Ann Dermatol Vol. 33, No. 4, 2021
pISSN 1013-9087 · eISSN 2005-3894
https://doi.org/10.5021/ad.2021.33.4.365

CASE REPORT

# Crohn's Disease with Cutaneous Polyarteritis Nodosa in a Child: A Case Report

Eun Hye Hong, Joon Woo Jung, Eun Joo Park, Kwang Joong Kim, Kwang Ho Kim

*Department of Dermatology, Hallym University Sacred Heart Hospital, Anyang, Korea*

A 10-year-old boy presented with a 1-day history of multiple painful erythematous skin lesions on his upper and lower extremities. He was admitted to the Department of Pediatrics with persistent right lower abdominal pain and diarrhea. Punch biopsy of a skin lesion on his lower leg showed necrotizing granulomatous vasculitis with septal panniculitis consistent with polyarteritis nodosa, and our differential diagnosis included cutaneous manifestations of Crohn's disease. Abdominal ultrasonography revealed distended colonic loops suggestive of inflammatory bowel disease. Upper and lower gastrointestinal endoscopy revealed lesions involving the duodenum, cecum, colon, and rectum. He developed multiple perianal fistulas during hospitalization. Additional laboratory tests revealed positive results for anti-saccharomyces cerevisiae and antinuclear antibodies. Based on his clinical presentation and laboratory findings, he was diagnosed with Crohn's disease associated with cutaneous polyarteritis nodosa. We report a rare case of a child who presented with cutaneous polyarteritis nodosa as an extraintestinal manifestation of Crohn's disease. **(Ann Dermatol 33(4) 365～368, 2021)**

**-Keywords-**
Crohn disease, Polyarteritis nodosa

Received November 12, 2019, Revised February 5, 2020, Accepted for publication March 2, 2020

**Corresponding author:** Kwang Ho Kim, Department of Dermatology, Hallym University Sacred Heart Hospital, 22 Gwanpyeong-ro 170beon-gil, Dongan-gu, Anyang 14068, Korea. Tel: 82-31-380-3765, Fax: 82-31-386-3761, E-mail: dermakkh@naver.com
ORCID: https://orcid.org/0000-0001-5315-6031



## INTRODUCTION

Crohn's disease is a form of chronic inflammatory bowel disease presenting with diarrhea, bleeding, and abdominal pain. Growth failure is often the earliest manifestation of Crohn's disease in children. This condition commonly affects the perianal area with formation of fistulas or abscesses. Its extraintestinal manifestations include ocular, hepatobiliary, and cutaneous complications[1]. Cutaneous manifestations vary from specific skin lesions such as metastatic Crohn's disease, skin lesions with histopathological features similar to Crohn's disease but without any association with the intestines, or reactive conditions secondary to chronic inflammation. The most common manifestation of reactive conditions is erythema nodosum and pyoderma gangrenosum. Notably, polyarteritis nodosa is a rare manifestation of Crohn's disease[2].

Polyarteritis nodosa is a rare cutaneous manifestation of inflammatory bowel disease, particularly in children, and the cutaneous symptoms may occur concomitantly with intestinal symptoms or precede the latter[3]. Histopathologically, polyarteritis nodosa presents as panniculitis, vasculitis, necrosis of vessels, fibrin deposition, and inflammatory cell infiltration. We report a rare case of a child presenting with cutaneous polyarteritis nodosa as an extraintestinal manifestation of Crohn's disease.

