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Neuromuscular Ultrasound Findings in Eosinophilic Fasciitis: A Case Series and Literature Review

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Introduction

Eosinophilic fasciitis (EF) is a rare condition characterized by inflammation, thickening, and sclerosis of the skeletal muscle fascia.¹ It can present with a variety of symptoms including induration and peau d'orange changes of the skin, stiffness in the distal limbs, arthralgias, myalgias, and joint contractures. Laboratory abnormalities may include elevated erythrocyte sedimentation rate (ESR), hypergammaglobulinemia, and peripheral eosinophilia.²⁻⁴

Electromyography (EMG) is either normal or demonstrates small voluntary motor unit potentials with early recruitment as can be seen in myopathic disorders. The diagnosis of EF typically relies on full thickness incisional biopsy, but interest in the application of imaging modalities including ultrasound (US) and magnetic resonance imaging (MRI) has increased in the past decade. We present three patients in whom imaging assisted in the diagnosis of EF. A systematic review of the literature identified additional cases.

Patient 1

A 16-year-old boy presented with 3 months of wrist stiffness and mild aching in the forearms. He had peau d'orange changes over the forearms and hands. His upper arms and distal legs were also mildly indurated. Mobility was limited at the wrists, fingers, and ankles. Muscle bulk and strength testing in all limbs were normal. Sensation was slightly decreased to all modalities in the toes. Deep tendon reflexes were normal.

Laboratory studies revealed an eosinophil relative count of 13% (normal 0-6%). Creatine kinase (CK), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and serology for antinuclear antibody (ANA) and anti-Scl-70 were normal. Nerve conduction studies

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(NCS) in the right forearm were normal and EMG showed decreased insertional activity and short duration, low amplitude voluntary motor unit potentials with no abnormal spontaneous activity and normal recruitment. US findings are in the Table and Figure. Full thickness biopsy of the proximal forearm showed chronic inflammation around vessels, within the fascia, and focally in skeletal muscle, with plasma cells, lymphocytes, and rare eosinophils. He was diagnosed with EF and treated with 60 mg of prednisone daily. Follow-up US findings at 6 weeks are in the Table.

Patient 2

A 57-year-old woman presented with several years of cramping and myalgias, as well as five months of increasing stiffness of her limbs. Her skin showed generalized thickening and induration. She had peau d'orange changes of the thighs. Range of motion was reduced at the wrists and ankles. She had normal strength, sensation, and reflexes.

Laboratory studies revealed an eosinophil count of 25.2%. CK, CRP, ANA, anti-SSA/SSB (Sjögren's syndrome A/B), and anti-Scl-70 were normal. X-anti-neutrophil cytoplasmic antibody titer was 1:80, and ESR was 34 mm/hour. NCS and EMG of the left upper and lower limbs were normal. MRI and US results are in the Table and Figure. A full thickness biopsy over the vastus lateralis showed superficial fibrosis of the skin and fascia, perivascular cuffs of inflammatory cells, and subcutaneous inflammation including occasional eosinophils without myositis.

She was diagnosed with EF and began treatment with prednisone 60 mg daily and methotrexate 20 mg weekly. She had slow, but sustained improvement. Repeat US findings at 7 months are in the Table. Her eosinophil count decreased to 3.1%.

Patient 3

A 21-year-old man presented with restricted movement of the wrists when trying to do push-ups, and with fatigue and weight loss. He subsequently developed pain in the forearms and thickening of the skin of the forearms and hands, as well as skin dimpling around the waist and back. Examination showed thickening of the skin over the lower back, hands, and forearms. He had peau d'orange changes of the skin around the waist and low back. Wrist extension was reduced bilaterally. He had normal strength, sensation, and reflexes.

Laboratory studies revealed an eosinophil count of 22%. CK, ESR, serum immunofixation, urine protein electrophoresis, and serology for anti-ribonucleoprotein, anti-SSA/SSB, anti-Scl 70, anti-Smith, anti-double stranded DNA, and rheumatoid factor were normal. ANA was 1:160. Electrodiagnostic studies of the right upper limb were normal. MRI and US findings are in the Table. Subsequent right forearm biopsy revealed thickened fascia and fibroseptae with a predominantly lymphocytic inflammatory infiltrate and rare eosinophils.

