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CASE BASED REVIEW

# A case of eosinophilic fasciitis without skin manifestations: a case report in a patient with lupus and literature review

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**Abstract**
Eosinophilic fasciitis (EF) is a rare connective tissue disease that causes inflammation and fibrosis of the fascia, inducing pain and motor dysfunction. Characteristic skin manifestations, such as edema, erythema, induration, peau d'orange appearance, and the groove sign, are of diagnostic significance and observed in the majority of patients with EF. We herein report a case of EF without these characteristic skin manifestations. A 66-year-old Japanese woman developed progressive limb pain and motor dysfunction. No skin changes were observed. We diagnosed the patient with EF based on the clinical course, magnetic resonance imaging, and en bloc biopsy containing fascia and muscle. Oral prednisolone therapy markedly attenuated limb pain and motor dysfunctions. Through a systemic search of the medical literature, we retrieved 4 juvenile cases and 8 adult cases of EF without characteristic skin manifestations during the clinical course. We herein present a systemic review on EF without skin manifestations and discuss differences between the two proposed sets of diagnostic criteria of EF.

**Keywords** Eosinophilic fasciitis · Diagnostic criteria · Skin manifestations · Systemic lupus erythematosus

## Introduction

Eosinophilic fasciitis (EF) is a rare connective tissue disease that was initially described by Shulman in 1974 [1]. There have since been approximately 400 cases of EF reported worldwide [2, 3]. EF is regarded as a fibrosing and sclerosing disorder that is associated with skin changes, such as edema, erythema, induration, peau d'orange appearance, and the groove sign, as the most characteristic manifestations of the disease [2–6]. According to previous findings, more than 95% of patients with EF show some of these skin manifestations [2, 3, 7–13]. Many patients also present with limb pain, joint contracture, muscle weakness, and a limited range of motion, which may be attributed to fascial inflammation and fibrosis [2, 6]. Characteristic laboratory findings for EF include peripheral eosinophilia, hypergammaglobulinemia, an elevated aldolase level, and increases in inflammatory markers, such as the erythrocyte sedimentation rate and C-reactive protein [2, 6]. Although the factors assumed to cause the disease include strenuous exercise, hemodialysis, borreliosis, and drugs, such as phenytoin, heparin, and nivolumab, the pathophysiological mechanisms underlying disease onset currently remain unclear [2]. We herein describe a Japanese case of EF without skin manifestations.

## Case report

A 66-year-old Japanese woman was admitted to our hospital with progressive limb pain, a limited range of motion, and

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muscle weakness. Approximately 4 months prior to her admission, she played volleyball. Three months before her presentation, she began to note restricted abduction in both shoulders. The intensity and range of pain gradually spread from the proximal part to the distal part, and similar symptoms appeared in the forearms, fingers, and lower limbs. She was a housewife and undergoing follow-up visits for proteinuria, hypertension, and thrombocytosis. Similar symptoms were not previously detected and there was no family history. Approximately 10 years prior to her presentation, she had undergone surgery for carpal tunnel syndrome. She was administered valsartan 80 mg/day for hypertension. She did not drink, smoke, or use recreational drugs.

A physical examination revealed no skin manifestations on the limbs, trunk, or face. She was 1.57 m in height and weighed 53.0 kg. Her body weight had decreased by 4–5 kg from 4 months prior to her presentation. Her body temperature was 37.2°C and other vital signs were normal. Pain was induced by the application of force to the deltoid muscles, biceps, carpal extensors, carpal flexors, finger flexors, and quadriceps. In the 0–5 manual muscle strength test, the biceps, carpal extensors, carpal flexors, and finger flexor muscle groups showed mild declines at 4 points on both sides. Grip strength was 4 kg on the right and 3 kg on the left. There were no abnormalities in deep tendon reflexes. Needle electromyography showed no abnormalities in the right biceps brachii group, right first dorsal interosseous muscle group, or right rectus femoris group. Nerve conduction tests showed delayed distal latency, a decreased compound muscle action potential, and decreased sensory nerve action potential in the right median nerve, which were consistent with her previous history of carpel tunnel syndrome. There were no abnormalities in the ulnar, tibial, or sural nerves.