## CASE REPORT

A 10-year-old boy presented with painful erythematous skin lesions on his extremities (Fig. 1). He was admitted to the Department of Pediatrics with crampy abdominal pain and diarrhea. Multiple erythematous tender nodules were scattered on his extremities with a heating sensation and joint pain. The patient's past medical history revealed growth failure. Laboratory tests showed increased white

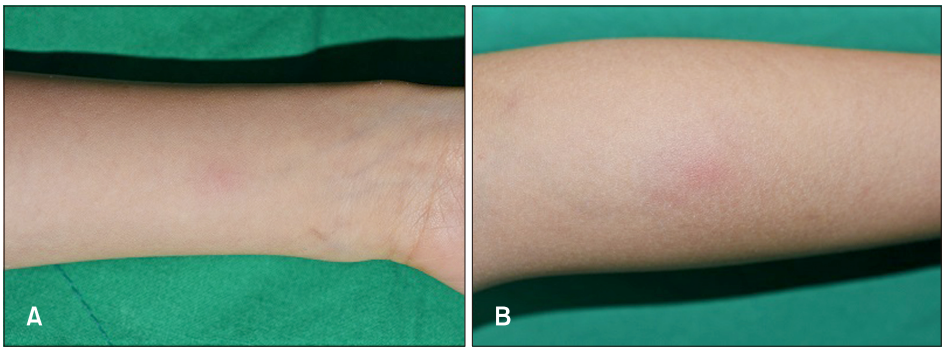


**Fig. 1.** Multiple painful erythematous nodules on the forearm (A) and lower leg (B).

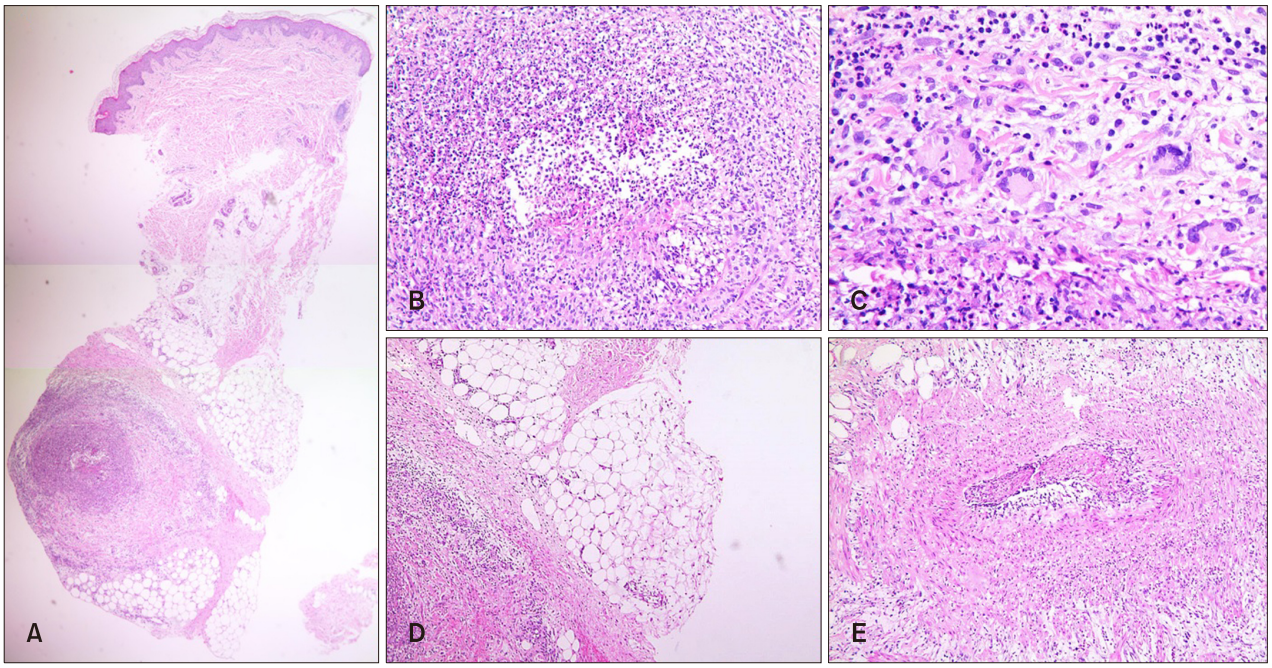


**Fig. 2.** (A) Low magnification of histologic findings of the lesions on the forearm (H&E, ×40). Histopathologic findings of the lesions on the forearm (B∼D) and lower leg (E). (B) Necrosis of blood vessel with neutrophil infiltration in deep dermis (H&E, ×200). (C) Granulomatous inflammation accompanying multinucleated giant cells around necrotizing vessel (H&E, ×400). (D) Septal panniculitis (H&E, ×100). (E) Necrosis of blood vessel with fibrin deposits and surrounding inflammatory cells in deep dermis (H&E, ×100).

