



Intralymphatic Histiocytosis Associated with Osteoarthritis: A Case Report

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Intralymphatic histiocytosis (ILH) is a rare cutaneous condition with uncertain pathogenesis. It is characterized by dilated lymphatic vessels that contain histiocytes within their lumina. Although the etiology of ILH remains unknown, it has been associated with various inflammatory and neoplastic diseases, such as rheumatoid arthritis (RA), reaction to metal joint implants and Merkel cell carcinoma, breast cancer and colon cancer. An 83-year-old female presented with an erythematous patch on the left forearm that had appeared six months previous. She had suffered from osteoarthritis (OA) and the cutaneous lesion was located in the vicinity of the affected joint. Skin biopsy from the lesion showed dilated dermal vessels and some ectatic vessels that contained many mononuclear histiocytes. Based on the clinical and histopathological findings, we diagnosed her with ILH with OA. Two sessions of intralesional triamcinolone acetonide injection (5 mg/ml) were administered to treat the skin lesion, which gradually improved over a period of a few months. We here report a rare case of ILH associated with degenerative OA.

Keywords: Intralymphatic histiocytosis, Osteoarthritis

INTRODUCTION

Intralymphatic histiocytosis (ILH) is a rare, chronic, cutaneous condition characterized by dilated lymphatic vessels that contain collections of histiocytes within their lumina. The cutaneous lesions have various presentations, including erythematous plaques, patches, excoriated papules, and livedo reticularis-like lesions. Such lesions are predominantly located on the upper and lower limbs¹. Although the etiology of ILH remains unknown, it has been associated with various inflammatory and neoplastic diseases, including rheumatoid arthritis (RA); reaction to metal joint implants; and Merkel cell carcinoma, breast cancer, and colon cancer¹⁻⁵. We describe a rare case of ILH in a patient with degenerative osteoarthritis (OA).

CASE REPORT

An 83-year-old female presented with an erythematous patch

on the left forearm that had appeared six months previous. She had no history of any other disease except chronic degenerative OA of the elbows, and the cutaneous lesion was located in the vicinity of the affected joint. Physical examination revealed a relatively well demarcated erythematous patch on the left elbow to forearm, and the affected skin was warm to touch (Fig. 1A, B). The patient reported no subjective symptoms regarding the skin lesion, and she only complained of arthralgia due to OA of the left elbow. Laboratory evaluations showed increased levels of erythrocyte sedimentation rate (112 mm/h) and C-reactive protein (31.7 mg/L). RA factor was normal (<10.80). Skin biopsy from the lesion showed numerous irregular dilated vessels throughout the dermis (Fig. 2A) and some ectatic vessels that contained many mononuclear histiocytes (Fig. 2B). Immunohistochemically, the aggregated histiocytes were positive for CD68 (Fig. 2C), and the most dilated vessels were positive for D2-40, a selective marker for lymphatic endothelium (Fig. 2D). After diagnosis of ILH, the patient was inves-



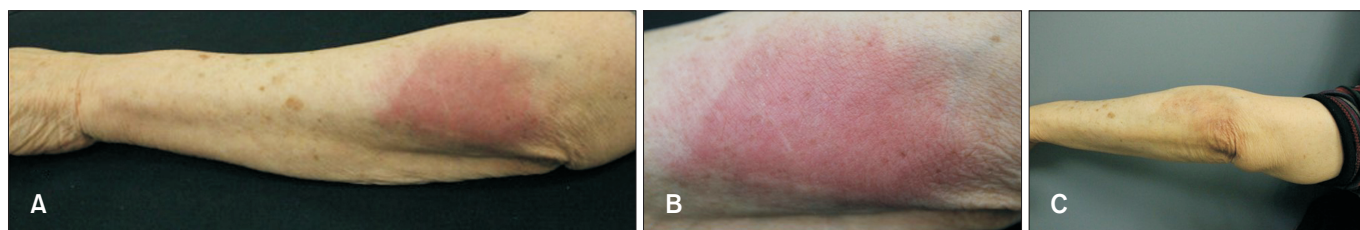


Fig. 1. Poorly demarcated erythematous patch on left forearm (A), close-up view of the lesion (B), 2 months after steroid intralesional injection (C).

