

Castleman disease-like mesenteric lymphadenopathy in a Korean adult ultimately diagnosed with familial Mediterranean fever

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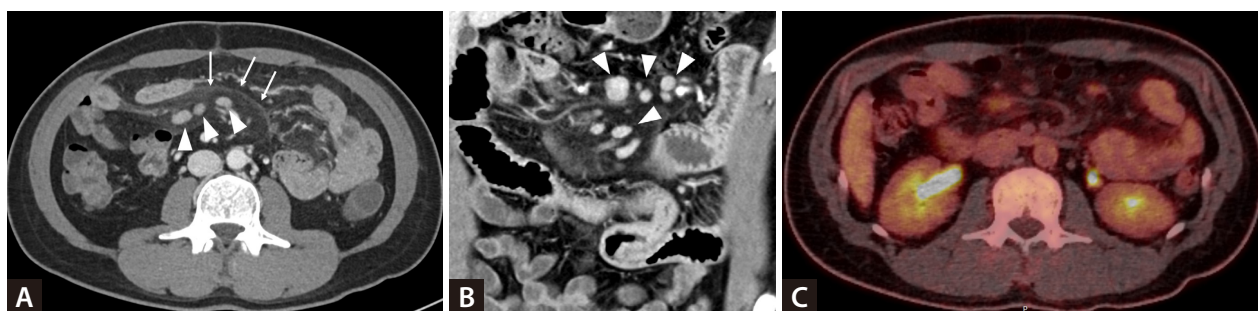


Figure 1. Radiologic findings of mesenteric lymphadenopathy. Axial (A) and coronal (B) images from contrast-enhanced abdominal computed tomography (CT) reveal multiple homogeneously enhancing enlarged mesenteric lymph nodes (arrowheads) measuring up to 1.2 cm in short-axis diameter, accompanied by engorged mesenteric vessels and thickened mesenteric folds (white arrows). (C) Fluorodeoxyglucose (FDG) positron emission tomography–CT obtained during a symptom-free interval shows only minimal FDG uptake in the mesenteric lymph nodes (SUVmax 3.1–3.4), indicating low metabolic activity outside of acute inflammatory attacks.

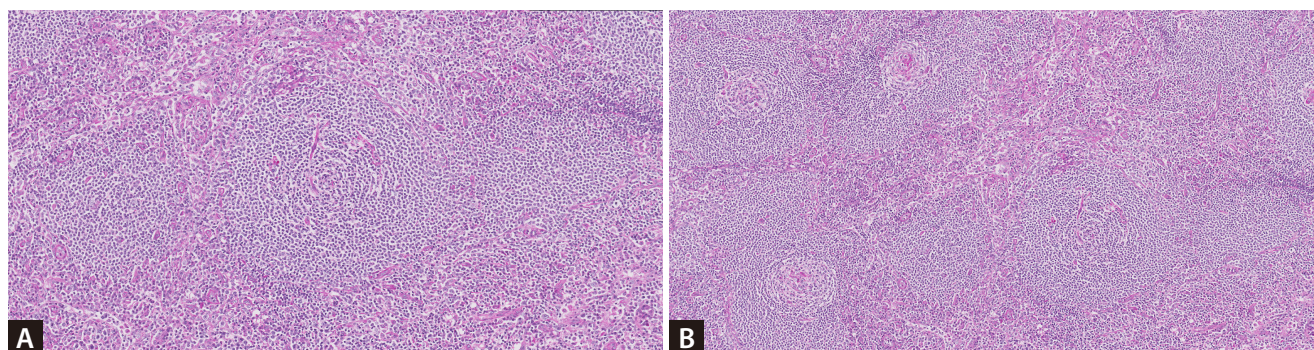


Figure 2. Histological findings of the mesenteric lymph node. The lymph node biopsy reveals atretic germinal centers infiltrated by hyalinized vessels, resulting in the formation of "lollipop follicles" (A) (H&E, $\times 100$). Additionally, the mantle zones are thickened, exhibiting an onion-skin appearance (B) (H&E, $\times 40$).

A 47-year-old Korean man was referred for evaluation of recurrent febrile episodes accompanied by abdominal pain. Since his early 30s, he had experienced attacks every two

to three months, with fevers up to 38.4°C lasting three days and periumbilical abdominal pain persisting about a week. Each episode was associated with elevated inflam-

matory markers, including C-reactive protein up to 20.03 mg/dL and erythrocyte sedimentation rate up to 46 mm/h, while symptom-free intervals showed no abnormalities. He reported no arthralgia, rash, or serositis.

Repeated abdominal computed tomography (CT) scans revealed multiple enlarged mesenteric lymph nodes with homogeneous enhancement, engorged mesenteric vessels, and thickened mesenteric folds (Fig. 1A, B), raising suspicion for Castleman disease. An excisional biopsy at an outside hospital was initially interpreted as “consistent with Castleman disease,” leading to a provisional diagnosis of idiopathic multicentric Castleman disease and a recommendation for siltuximab therapy. However, the patient’s periodic febrile attacks were atypical for Castleman disease. In addition, fluorodeoxyglucose positron emission tomography–CT obtained during a symptom-free interval demonstrated only minimal metabolic activity in the mesenteric lymph nodes (Fig. 1C), prompting reconsideration of the diagnosis.

Pathologic re-evaluation indicated reactive lymphoid hyperplasia with focal Castleman-like features, including atretic germinal centers with penetrating hyalinized vessels and thickened mantle zones exhibiting an onion-skin appearance (Fig. 2). Given the long-standing relapsing inflammatory episodes, periodic fever syndromes were considered [1]. Next-generation sequencing identified heterozygous P369S and R408Q variants in the *MEFV* gene, both classified as variants of uncertain significance [2]. A diagnosis of familial Mediterranean fever (FMF) was established by integrating the pattern of recurrent fever, abdominal pain, reactive mesenteric lymphadenopathy, and *MEFV* variants [3]. Colchicine therapy led to complete remission of attacks and normalization of inflammatory markers within six months. This case illustrates that FMF, although rare in Korea, can radiologically and pathologically resemble Castleman disease.

REFERENCES

1. Lachmann HJ. Periodic fever syndromes. *Best Pract Res Clin Rheumatol* 2017;31:596-609.
2. Zadeh N, Getzug T, Grody WW. Diagnosis and management of familial Mediterranean fever: integrating medical genetics in a dedicated interdisciplinary clinic. *Genet Med* 2011;13:263-269.
3. Lancieri M, Bustaffa M, Palmeri S, et al. An update on familial Mediterranean fever. *Int J Mol Sci* 2023;24:9584.

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Conflicts of interest

The authors disclose no conflicts.

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Ethical statement

This case was approved by the Institutional Review Board of Seoul St. Mary’s Hospital (IRB No. KC25ZISI0903). Written informed consent was waived.