>31</p> <p>1.4 mg/dL, 가</p> <p>ANCA (antineutrophil cytoplasmic antibodies)</p> <p>IgA</p> <p>(leukocytoclastic vasculitis)</p> <p>Henoch-Schönlein , 38</p> <p>(abscess)</p> <p>7 33</p>

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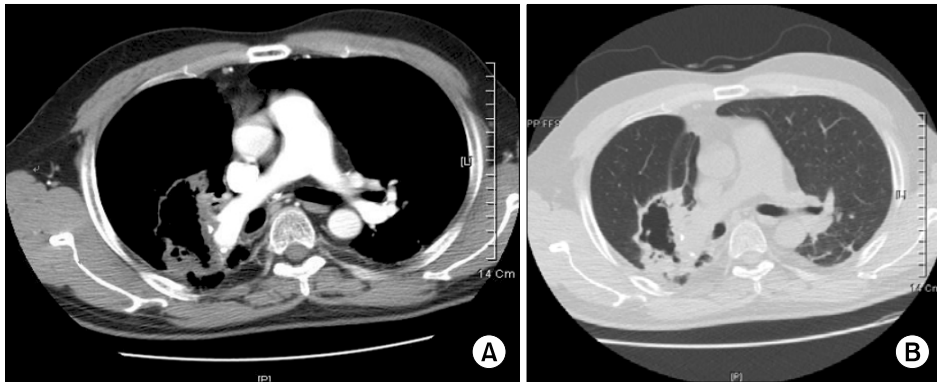
**Figure 1.** On patient's facial picture, a saddle nose is observed.



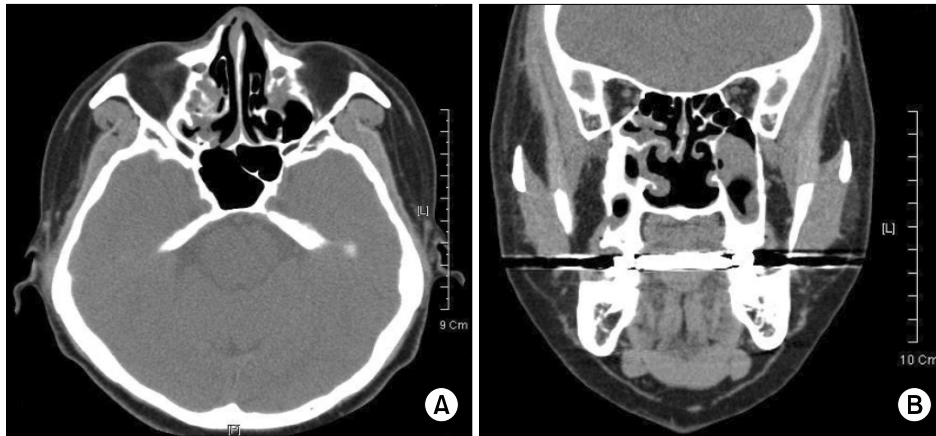
**Figure 2.** Chest radiography shows cavitary lung mass at the right upper lung field.

가 , 가 , 37 (infected bullae) , (chronic organizing pneumonia with fibrosis and necrosis) . C-ANCA (circulating antineutrophil cytoplasmic antibodies) . 42 2.0 mg/dL, 2.1 g/day, C-ANCA IgA (diffuse mesangial proliferative glomerulonephritis with focal necrotizing glomerulitis) 6 , 4 10 mg . , 가 신체검사 소견: 120/70 mmHg, 75 / , 20 / , 36.5°C , (Figure 1)가 . , (Figure 4).

검사실 소견: 8,770/mm<sup>3</sup> ( 83.4%), 11.4 g/dL, 371,000/mm<sup>3</sup> , aPTT 40.7 , PT 14.1 . AST 20 U/L, ALT 17 U/L, 6.8 mg/dL, 3.2 mg/dL, 39 mg/dL, 3.1 mg/dL, Na 141 mEq/L, K 5.1 mEq/L, Cl 108 mEq/L, 10.6 mg/dL, 9.1 mg/dL, 4.4 mg/dL . 1.015, pH 5.0, 0~1/HPF, 0~1/HPF , 24 143.5 mg/day, 34.2 mL/min , C-ANCA가 , P-ANCA (perinuclear antineutrophil cytoplasmic antibodies) 가 방사선학적 소견: 가 (Figure 2). 가 5.6x3.4 cm (Figure 3), (Figure 4). 임상 경과: 가



**Figure 3.** Chest CT scans show an irregular shaped mass consisted of various size of cavities with surrounding fibrosis and speckled margins (A; mediastinal setting view, B; lung setting view).