blood cells, an elevated erythrocyte sedimentation rate, serum C-reactive protein and platelets, and decreased hemoglobin. Stool tests showed increased calprotectin levels. Abdominal ultrasonography revealed diffuse thickening of the colonic wall, and he was treated symptomatically for suspected enteritis. He underwent a punch biopsy of a skin lesion. Examination of a forearm lesion showed necrosis of blood vessels (Fig. 2A, B) and surrounding granulomatous inflammation with accompanying neutrophils and multinucleated giant cells (Fig. 2C). The fat layer showed inflammatory cell infiltration into the septum (Fig. 2D). Histopathological examination of a specimen obtained from his lower leg also revealed perivascular inflammatory cell infiltration in the deep dermis and subcutis, necrosis of blood vessels with fibrin deposits and surrounding neutrophils (Fig. 2E). Evidence of granulomatous vasculitis with panniculitis on histopathological examination led to a high index of clinical suspicion for cutaneous polyarteritis nodosa. Considering his persistent abdominal pain and diarrhea, we could not exclude cutaneous Crohn's disease, and further evaluation for pediatric inflammatory bowel disease was performed. Repeat abdominal ultrasonography revealed distended colonic loops suspicious for inflammatory bowel disease. Upper and lower gastrointestinal endoscopy revealed lesions involving the duodenum, cecum, colon, and rectum. Laboratory investigations revealed positive results for anti-saccharomyces cerevisiae antibodies, and we detected a

perianal fistula. Based on his clinical presentation and the aforementioned investigations, he was diagnosed with Crohn's disease presenting with an extraintestinal manifestation of cutaneous polyarteritis nodosa. He was then transferred to an another hospital and got treatments including low dose prednisolone. His skin lesions and symptoms showed a waxing and waning pattern.
We received the patient's consent form about publishing all photographic materials.

## DISCUSSION

Crohn's disease is a form of chronic inflammatory bowel disease that can affect any part of the gastrointestinal tract and usually shows a clinical presentation of diarrhea, abdominal pain, and bleeding. Pediatric Crohn's disease often manifests with fever and growth failure as the initial symptoms. In laboratory tests, indicators of inflammation such as white blood cells, erythrocyte sedimentation rate and serum C-reactive protein can be elevated. A positive result of anti-saccharomyces cerevisiae antibody also has a diagnostic value of Crohn's disease. Perianal fistulas and abscess are common, and involvement of the skin, eyes, or liver can occur as extraintestinal manifestations[1]. In this case, the patient manifested with growth failure, which may be an initial symptom of underlying Crohn's disease.
Kurtzman et al.[2] reported that cutaneous manifestations of Crohn's disease can be categorized as specific lesions directly associated with Crohn's disease, reactive and associated conditions, nutritional deficiency-induced lesions, as well as therapy-induced lesions. Metastatic Crohn's disease is directly associated with intestinal Crohn's disease that presents with erythematous plaques, nodules, and ulceration. Histopathological features of noncaseating granulomatous inflammation involving the papillary and reticular dermis are observed in intestinal as well as cutaneous lesions of Crohn's disease[2]. Erythema nodosum is the most common reactive cutaneous manifestation of Crohn's disease that presents with painful and indurated erythematous plaques on the extremities. Histopathological examination of these lesions shows septal panniculitis without vasculitis[4]. Pyoderma gangrenosum is also a relatively common condition; however, polyarteritis nodosa is rare, particularly in children.
Polyarteritis nodosa is a rare cutaneous manifestation of inflammatory bowel disease, which may precede or coincide with the onset of intestinal manifestations[3]. Histopathological examination is not different from polyarteritis nodosa without Crohn's disease. It shows vasculitis with panniculitis and necrosis of vessels with fibrin deposition. Previous studies[5] have reported polyarteritis nodosa as a cutaneous manifestation of Crohn's disease. Of the 15 cases reported, skin lesions preceding Crohn's disease developed in 3[6-8]. In our patient, skin lesions consistent with polyarteritis nodosa occurred almost simultaneously with intestinal symptoms. Considering the patient's abdominal symptoms accompanied by cutaneous polyarteritis nodosa, we suspected underlying inflammatory bowel disease in this case and performed further evaluation for inflammatory bowel disease. Additional examination led to a diagnosis of Crohn's disease.
Polyarteritis nodosa can be associated with hepatitis B or C virus infection as well as Crohn's disease. As vaccination against hepatitis B virus is routinely conducted, this association has been weakened[9]. However, it must be determined whether it is associated with hepatitis B or C virus infection before starting treatment for polyarteritis nodosa.
Crohn's disease may clinically present with a variety of cutaneous lesions. Clinicians should be aware that the cutaneous manifestations of Crohn's disease may not always occur simultaneously with abdominal symptoms and that these may precede the onset of Crohn's disease. Patients with suspected skin lesions must undergo thorough evaluation to assess abdominal pain and/or diarrhea. If needed, further evaluation should be performed to confirm underlying inflammatory bowel disease. We report a rare case of Crohn's disease in a child presenting with an extraintestinal manifestation of cutaneous polyarteritis nodosa. Diagnostic evaluation was recommended by the attending dermatologist for suspected pediatric inflammatory bowel disease.

