

Crohn's Disease Associated with Pachydermoperiostosis

Won Ho Kim, M.D., Yoon Woong Go, M.D., Chung Ryul Lee, M.D.,
E Wha Kang, M.D., Kea Won Kwon, M.D.* and Ho Geun Kim, M.D.*

Departments of Internal Medicine and Pathology,
Yonsei University College of Medicine, Seoul, Korea*

Pachydermoperiostosis is a rare hereditary syndrome characterized by finger clubbing, periosteal change, pachydermia and autonomic nervous system symptoms such as facial flushing and hyperhidrosis. This syndrome is known to be associated with a variety of diseases such as cranial suture defect, female escuchen, bone marrow failure, hypertrophic gastropathy and Crohn's disease. Crohn's disease is a chronic idiopathic inflammatory disease of the intestine and frequently presents many kinds of extraintestinal manifestations including finger clubbing. Recently, Compton et al. reported 3 patients with Crohn's disease associated with pachydermoperiostosis in a family as a new syndrome. Recently, we have also experienced 3 patients with pachydermoperiostosis in a family. One of them developed Crohn's disease at 45 years of age. The elder sister of the proband had protein losing enteropathy without a significant morphological change of gastrointestinal tract. The younger brother had experienced repeated gastrointestinal bleeding caused by multiple ileal ulcer. (**Kor J Gastroenterol 1999;33:723 - 729**)

Key Words: Crohn's disease, Pachydermoperiostosis

osteoarthritis)

.12

(pachydermoperiostosis)

, (pachydermia) (periostosis) 가
(hypertrophic

: 1998 4 9 , : 1998 7 23

: , 120-752, 134

Tel: (02) 361-5410, Fax: (02) 393-6884

가 가

가 가

(superior mesenteric artery synd-

cutis verticis gyrata rome)

.12 가 . 가 24 가 32

가 가 , , 가 5 2 5

.34 3

, 가 .4 가 가

2 가 가 4 3 (4)

(escuchen), , 가 (2)가 가

.59 가 10 (3)

.11 38% 가 , (2) 32 (grease) , 36

가 7 가 , .1213

가 Comptom 14 (4)

1 가 10 , 18 , 20

, 22

. 23

45 가 5 3 , ,

25 ,

, 30 가 ,

, 38 . 29 가

가 , 가 .
 (Fig. 1). (Fig. 2a)
 52 kg, 165 cm, 가 (Fig. 2b).
 110/70 mmHg, 80 , 37.0 가

Fig. 1. Pedigree. Left upper portion denotes Cronh's disease and right lower portion represents pachydermoperiostosis. Black filled portion mean confirmed diasease and shaded area means suggestive disease but not confirmative.
 SLE, systemic lupus erhythematosus; CD, Crohn's disease; PDP, pachydermoperiostosis.

Fig. 2. Gross appearances of proband. (A) Face photography shows greasing skin, deep furrow of forehead and protruding chin. (B) Hand photography shows clubbing of fingers.

8.8 g/dL, 29.5%, 457,000/
 μL , 11,300/ mm^3 71%, 17%,
9%, 4%), ESR 25 mm/hr, 50
 $\mu\text{g/dL}$, (TIBC) 230 $\mu\text{g/dL}$,
(UIBC) 180 $\mu\text{g/dL}$, prothrombin time
91%
AST 9 IU/L, ALT 7 IU/L, 0.3 mg/dL,
132 mg/dL, 5.8 g/dL, 2.1 g/dL,
BUN 6.2 mg/dL, creatinine 0.9 mg/dL, calcium 7.1
mg/dL, inorganic phosphorus 3.5 mg/dL, amylase 42
U/dL, Na 139 mEq/L, K 3.6 mEq/L, Cl 103 mEq/L

9 mm

가 (Fig. 3)

가

가

5.48 ng/mL

가

VDRL

(Fig. 4).

Fig. 3. Bone X-ray of leg demonstrates. Thickening of cortex and diffuse irregular periosteal reaction with new bone formation of tibia and fibula are found.

Fig. 4. Barium enema shows irregular narrowing of the lumen with loss of haustral marking. The mesenteric border is shortened and many sacculations are seen on the antimesenteric border. Numerous Barium collections with mucosal destruction pattern are noted on the entire colon.