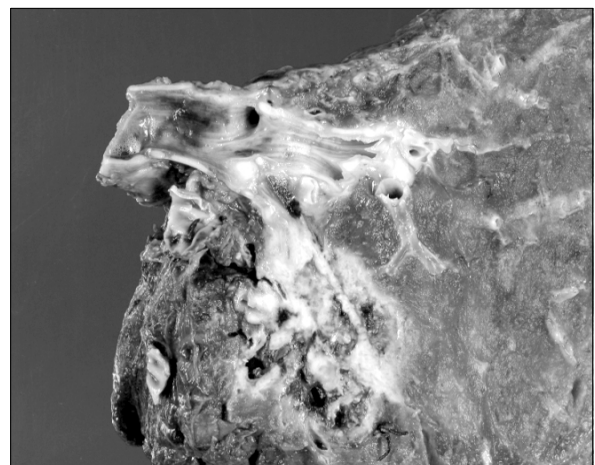
**Figure 4.** Facial CT shows small fluid density and thickening of bony wall in the right and left maxillary sinuses. Axial (A) and coronal (B) view.

수술 및 병리 소견:

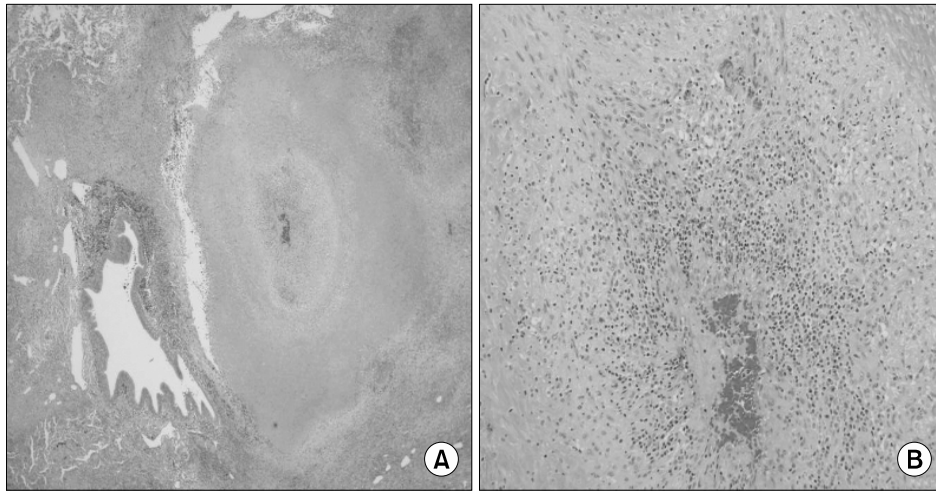
5×4 cm 가  
(Figure 5).

(histiocyte) (multinucleated giant cell)  
(Figure 6)

치료 및 경과 관찰: 70 mg  
125 mg , 1  
C-ANCA



**Figure 5.** Cut surface shows a rather ill-defined mass, which is consisted of varying sized multiple cyst with dense fibrous walls in the middle lobe, measuring 5×4 cm in size.



**Figure 6.** On microscopic examination, the granulomatous necrotizing vasculitis of large arteries and veins (A; H&E stain,  $\times 200$ ), surrounded by irregular shaped fibrinous necrosis and histiocytes and multinucleated giant cells (B; H&E stain,  $\times 400$ ).

고 찰

70% P-ANCA 7.

가 (vasculitis and granulomatous inflammation)

50 가 25% (leukocytoclastic vasculitis)

3 (necrotizing segmental glomerulonephritis)

8,9 Hensch-Schönlein (leukocytoclastic vasculitis) , IgA

2.

4 가 가 , (microscopic polyangiitis) Hensch-Schönlein

5. Woodworth 68

C-ANCA , , 5

가 ,

5,6 ANCA C-ANCA P-ANCA , 4~78 , 90% C-ANCA ,

5  
 Miyata Henoch-Schönlein C-ANCA  
 (leukocytoclastic vasculitis) IgA 가  
 C-ANCA  
 Henoch-Schönlein 15  
 가 가  
 가 가  
 가 가  
 21  
 31 Henoch-Schönlein 가  
 가  
 C-ANCA가 37 42  
 42  
 )  
 가 ANCA Henoch-Schönlein  
 ANCA  
 가 Henoch-Schönlein  
 IgA (3+)  
 IgA가 8

C-ANCA

요 약

Henoch-Schönlein

IgA Henoch-Schönlein 15  
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SB Song et al: Wegener's granulomatosis with delayed diagnosis

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