Case Report
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대한영상의학회지

A Case of Eosinophilic Fasciitis: Imaging Findings from Early Diagnosis to Complete Remission
호산구성 근막염 1예: 조기 진단에서 완치까지의 영상 소견

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Eosinophilic fasciitis (EF) is a rare disease characterized by symmetric and painful swelling with progressive induration and thickening of the skin and soft tissues of the limbs and trunk. Herein, we report a case of a 31-year-old woman who presented with painful swelling in both lower legs which persisted for 6 days. She underwent ultrasonography (US) in an out-patient department to rule out deep-vein thrombosis. The US did, however, reveal perifascial fluid in the thickened superficial fascia and interstitial fluid in the subcutaneous layer of both lower legs. Magnetic resonance imaging findings were identical to the US and additionally showed no involvement of the muscles or deep fascia. Laboratory data, showing peripheral eosinophilia and a US-guided gun-biopsy showing lymphocytic and eosinophilic infiltration were both indicative of EF. The patient was treated with corticosteroids, resulting in a remarkable improvement in both the lower-leg edema and peripheral eosinophilia. There was no recurrence after 7 years of follow-up.

Index terms
Eosinophilia
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Ultrasonography
MRI

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INTRODUCTION

Eosinophilic fasciitis (EF) is a rare disease of unknown etiology and pathogenesis manifesting in a scleroderma-like clinical presentation. The characteristic laboratory findings are peripheral eosinophilia, hypergammaglobulinemia and an elevated erythrocyte sedimentation rate, first described by Shulman. A full-thickness wedge biopsy of the clinically affected skin is essential for an accurate diagnosis. Almost all cases of EF show a good responsiveness to corticosteroid therapy. Notwithstanding the demonstrated effectiveness of solitary magnetic resonance imaging (MRI) for the diagnosis of EF, herein we report a case of EF from early diagnosis to complete remission. The report focuses on the illustrative findings of combined ultrasonography (US) and MRI findings, with a literature review.

CASE REPORT

A 31-year-old woman visited our hospital with painful lower-leg swelling persisting for 6 days. She showed no fever, dyspnea or chest pain and was not on any medication. A review of her medical history revealed neither a recent trauma or allergy nor any cardiac or other disease. On physical examination, both lower legs showed diffuse pitting edema without skin discoloration or heating sensation. An US (iU 22 unit; Philips Medical Systems, Bothell, WA, USA), performed on an out-patient basis ruled out an acute deep-vein thrombosis, but showed interstitial fluid in the thickened subcutaneous layer and perifascial fluid along the superficial fascia of both lower legs (Fig. 1A). However, no significantly enlarged lymph nodes or intramuscular focal lesion was observed in either lower extremity. The preliminary