A laboratory analysis on admission revealed the following: total leukocyte count 4,800/μL (neutrophils 86.8%, lymphocytes 8.7%, monocytes 3.7%, eosinophils 0.6%, basophils 0.2%); hemoglobin 9.4 g/dL; RBC count $350 \times 10^4$/μL; platelet count $61.6 \times 10^4$/μL; total protein 6.4 g/dL; creatine kinase 5 U/L; lactate dehydrogenase 241 U/L; aldolase 9.0 U/L (reference, ≤ 5.9 U/L); ferritin 287 ng/mL; fibrinogen degenerative products 14.3 mg/dL; fibrinogen 419 mg/dL; C3 113 mg/dL; C4 24.7 mg/dL; $CH_{50}$ 48.5 mg/dL; C-reactive protein 6.69 mg/dL; matrix metalloproteinase-3 233.6 ng/mL; sIL-2 receptor 1769 U/mL; antinuclear antibody ≥ 1:2560; anti-ds-DNA antibody 17.8 U/mL. The results of other laboratory tests are shown in Table 1. Urinalysis showed 1+ urine protein and no cellular casts. The 24-h urine collection test was performed twice, and urine protein levels were 0.629 g/day and 0.444 g/day, respectively. Abdominal and thoracic computed tomography images showed no visceral abnormalities, such as interstitial pneumonia and malignant tumors. T2 short tau inversion (STIR) magnetic resonance imaging (MRI) showed hyperintensity of the

**Table 1** Laboratory findings on admission

| | Result | Range |
|---|---|---|
| Hematology | | |
| Leukocytes | 4800/μL | 3300–8600 |
| Neutrophils | 86.8% | 34.0–75.0 |
| Lymphocytes | 8.7% | 17.0–55.0 |
| Monocytes | 3.7% | 1.0–11.0 |
| Eosinophils | 0.6% | 0–8.0 |
| Basophils | 0.2% | 0–3.0 |
| Erythrocytes | $3.50 \times 10^9$/μL | 3.86–4.92 |
| Hemoglobin | 9.4 g/dL | 11.6–14.8 |
| Hematocrit | 30.3% | 35.1–44.4 |
| MCV | 86.6 $\mu m^3$ | 83.6–98.2 |
| MCH | 26.9 pg | 27.5–33.2 |
| MCHC | 31.0 g/dL | 31.7–35.3 |
| Platelets | $61.6 \times 10^4$/μL | 15.8–34.8 |
| Biochemistry | | |
| Total protein | 6.4 g/dL | 6.6–8.1 |
| Albumin | 3.1 g/dL | 4.1–5.1 |
| AST | 13 U/L | 13–30 |
| ALT | 8 U/L | 7–23 |
| LDH | 241 U/L | 124–222 |
| Ferritin | 287 ng/mL | 4–87 |
| Total bilirubin | 0.4 mg/dL | 6.6–8.1 |
| Creatine kinase | 5 U/L | 41–153 |
| Aldolase | 9 U/L | 2.7–5.9 |
| BUN | 11 mg/dL | 8–20 |
| Creatinine | 0.78 mg/dL | 0.46–0.79 |
| Uric acid | 5.2 mg/dL | 2.6–5.5 |
| Sodium | 138 mmol/L | 138–145 |
| Potassium | 4.4 mmol/L | 3.6–4.8 |
| Chloride | 101 mmol/L | 101–108 |
| Glucose | 116 mg/dL | 73–109 |
| C-Reactive protein | 6.69 mg/dL | 0–0.14 |
| KL-6 | < 100 U/mL | < 500 |
| FT3 | 2.22 pg/mL | 1.88–3.18 |
| FT4 | 1.28 ng/dL | 0.70–1.48 |
| TSH | 1.510 μU/mL | 0.350–4.940 |
| Coagulation | | |
| PT-INR | 1.12 | 0.88–1.12 |
| APTT | 41.7 sec | 21.0–42.0 |
| Fibrinogen | 419 mg/dL | 140.0–340.0 |
| FDP | 14.3 mg/dL | < 5.0 |
| Serology | | |
| C3 | 113 mg/dL | 73–138 |
| C4 | 24.7 mg/dL | 11–31 |
| $CH_{50}$ | 48.5 mg/dL | 30.0–46.0 |
| MMP-3 | 233.6 ng/mL | 17.3–59.7 |
| Rheumatoid factor | < 10.0 U/mL | ≤ 15.0 |
| IgG | 1589 mg/dL | 861–1747 |
| IgA | 154 mg/dL | 93–393 |

