

A Case of Crohn's Disease Showing a Skin Lesion with a Cobblestone-like Appearance in the Perianal Region

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Crohn's disease, a chronic relapsing, multisystemic, inflammatory disorder, may involve any part of the gastrointestinal tract and shows a cobblestone-like appearance on intestinal mucosa. There are also extraintestinal features, including lesions of the skin, eye, and joints. Ulcers, fissures, sinus tracts, abscesses, and vegetant plaques have been reported for the perianal skin lesions of Crohn's disease.

We experienced a case of Crohn's disease in a 21-year-old Korean female showing a skin lesion with a cobblestone-like appearance in the perianal region.

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Key Words : Crohn's disease, Skin lesion, Cobblestone-like appearance

Crohn's disease is a chronic relapsing, multisystemic, inflammatory disorder of obscure etiology and may involve any part of the gastrointestinal tract from the mouth to anus, especially the terminal ileum.

There are also extraintestinal features, including lesions of the skin, eye, and joints¹. The skin is the most common site of extraintestinal involvement. The perianal, perineal, and peristomal areas are the most frequently affected sites¹. Ulcers, fissures, sinus tracts, abscesses, and vegetant plaques may extend in continuity from sites of intra-abdominal involvement to the perineum. A characteristic pathologic feature is the noncaseating granuloma which consists of lymphocytes, epithelioid cells and multinucleated giant cells.

The authors report a case of Crohn's disease that showed a skin lesion with a cobblestone-like ap-

pearance in the perianal region.

CASE REPORT

A 21-year-old female visited our dermatologic department with an 8-year history of erythematous plaque and skin-colored protruding masses in the perianal region. She had a prickling and itching sensation in the perianal region. She suffered from intermittent abdominal pain and diarrhea for 9 years. She had a low body weight(38kg), relative short stature(152cm), and irregular menstrual cycle.

She was presented with erythematous scaly plaques and cobblestone-like masses in the perianal region(Fig.1). The skin lesion enlarged gradually and showed oozing on the lesional surface. Her family and past medical history were non-contributory. On laboratory findings, hemoglobin and hematocrit were slightly decreased. LFT and urinalysis were within normal ranges. Colonoscopy, small bowel series and colon study showed luminal narrowing and cobblestone appearance, especially in the distal jejunum, terminal ileum, and ascending colon(Fig. 2, 3). The histopathologic findings of the perianal lesional skin and ascending colon showed noncaseating granulomatous reactions

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Fig. 1. Erythematous scaly plaque and protruding cobblestone-like masses in the perianal region.

Fig. 2. Colonoscopic finding shows severe stenosis surrounded by healing ulcers and inflammatory polyps at the proximal ascending colon. Sacular dilatation is also seen in the vicinity of stenosis.

Fig. 3. Colon study shows marked luminal narrowing with aggregated nodularities at the terminal ileum.

Fig. 4. Microscopic examination from protruding cobblestone-like mass in the perianal region shows non-caseating granulomatous lesions which consist of epithelioid cells and lymphocytes with a few giant cells(H&E, $\times 200$).

composed of lymphocytes, epithelioid cells, and a few giant cells in H & E stain(Fig. 4, 5). A skin biopsy was also performed with AFB stain and PAS stain. The results were negative.

On the basis of the clinical and laboratory findings, we were able to rule out tuberculosis, deep fungal infections, sarcoidosis, and other granulomatous disorders and make the diagnosis of Crohn's disease of the perianal region. After the patient was treated with systemic corticosteroid for three months, the

gastrointestinal symptom was slightly relieved, and the skin lesion was slightly regressed.

DISCUSSION

Crohn's disease is frequent in western countries, but it is rare in Korea. Associated mucosal and skin findings occur in 22% to 44% of patients^{1,2}, which include cutaneous Crohn's disease, contiguous perianal Crohn's disease, oral Crohn's disease, reactive skin findings, and nutritional skin changes^{1,7}.

Four types of cutaneous involvement have been described⁴. (1) direct extension from involved

Fig. 5. Microscopic examination from ascending colon shows the same histologic findings as the skin lesion (H&E, $\times 200$).

bowel leading to perineal ulceration, abscess, fistula, vegetant plaque, and skin tag^{1,2}; (2) extraintestinal involvement of the vulva, the submammary area, the penis, the abdominal wall, and the extremities; (3) vascular reactions leading to pyoderma gangrenosum, erythema nodosum, palmar erythema, and polyarteritis nodosa^{1,2}; (4) skin lesions secondary to nutritional deficiency such as acrodermatitis enteropathica (zinc deficiency)⁸.

In this case, the perianal skin lesions were supposed to be a direct extension from the involved bowel and appeared as cobblestone-like masses. Although various cutaneous lesions can occur on oral mucosa, perianal region, and vulva, the only well documented cobblestone appearance in patients with Crohn's disease is on the oral lesion^{1,3,9}. In Korean literature, about 130 cases of Crohn's disease have been reported, but extraintestinal manifestations were relatively rare compared with that of Caucasians¹⁰. Anal fistulas, ulcers, and fissures were reported as skin lesions in the perianal region^{11,12}, and cobblestone-like masses in the perianal region have not been reported.

The significance of the cutaneous manifestation of Crohn's disease is to serve as an early marker of the disease and it can aid in the diagnosis. Cutaneous changes may also help distinguish Crohn's disease from other inflammatory bowel diseases. When histologic evidence of granulomatous inflammation is found in the oral cavity, perianal region, or isolated ulcerated or nodose lesions, Crohn's disease is the likely diagnosis. Some nutritional changes, finger clubbing, and palmar ery-

thema should also point toward Crohn's disease¹.

The diagnosis of cutaneous Crohn's disease is generally obvious if the patient has associated gastrointestinal and perianal manifestations. However, the clinical differential diagnosis should include other granulomatous disorders such as cutaneous sarcoidosis, mycobacterial infections, actinomycosis, deep fungal infections, lymphogranuloma venereum, granuloma inguinale, cellulitis, chronic lymphedema resulting from obstruction, schistosomiasis, hidradenitis suppurativa, and foreign body reactions³.

The most effective treatment of cutaneous Crohn's disease is oral metronidazole^{3,13}. Other treatments include topical and intralesional corticosteroids, systemic steroids, sulfasalazine, cytotoxic agents such as azathioprine or 6-mercaptopurine, and surgery^{1,13,14}. Surgical removal of the diseased bowel does not necessarily improve cutaneous Crohn's disease¹⁵.

We report a patient with Crohn's disease, showing the cobblestone-like appearance in the perianal region as well as in the gastrointestinal tract.

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