## CONFLICTS OF INTEREST

The authors have nothing to disclose.

## FUNDING SOURCE

None.

## ORCID

Eun Hye Hong, https://orcid.org/0000-0002-3566-1007
Joon Woo Jung, https://orcid.org/0000-0001-8977-7980
Eun Joo Park, https://orcid.org/0000-0002-9924-515X
Kwang Joong Kim, https://orcid.org/0000-0003-4158-6100
Kwang Ho Kim, https://orcid.org/0000-0001-5315-6031

## REFERENCES

1. Kliegman R, Stanton B, St Geme III J, Schor N, Behrman R.

Nelson textbook of pediatrics. 19th ed. Philadelphia: Elsevier Saunders, 2011:1300-1304.
2. Kurtzman DJ, Jones T, Lian F, Peng LS. Metastatic Crohn's disease: a review and approach to therapy. J Am Acad Dermatol 2014;71:804-813.
3. Pellicer Z, Santiago JM, Rodriguez A, Alonso V, Antón R, Bosca MM. Management of cutaneous disorders related to inflammatory bowel disease. Ann Gastroenterol 2012;25: 21-26.
4. Hagen JW, Swoger JM, Grandinetti LM. Cutaneous manifestations of Crohn disease. Dermatol Clin 2015;33:417-431.
5. Komatsuda A, Kinoshita K, Togashi M, Maki N, Masai R, Niitsu H, et al. Cutaneous polyarteritis nodosa in a patient with Crohn's disease. Mod Rheumatol 2008;18:639-642.
6. Dyer NH, Verbov JL, Dawson AM, Borrie PF, Stansfeld AG. Cutaneous polyartheritis nodosa associated with Crohn's disease. Lancet 1970;1:648-650.
7. Gudbjörnsson B, Hällgren R. Cutaneous polyarteritis nodosa associated with Crohn's disease. Report and review of the literature. J Rheumatol 1990;17:386-390.
8. Voulgarelis M, Chorti M, Kittas C, Karachristos A, Ikonomopoulos G, Skopouli FN. Fever and abdominal pain in a 45-year-old woman with cutaneous necrotising vasculitis. Clin Exp Rheumatol 1998;16:72-76.
9. Guillevin L, Mahr A, Callard P, Godmer P, Pagnoux C, Leray E, et al. Hepatitis B virus-associated polyarteritis nodosa: clinical characteristics, outcome, and impact of treatment in 115 patients. Medicine (Baltimore) 2005;84: 313-322.