**Table 1** (continued)

| | Result | Range |
|---|---|---|
| IgM | 59.6 mg/dL | 50–269 |
| sIL-2 receptor | 1,769 U/mL | 127–582 |
| HBs antigen | Negative | Negative |
| Anti-nuclear antibody | Titer of ≥ 1:2560 Homogeneous, speckled | ≤ 1:40 |
| Anti-ds-DNA antibody | 17.8 U/mL | ≤ 12.0 |
| Anti-ss-DNA antibody | 330.8 U/mL | ≤ 25.0 |
| Anti-RNP antibody | < 2.0 U/mL | < 10.0 |
| Anti-Smith antibody | < 1.0 U/mL | < 10.0 |
| Anti-CL-beta-2 GPI | < 0.7 U/mL | ≤ 3.5 |
| Lupus anticoagulant | 1.1 | < 1.3 |
| Anti-SS-A antibody | Negative | Negative |
| Anti-SS-B antibody | Negative | Negative |
| Anti-Scl-70 antibody | Negative | Negative |
| Anti-centromere antibody | < 2.0 U/mL | < 10.0 |
| Anti-CCP antibody | 0.8 U/mL | < 4.5 |
| Anti-ARS antibody | Negative | Negative |
| Anti-Jo-1 antibody | < 1.0 U/mL | < 10.0 |
| Anti-MDA-5 antibody | Negative | Negative |
| Anti-Mi-2 antibody | Negative | Negative |
| Anti-TIF-1-gamma-antibody | Negative | Negative |
| Anti-MPO-ANCA | 0.1 U/L | < 3.5 |
| Anti-PR3-ANCA | 0.1 U/L | < 3.5 |

*Abbreviations*: *AST*, aspartate aminotransferase; *ALT*, alanine aminotransferase; *LDH*, lactate dehydrogenase; *BUN*, blood urea nitrogen; *KL6*, Krebs von den Lungen-6; *FT3*, free triiodothyronine; *FT4*, free thyroxine; *TSH*, thyroid-stimulating hormone; *PT-INR*, international normalized ratio for the prothrombin time; *APTT*, activated partial thromboplastin time; *FDP*, fibrinogen degenerative products; *C3*, complement component 3; *C4*, complement component 4; $CH_{50}$, 50% hemolytic complement; *MMP-3*, matrix metalloproteinase-3; *sIL-2 receptor*, soluble interleukin-2 receptor; *HBs antigen*, hepatitis B surface antigen; *Anti-RNP antibody*, anti-ribonucleoprotein antibody; *Anti-CL-beta-2 GPI*, anti-cardiolipin beta 2-glycoprotein I complex antibody; *Anti-SS-A antibody*, anti-Sjögren's syndrome-related antigen A; *Anti-SS-B antibody*, anti-Sjögren's syndrome-related antigen B; *Anti-Scl-70 antibody*, anti-scleroderma-70 antibody; *Anti-CCP antibody*, anti-cyclic citrullinated peptide antibody; *Anti-ARS antibody*, anti-aminoacyl tRNA synthetase antibody; *Anti-Jo-1 antibody*, anti-histidyl tRNA synthetase antibody; *Anti-MDA5 antibody*, anti-melanoma differentiation-associated gene 5 antibody; *Anti-TIF-1-gamma-antibody*, anti-transcriptional intermediary factor 1-gamma antibody; *Anti-MPO ANCA*, anti-myeloperoxidase anti-neutrophil cytoplasmic antibody; *Anti-PR3 ANCA*, anti-proteinase 3 ANCA