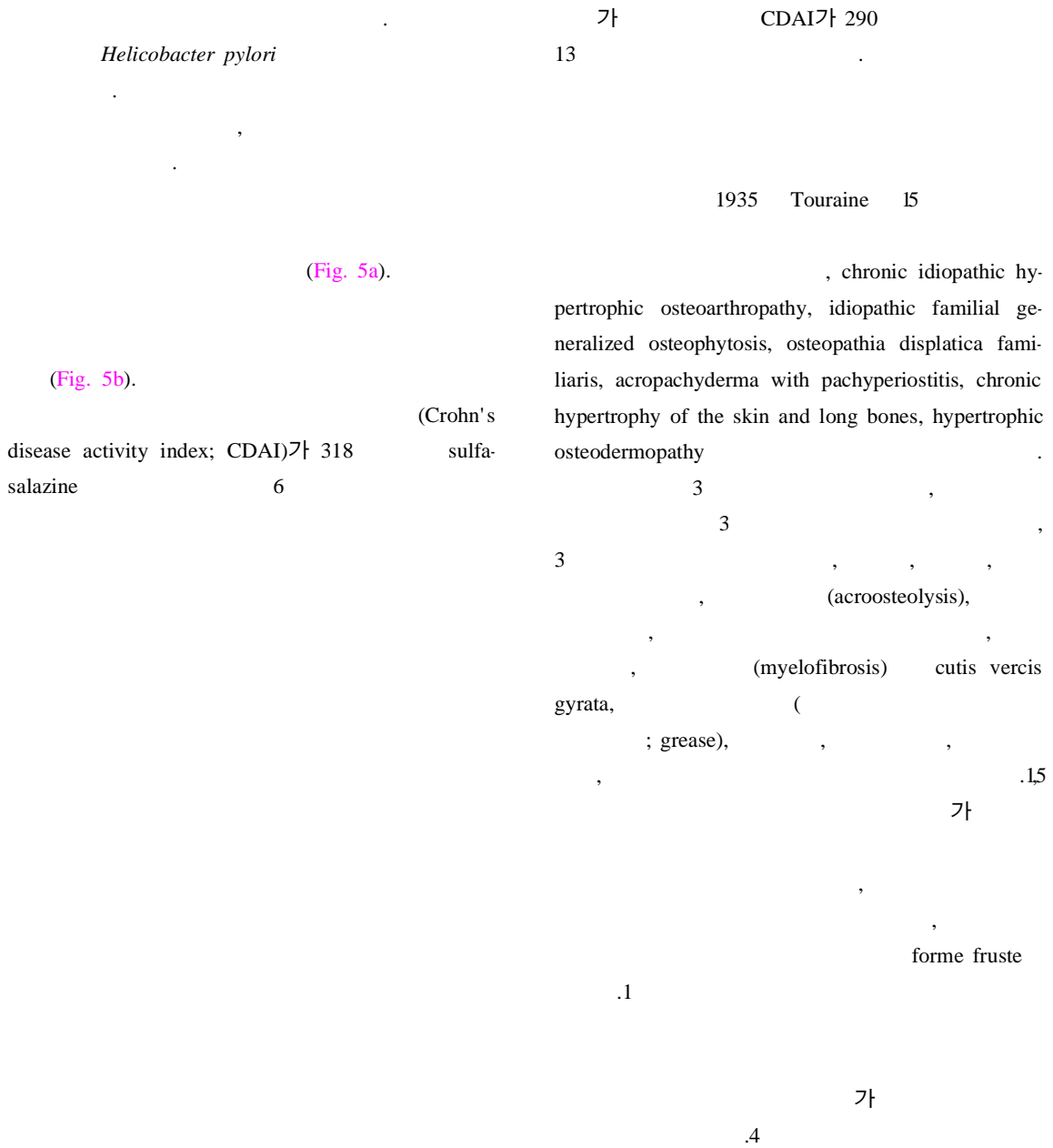


Fig. 5. Colonoscopic and microscopic appearances of the colon. (A) Multiple aphthous ulcers with erythematous and edematous margin are observed on the entire colon during colonoscopy. Mucosa between the ulcers were slightly edematous. (B) Histologic finding of biopsied colonic mucosa showing acute and chronic inflammation combined with crypt distortion (H&E stain, $\times 100$).

가 , 4 가
 가 , VDRL .10
 가 가
 가
 .21617 가
 8.9:1 ,
 (acidic mucopolysaccharides)
 osteocalcin 가
 (osteoblast) 가
 .18
 가
 .17 가
 가
 .69
 .10 1/3
 1/3 1/3
 가
 50% 가 15%
 가
 DR1 DQ5 HLA
 (tumor necrosis factor;
 TNF) (microsatellite)
 (intercellular adhesion mole-

cule; ICAM)-1 (polymorphism)
 .11
 38%
 가
 7 가 .1213
 가 3
 3
 6 20
 14
 4
 , 2
 가
 가

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