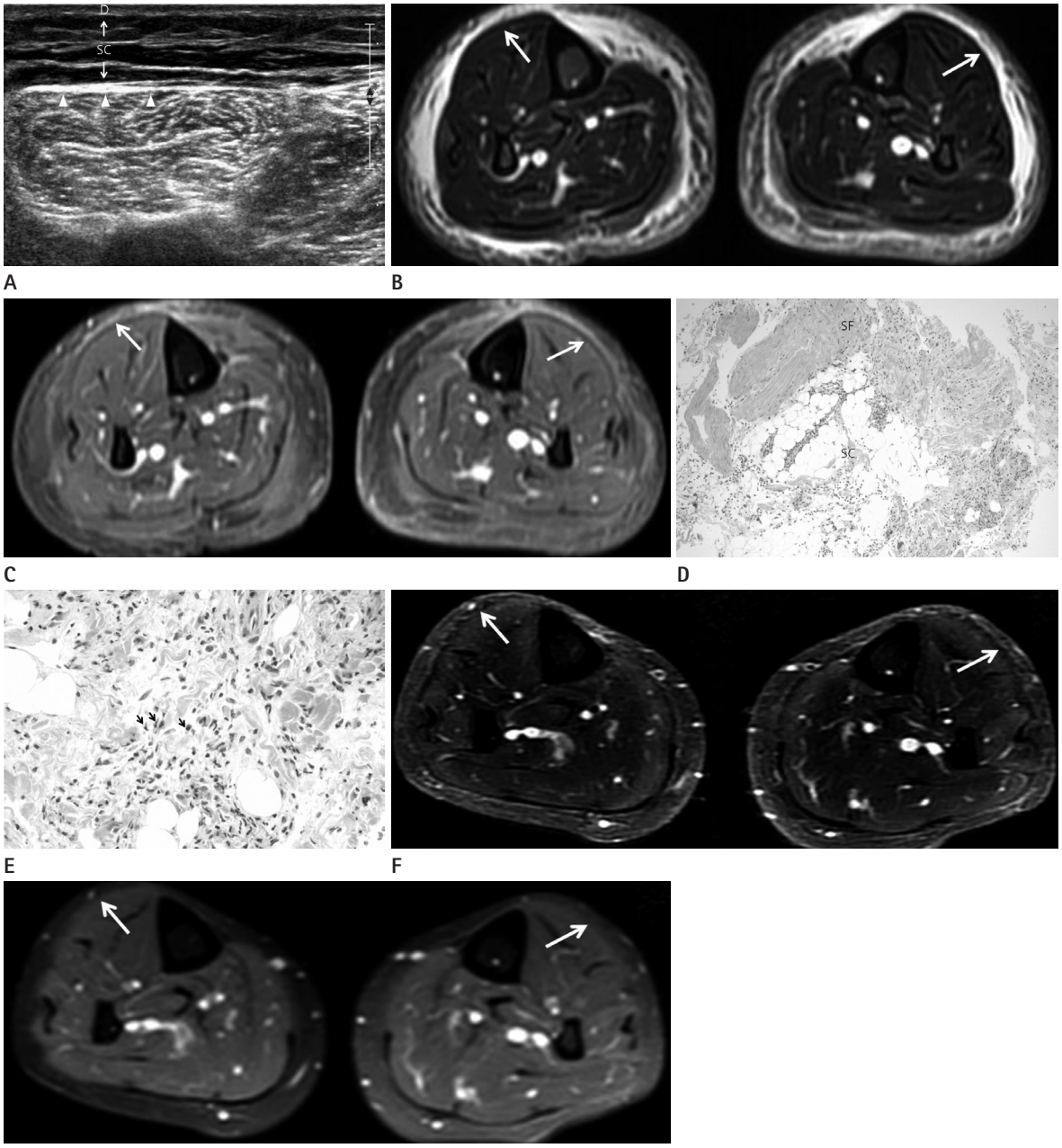
sonographic diagnosis was infectious or inflammatory fasciitis rather than venous congestion. The initial laboratory findings included a white blood cell count (WBC) of 33650 (normal; 4000–10000) cells/μL, with 68% (normal; 1–4%) eosinophils. The erythrocyte sedimentation rate was 4 (normal; 1–4) mm/hr and the C-reactive protein level was 0.31 mg/dL. Other laboratory data, including results of the biochemical study were normal. Due to the negative result of stool exam, a parasitic infection owing to the patient's peripheral eosinophilia was excluded. A thyroid test revealed a normal function. The results of an electrocardiogram and a pulmonary function test were normal as well. The patient was negative for rheumatoid factor and antinuclear antibody. Four days later, the patient underwent MRI (MR Signa Excite; GE Healthcare, Milwaukee, WI, USA). On axial fat-suppressed T2-weighted imaging (T2WI) (Fig. 1B), the fluid-signal intensity in the subcutaneous fat layer and superficial fascia of both lower legs suggested diffuse inflammatory changes. The fat-suppressed enhanced T1-weighted imaging (Fig. 1C) also showed a mild superficial fascial enhancement corresponding with the T2WI and US locations. On US and MRI, no focal lesion was detected either in the muscles or deep fascia of the lower legs. Considering the laboratory and imaging findings of prominent fascial involvement and peripheral eosinophilia, EF was regarded as a first differential diagnosis. An US-guided gun biopsy was subsequently performed on the right lower leg and the specimen showed fascial thickening by an inflammatory infiltration of lymphocytes, plasma cells and eosinophils (Fig. 1D, E), sufficient for the confirmative diagnosis. Accordingly, the treatment was started with prednisolone (40 mg daily). After two days of treatment, the symmetric pitting edema had dramatically improved the differential eosinophil count had sharply decreased from 28.9% of WBC (17050 cells/μL) to 4% (9700). The prednisolone course was tapered until the 10th week and was halted thereafter. After the 15th week, a relapse occurred and the patient complained of mild, doughy swelling in both lower legs upon revisiting the hospital. The differential eosinophil count had increased to 19% of WBC (15912 cells/μL). An oral steroid therapy was started once again, gradually tapered and discontinued for 3 months. 6 months later, the follow-up MRI showed an improved fasciitis without residual fibrosis (Fig. 1F, G). The patient remained free of symptoms as she continued to be monitored on an out-patient basis over the course of a long-term follow-up period (7 years).