fascial areas at the upper right arm, left hip, left thigh, and inner left chest arm to the chest wall (Fig. 1). MRI showed no abnormalities in the central nervous system. En bloc biopsy containing fascia and muscle from the left semimembranosus revealed normal epidermis, dermis, and superficial layers of subcutaneous tissue, while several foci of mononuclear cell infiltration were observed in the deep layer of subcutaneous tissue and deep fascia (Fig. 2). Alkaline phosphatase staining revealed that enzymatic activity was markedly elevated in the fascia. Mild to moderate variation was observed in muscle fiber size. Relatively atrophic muscle fibers were clustered at the perifascicular area facing the fascia. Immunohistochemistry showed scattered muscle fibers, particularly perifascicular fibers, expressing MHC-I, while some perifascicular fibers also expressed MHC-II. Myxovirus-resistant protein A (MxA) was not expressed in muscle fibers. Complement C5b-9 was not deposited on the capillaries. Renal biopsy showed benign nephrosclerosis.

We diagnosed the patient with EF based on her symptoms, laboratory data, MRI, and biopsy results, all of which were consistent with EF, except for the lack of skin manifestations. The oral administration of prednisolone at 25 mg/day markedly attenuated pain, muscle weakness, and inflammatory responses (Fig. 3). She was discharged 22 days after the initiation of treatment when sufficient prednisolone tapering (< 20 mg/day) and recovery of symptoms were achieved. There have been no recurrence and newly developed cutaneous manifestations for 12 months with the tapering of prednisolone.

## Discussion

The present case did not have any of the characteristic skin manifestations of EF, such as edema, erythema, sclerosis,

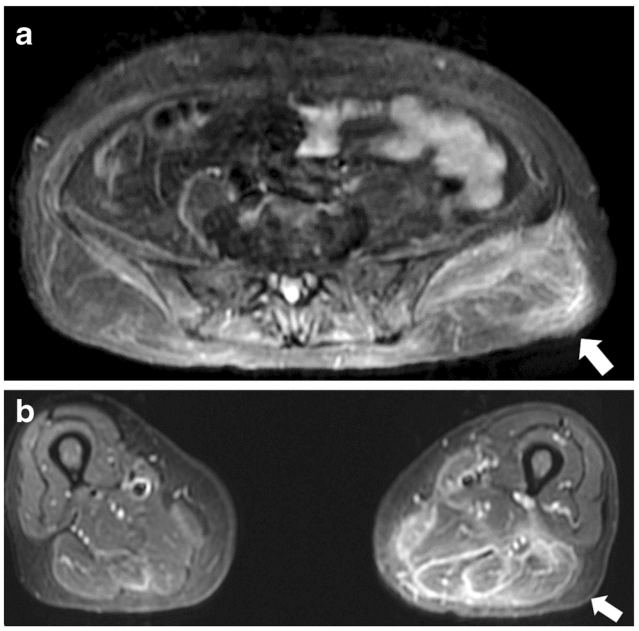


**Fig. 1** MRI T2 STIR images. **a** Hyperintensity signals in the left gluteus maximus, including the fasciae (arrow). **b** The fasciae of the left thigh muscles also show hyperintensity signals (arrow)