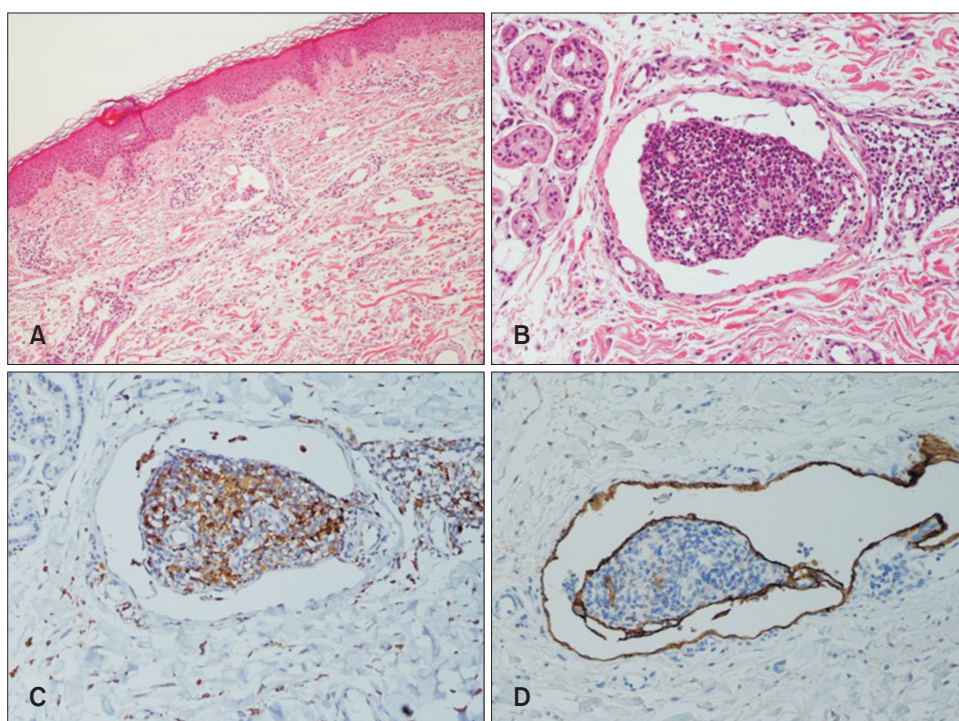


Fig. 2. Numerous irregular dilated vessels throughout the dermis (H&E, ×40) (A), dilated vessels with intraluminal collections of mononuclear cells (H&E, ×400) (B), histiocytes stained strongly positive for CD68 (CD68, ×400) (C), endothelial cells of a dilated vessel positive for D2-40 (D2-40, ×400) (D).

tigated for other systemic diseases. No concomitant diseases were found. Two sessions of intralesional triamcinolone acetate injection (5 mg/ml) were used to treat the skin lesion, which gradually improved over a period of a few months (Fig. 1C). We received the patient's consent form about publishing all photographic materials.

DISCUSSION

ILH, also known as intravascular histiocytosis, is a rare cutaneous condition first described by O'Grady et al.⁶ in 1994. Initially, there was controversy over whether the vessels were blood vessels or lymphatic vessels, but additional immunohistochemical markers characteristic of lymphatic endothelium, such as D2-40, podoplanin, Lyve-1, and Prox-

1, confirmed that most of the dilated vessels were lymphatic^{1,2,4,5}. ILH is usually associated with underlying chronic inflammatory disorders or neoplastic diseases, such as rheumatoid arthritis, Crohn's disease, Merkel cell carcinoma, breast cancer, and reaction to metal joint implants^{1-5,7-9}. In particular, there are many cases associated with rheumatoid arthritis. In the largest published review of ILH, almost half of the 42 patients had rheumatoid arthritis, and the lesions were located in the vicinity of the affected joint¹⁰. Although the cause of ILH in relation to chronic inflammatory disease is still being discussed, Requena et al.⁴ suggested that it is caused by lymphangiectasis resulting from obstruction of lymphatic drainage due to congenital abnormal vessel development or acquired damage of lymphatic vessels from infection, trauma, surgery, or radiation. Lymphatic stasis

may lead to poor clearance of an antigen, localized immune dysfunction, and persistent inflammation. Aggregations of histiocytes within the lymphatic vessels may indicate the presence of a persistent antigen that stimulates histiocytes to proliferate and aggregate. Pruim et al.¹¹ explained that ILH is due to lymphatic drainage of histiocytes from inflamed joints, which explains why ILH is often associated with arthritis. ILH associated with degenerative OA has been very rarely reported, and to our knowledge, this is the third report of ILH associated with OA². However, there may be under-reported cases not recognized due to either the rarity of OA-related cutaneous manifestations or an unfamiliar association between them.

ILH, more common in female than in male, can present as poorly demarcated erythematous patches, plaques, papules, and nodules and also can be seen as livedo-like lesions. Three cases which have been reported in Korea also presented in the form of erythematous patches or plaques as shown in this case⁷⁻⁹. No effective treatment of ILH has been established. In addition, there is a lack of evidence that indicates improvement in skin lesions when the associated disease improves. In many cases, a topical steroid was tried but had little effect. The patient in the present case was successfully treated with triamcinolone acetonide intralesional injections. Since ILH is a problem of the dermal lymphatics, it was assumed that an intralesional injection, which directly delivered the drug to the dermis, would be effective, unlike topical applications.

ILH clinically manifests as erythematous patch or plaque lesions and histopathologically demonstrates intralymphatic aggregations of histiocytes, and it may occur around a joint with chronic inflammation. Therefore, it is important for physicians to consider a histologic examination for a characteristic rash around a joint with chronic inflammation, preventing patients from undergoing further unnecessary diagnostic testing.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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