DISCUSSION

EF is an uncommon scleroderma-like disease known as Shulman's syndrome or diffuse fasciitis with eosinophilia (1). EF can occur at any age and the incidence rates by gender are roughly equal. EF's clinical presentation typically entails an abrupt onset of painful and erythematous swelling of the trunk and extremities, though variable and nonspecific. EF can affect both the upper and lower extremities, usually with bilateral symmetric involvement. Patients might initially complain of swelling and, later, weakness and thickening of the skin, which, eventually, is firmly bound to the underlying tissue in refractory fibrosis. EF usually is benign; according to reports, 70–90% of patients respond well to steroid treatment (2, 3).

An early diagnosis of EF, prior to the confirmative diagnosis by full-thickness skin-to-muscle biopsy, can be established on the basis of peculiar laboratory abnormalities such as peripheral eosinophilia, hypergammaglobulinemia, elevated erythrocyte sedimentation rate and characteristic imaging patterns of prominent fascial thickening, hyperintense signals within the fascia on fluid-sensitive sequences and fascial enhancement after IV-contrast administration on MRI (4).

In the present case, fascial thickening with EF was confirmed by an US-guided biopsy, even though the full-thickness skin-to-muscle biopsy is known to be a confirmative method for diagnosis. The radiologic, clinical and pathologic findings were consistent with EF. However, the inflammatory changes involving the superficial fascia and subcutaneous fat layer, but not involving the deep fascia or muscle might be indicative of an acute-phase of EF prior to the formation of fibrosis or a morphea-like lesion of the skin as reported in previous cases (5-7). Although a relapse occurred following the cessation of medication, an early restart of medication might have helped to prevent a refractory fibrosis. Previous EF studies just noted the described fascial thickening and enhancement in the superficial and deep fascia (on enhanced MRI) and an induration or morphea-like skin thickening (on physical examination). In those cases lasted the symptom duration several months (5-7). In our presented case, the patient experienced duration of symptoms for six days only before medication start and showed a complete remission with



A B C D E F G

Fig. 1. Eosinophilic fasciitis (EF) in both lower legs of 31-year-old woman.
A. Ultrasonography (US) of right-lower leg shows diffuse interstitial fluid in thickened subcutaneous layer (space between arrows) and superficial fascia (arrowheads). There is no intramuscular focal lesion or perifascial fluid in the deep fascia, nor is there any evidence of deep-vein thrombosis (not shown).
B, C. Magnetic resonance imaging (MRI) of both lower leg shows interstitial fluid (**B**, arrows) in subcutaneous layer and superficial fascia on fat-suppressed T2-weighted image **(B)** and subtle linear enhancement (**C**, arrows) on fat-suppressed contrast-enhanced T1-weighted image **(C)**. There is no signal change or enhancement within the muscle or deep fascia.
D. Photomicrograph (H&E stain, × 100) of fascia-subcutaneous fat junction showing thickened fascia and entrapment of subcutaneous fat by heavy inflammatory cell infiltration.
E. High-power photomicrograph (H&E stain, × 400) of fascia showing heavy inflammatory cell infiltration with numerous eosinophils (arrows), lymphocytes and occasional plasma cells.
F, G. MRI was obtained after 6 months of corticosteroids treatment. Fat-suppressed T2-weighted image **(F)** and fat-suppressed contrast-enhanced T1-weighted image **(G)** showing complete remission. The patient is symptom-free, and the laboratory findings are normal. The arrows show the sites of abnormalities identified in a previous MRI.
Note.—D = dermal layer, SC = subcutaneous layer, SF = superficial fascia