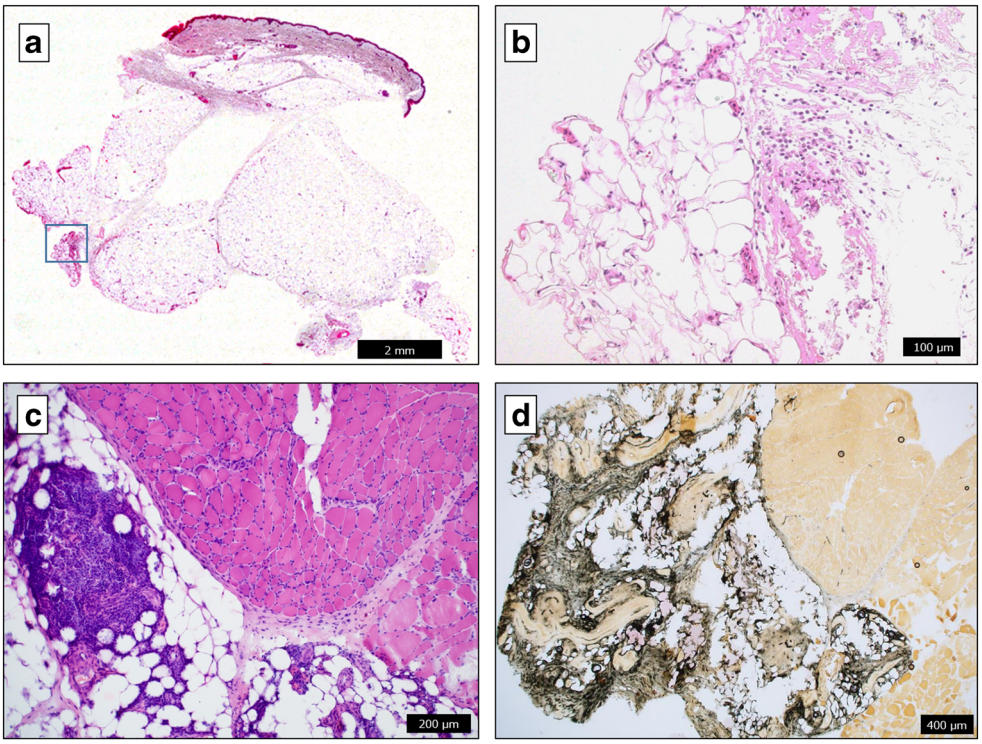


**Fig. 2** Pathological findings of en bloc biopsy specimens of the left thigh containing fascia and muscle. **a** Normal appearance of the epidermis, dermis, and subcutaneous tissue. A high-magnification view of the boxed region is shown in **b**. **b** Moderate mononuclear cell infiltration in the deep layer of subcutaneous tissue. **c** Severe mononuclear cell infiltration into the fascia. Clustered relatively atrophic muscle fibers at the perifascicular area facing the fascia. Mild to moderate variations in muscle fiber sizes. **d** Markedly increased alkaline phosphatase activity in the fascia. Hematoxylin and eosin staining (**a**–**c**) and immunohistochemistry of alkaline phosphatase **d**

peau d'orange appearance, and the groove sign, during the clinical course. To the best of our knowledge, there have been only 12 (8 adults and 4 children) cases of EF without any skin manifestations during the disease course among the approximately 400 cases of EF reported to date [2, 3, 7–13]. In 4 cases, skin manifestations were initially absent, but developed with disease progression [14–17]. The skin manifestations of EF have been assumed to begin with edema and then progress to more severe changes, such as skin sclerosis, peau d'orange appearance, and the groove sign [2, 6]. Due to the gradual progression of the disease, some cases of EF may lack apparent skin manifestations in the early stage of the disease. Previous studies on en bloc biopsy samples have shown normal epidermis and dermis in contrast to inflammation of the fascia frequently observed in early EF cases. Epidermal structural loss, dermal thickening, and hyalinization frequently appear according to the clinical severity of EF [2, 18]. The normal epidermal and dermal appearance in the biopsy pathology of the present case suggest an early disease stage.

Based on this assumption, early treatment intervention may reduce the appearance of skin changes. Since the duration of symptoms prior to diagnosis is assumed to be associated with a poor treatment response in skin manifestations and motor dysfunctions, the importance of an early diagnosis and treatment have been emphasized [6, 13]. Recent studies in the

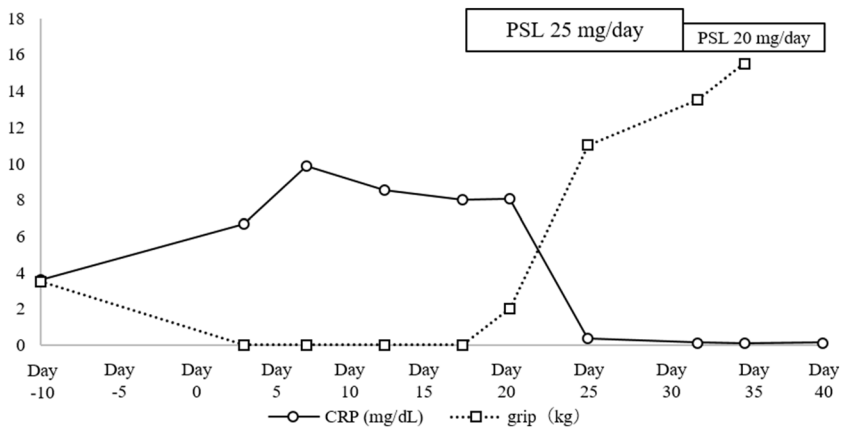


**Fig. 3** Clinical course of the patient. The significant amelioration of inflammation and muscle weakness was observed after the treatment with oral prednisolone. The patient was hospitalized on day 1, diagnosed with EF, and started the PSL treatment on day 18. CRP, C-reactive protein; PSL, prednisolone

USA have shown that the average duration from the onset of symptoms to EF diagnosis is 6 or 11 months [3, 5]. In contrast, diagnosis and treatment initiation in the present case were as early as approximately 4 months from the disease onset. This early diagnosis and treatment initiation may contribute to the lack of obvious cutaneous manifestations in our case.

Oral prednisolone is the gold standard treatment for EF; however, randomized control trials have not yet been conducted [2, 6, 13]. The present case also showed the marked amelioration of pain, muscle weakness, and the limited range of motion after the initiation of oral prednisolone. The relatively earlier diagnosis and initiation of immunotherapy may have contributed to the good outcome of the present case.

Based on progressive limb pain, the limited range of motion, and muscle weakness in our patient, we initially suspected myogenic diseases, neurogenic diseases, rheumatoid arthritis, polymyalgia rheumatica, and/or ANCA-related diseases. However, her symptoms and laboratory data did not completely satisfy any of the diagnostic criteria of the above diseases. Muscle pathology showed that perifascicular fibers were atrophic, mimicking perifascicular atrophy, a diagnostic finding for dermatomyositis. However, dermatomyositis was excluded because MxA, a diagnostic marker of dermatomyositis, was negative [19]. Another possibility was perifascicular necrosis, a typical finding of anti-aminoacyl tRNA synthetase syndrome (ASS), because regenerating fibers may be small. The expression of MHC-II in perifascicular fibers is frequently reported in ASS [20]. However, none of the anti-synthetase antibodies, including those to isoleucyl-tRNA synthetase (OJ), phenylalanyl transfer RNA synthetase (Zo), and tyrosyl-tRNA synthase (Ha), was detected, even by the RNA immunoprecipitation method [21, 22].