no refractory fibrosis if uniquely compared with those previous studies. Lee et al. (8) reported two cases with no residual sequelae very similar to our case. The complete remission of EF in the present case might have resulted from the early steroid treatment as well as the gradual tapering to prevent relapse. In any case, the monitoring of disease activity, besides symptoms, should be performed with respect to peripheral eosinophilia and by MRI.

The diagnosis of acute-phase EF can be facilitated by exclusion of other diseases with acute limb swelling, such as cardiac or renal disease, acute deep-vein thrombosis, infection (cellulitis, necrotizing fasciitis, pyomyositis), inflammatory processes mimicking skin infections (dermatomyositis, scleroderma, graft-versus-host disease), tumors mimicking soft-tissue inflammatory disease (lymphoma), rhabdomyolysis or acute muscle denervation. The MRI provides information crucial to the determination of the depth of soft-tissue involvement, which supplements laboratory data and contributes to differential diagnosis. For example, in renal and cardiac diseases, fascial thickening is absent and in dermatomyositis and the scleroderma, the fascia usually is not involved. In case of tumors mimicking an inflammatory disease of soft tissue (lymphoma), an infiltrating or circumscribed mass is shown; in rhabdomyolysis and acute muscle denervation, the main characteristic abnormality is noted in the muscle. Meanwhile, the US plays the main role in the identification of a fascial abnormality and the exclusion of several diseases such as deep-vein thrombosis, myositis and pyomyositis (9). In the presented case for example, the initial US ruled out a deep-vein thrombosis.

Most previously as EF reported cases showed a relative long duration of symptoms from months to years which we call chronic. In those cases, the imaging findings were thickening of both superficial and deep fascias. In our experienced case, the time interval between the initial manifestation of symptoms to the imaging work-up was short compared with previous cases, resulting in acute to subacute imaging findings; interstitial edema and superficial fascial thickening, resembling a case reported before (8).

In summary, we report a case of EF with early diagnosis and complete remission. Even though the imaging-based diagnosis of EF is challenging due to its rarity in cases of acute limb swelling, cross-sectional imaging modalities such as MR and US are helpful for the early diagnosis and monitoring of EF along with the characteristic laboratory findings.

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호산구성 근막염 1예: 조기 진단에서 완치까지의 영상 소견

이재욱 · 이지영

호산구성 근막염(eosinophilic fasciitis)은 사지와 몸통의 대칭적인 부종과 지속적으로 진행하는 피부와 연부조직의 경화와 비후를 특징으로 하는 드문 질환이다. 31세 여자 환자는 6일 동안 지속된 하지의 부종 및 통증을 주소로 본원 외래로 내원하였다. 심부정맥혈전증을 감별하기 위해 초음파 검사를 시행하였고, 초음파 검사상 양측 종아리에서 표재성 근막의 비후 및 근막주변부의 수분저류를 관찰하였다. 자기공명영상검사상 초음파 검사와 동일한 소견을 보였고, 근육 및 심부근막에는 이환되지 않았음을 추가로 확인하였다. 혈액검사 결과상 말초 호산구 증가 소견을 보였고, 초음파 유도하 총 생검상 림프구와 호산구의 침착을 확인하여 호산구성 근막염으로 진단할 수 있었다. 환자는 스테로이드 치료를 받았고, 부종 및 말초 호산구 증가 소견은 현저히 호전되었으며, 7년의 경과 관찰에도 재발은 없었다.

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