It was challenging to establish whether our patient concurrently had systemic lupus erythematosus (SLE). We suspected SLE based on positivity for the anti-nuclear antibody and anti-ds-DNA antibody and a decreased lymphocyte count. However, the patient did not have malar rash, photosensitivity, discoid rash, oral ulcers, serositis, alopecia, or neurological symptoms. Although limb pain occurred with movement, joint tenderness, swelling, effusion, synovitis, and morning stiffness were not observed in any of the patient's joints. Proteinuria greater than 0.5 g/24 h was only observed once during the clinical course and spontaneously resolved before the initiation of treatment. Renal biopsy showed mild sclerosis, but not apparent lupus nephritis. None of the laboratory results satisfied the diagnostic criteria other than positivity for the anti-nuclear antibody and anti-ds-DNA antibody and a decreased lymphocyte count (Table 1). Based on these results, we concluded that this case was not classified as SLE based on the classification criteria for SLE by the American College of Rheumatology and Systemic Lupus Collaborating Clinics; however, it was still suspected. New classification criteria for SLE were recently published by the European League Against Rheumatism and the American College of Rheumatology [23], according to which the present case may be diagnosed as SLE since the patient satisfied the following criteria: positivity for anti-nuclear antibody at a titer of $\geq$ 1:80, proteinuria greater than 0.5 g/24 h, and positivity for anti-dsDNA antibody.

To date, 6 patients with EF related to SLE have been reported [24–29], all of whom had at least one of the characteristics of skin manifestations of EF. Furthermore, none of the formerly reported EF patients without skin manifestations concurrently had SLE.

There have been no universally accepted and validated diagnostic criteria for EF [2]. In many cases, the diagnosis of EF was reached based on characteristic skin findings, hyperintense fascia on MRI, and fascial thickening with inflammatory cell infiltration in en bloc biopsy containing fascia and muscle. We diagnosed our patient with EF based on her symptoms, laboratory data (particularly the elevated serum aldolase level in contrast to a normal serum creatinine kinase level, which is one of the characteristic laboratory findings of EF), MRI findings, and biopsy results, which were all consistent with those of EF, except for the lack of skin manifestations. Essential thrombocythemia and carpal tunnel syndrome in this patient further supported the diagnosis of EF because this hematological disorder and carpal tunnel syndrome have both been identified as common complications of EF [2].

Diagnostic criteria for EF have recently been proposed by two groups (Table 2) [30, 31]. One of the critical differences between the two diagnostic criteria is skin manifestations; the diagnostic criteria proposed by Ihn require skin manifestations to diagnose EF, while those by Pinal-Fernandez et al. do not. Based on the diagnostic criteria proposed by Pinal-Fernandez et al., the present case may be diagnosed as EF because the symptoms observed satisfied one major (criteria 2) and two minor (criteria 3 and 5) criteria. In contrast, the present case cannot be diagnosed as EF based on the diagnostic criteria proposed by Ihn because the main criterion was not satisfied. Table 3 shows the findings of 10 previously reported cases of EF without skin manifestations applied to the diagnostic criteria of each group. Five out of the 6 available cases were diagnosed as EF based on the diagnostic criteria proposed by Pinal-Fernandez et al., in contrast to none based on those by Ihn. These findings suggest that the diagnostic criteria proposed by Pinal-Fernandez et al. are more appropriate for the early diagnosis of EF without apparent skin manifestations. It is an interesting question how many EF cases without characteristic skin manifestations are missed or misdiagnosed as other diseases, although it is hard to estimate the precise frequency so far. Indeed, there is a case of EF initially diagnosed as postviral myalgia and later corrected to EF, since the patient

**Table 2** Two proposed diagnostic criteria for eosinophilic fasciitis

**A** Diagnostic criteria for eosinophilic fasciitis proposed by Pinal-Fernandez et al [30]

*Major Criteria*

1.Swelling, induration, and thickening of the skin and subcutaneous tissue that is symmetrical or non-symmetrical, diffuse (extremitis, trunk and abdomen) or localized (extremities)

2.Fascial thickening with accumulation of lymphocytes and macrophages with or without eosinophilic infiltration (determined by full-thickness wedge biopsy of clinically affected skin)

*Minor Criteria*

1.Eosinophilia >0.5 x $10^9$/L

2.Hypergammaglobulinemia >1.5 g/L

3.Muscle weakness and/or elevated aldolase levels

4.Groove sign and/or *peau d'orange*

5.Hyperintense fascia on MR T2-weighted images

Exclusion criteria: diagnosis of systemic sclerosis

Presence of both major criteria, or one major criterion plus 2 minor criteria, establishes the diagnosis of eosinophilic fasciitis

**B** Diagnostic criteria for eosinophilic fasciitis proposed by Ihn [31]

*Major Criterion*

Symmetrical plate-like sclerotic lesions are present on the four limbs. However, this condition lacks Raynaud's phenomenon, and systemic sclerosis can be excluded.

*Minor Criteria 1*

The histology of a skin biopsy that incorporates the fascia shows fibrosis of the subcutaneous connective tissue, with thickening of the fascia and cellular infiltration of eosinophils and monocytes.

*Minor Criteria 2*

Thickening of the fascia is seen using imaging tests such as magneticresonance imaging (MRI).

A definitive diagnosis is made when a patient has the major criterion and one of the minor criteria, or the major criterion and two of the minor criteria

initially showed only malaise and heaviness in extremities but later developed characteristic cutaneous manifestations of EF [14]. There might be some other cases of EF without characteristic cutaneous manifestations misdiagnosed as other diseases such as myalgia. The initiation of immunotherapy before the appearance of skin changes may have resulted in the good outcome in the present case, which also indicates the importance of an early diagnosis based on appropriate diagnostic criteria and en bloc biopsy, particularly in cases without skin manifestations.

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**Data availability** The datasets obtained and/or analyzed during the current study available from the corresponding author on reasonable request.

## Compliance with ethical standards

**Disclosures** None.

**Ethics approval** All procedures used in this research were approved by the Ethical Committee of University of Fukui.

**Consent for participate and publication** Informed consent for participation and publication of the case was obtained from the patient.

**Table 3** Evaluation of EF patients without skin manifestations based on diagnostic criteria proposed by Pinal-Fernandez et al. and Ihn

| No. | Age | Sex | EF diagnosis based on Pinal-Fernandez et al. criteria | | EF diagnosis based on Ihn criteria | | Ref. |
|---|---|---|---|---|---|---|---|
| 1 | 3 | F | Yes | Mj 2 + mn 1 + mn 5 | No | mn 1 + mn 2 | 12 |
| 2 | 4 | F | Yes | Mj 2 + mn 2 + mn 5 | No | mn 1 + mn 2 | 12 |
| 3 | 11 | F | Yes | Mj 2 + mn 1 + mn 2 + mn 5 | No | mn 1 + mn 2 | 11 |
| 4 | 14 | M | Yes | Mj 2 + mn 1 + mn 2 + mn 5 | No | mn 1 + mn 2 | 10 |
| 5 | 27 | F | Yes | Mj 2 + mn 3* + mn 5 | No | mn 1 + mn 2 | 8 |
| 6 | 42 | M | Yes | Mj 2 + mn 3* + mn 5 | No | mn 1 + mn 2 | 8 |
| 7 | 46 | M | No | Mj 2 + mn 5 | No | mn 1 + mn 2 | 9 |
| 8 | 69 | F | Yes | Mj 2 + mn 1 + mn 3 + mn 5** | No | mn 1 + mn 2 | 7 |
| 9–12 | N/A | N/A | Unevaluable | Unevaluable | Unevaluable | Unevaluable | 13 |
| Our case | 66 | F | Yes | Mj 2 + mn 3 + mn 5 | No | mn 1 + mn 2 | |

*Abbreviations*: *F*, female; *M*, male; *Mj*, major criterion; *mn*, minor criterion; *N/A*, not available; *Ref.*, reference

*Joints with a limited range of motion are considered to have muscle weakness

**Hyperintense fascia is evaluated by $^{18}$ fluoro-2-deoxyglucose positron emission tomography instead of T2-weighted magnetic resonance imaging

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