A diagnosis of EF was made and prednisone 60 mg daily was started. He subsequently regained mobility of the wrists, and his skin changes improved. Symptoms returned with tapering of prednisone to 40 mg but improved with the addition of methotrexate.

Discussion

Interest in imaging to aid in the diagnosis of EF, guide biopsy site, and monitor therapeutic response has recently increased. MRI is excellent in the evaluation of soft tissues, but it is limited by cost, duration, gadolinium exposure, and ease of access. US is economically feasible, available at the bedside, and allows for immediate visualization of multiple affected sites in real time.

The US findings in these new cases are almost all subjective (except for the median nerve cross-sectional area measurements), but they are striking and match with other cases in the literature, as noted in the Table. US demonstrated thickened subcutaneous tissue, thickened fascia, and increased echogenicity in the fascia and muscle. Additionally, hypoechoic halos were visualized surrounding the flexor tendons at the wrist in patients 1 and 3, which is likely secondary to inflammation. When using US to assess for EF, we recommend imaging at the site of thickened skin. Note should be made of the skin and fascial thickness, which can be assessed subjectively or measured using the US calipers. Though there are not normal values, US of a region that does not appear to be clinically affected can provide a comparison. Similarly, subjective note should be made of the fascia and muscle echogenicity, and without changing the gain, comparison can be made to a clinically unaffected region. This method of US assessment can be used to guide the biopsy site, and it can also be repeated following treatment as a subjective biomarker. Future investigations of US in EF should focus on quantitative assessments of subcutaneous tissue and fascial thickness, objective measures of fascial and muscle echogenicity, and other approaches such as Doppler, which may allow for assessment inflammation and/or neovascularization.

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Abbreviations

ANA	Antinuclear antibodies
CK	Creatine kinase
CRP	C-reactive protein
EF	Eosinophilic fasciitis
EMG	Electromyography
ESR	Erythrocyte sedimentation rate
MRI	Magnetic resonance imaging
NCS	Nerve conduction study
US	Ultrasound

Figure. Ultrasound and MRI Imaging in Patients 1 and 2

A: Ultrasound shows stacked flexor digitorum superficialis and flexor digitorum profundus tendons (T) adjacent to the slightly enlarged median nerve (arrow, 12 mm^2 , normal is $< 12 \text{ mm}^2$). A hypoechoic cuff is seen surrounding the tendon pair and is also evident around the other flexor tendons at the wrist. These areas of decreased echogenicity likely represent edema surrounding the flexor tendons, and provide evidence of inflammation within the carpal tunnel. B: T1-weighted MRI demonstrates diffuse subcutaneous as well as superficial and deep perifascial edema in the left thigh without evidence of myositis. C: Ultrasound of the left vastus lateralis pre-treatment shows increased echogenicity of muscle and fascia. D: Ultrasound of the left vastus lateralis seven months post-treatment shows reduced echogenicity in the same muscle and fascia as image C.

Table

Imaging findings for nine patients with eosinophilic fasciitis.

Reference	Interval from Symptom Onset	Site of US/MRI	Initial US	Initial MRI	Interval to Repeat Imaging	Follow-up US	Follow-up MRI
5	3 mo	wrists/wrists	thickened skin/SC tissue, and FR, hypoechoic cuff around flexor tendons	enhancement of flexor tendons, thickened FR, cuff of high signal around ECU tendon, extensor tendon changes less prominent	6 mo	not performed	↓ fascial thickening in distal forearm/ wrists, changes in tendons and FR resolved
6	unknown	calves/calves	halo and septal effusion	edema of calf muscles	1 wk	normal	some fluid in muscle septum
7	5 mo	forearm/forearm	thickening of fascia	hyperintensity of fascia	unknown	not performed	normal
10	11 months	leg/leg	thickened fascia	thickening of fascia, contrast enhancement of myofascial plane	N/A	not performed	not performed
8	8 mo	forearm, leg/forearm	thickened fascia R>L, ↑ EG of extensor muscles, normal EG biceps brachial, forearm flexors, leg muscles	slight thickening of fascia of flexors and extensors with contrast enhancement	2 yr	improvement compared to initial US but fasciae slightly thick compared to healthy control	not performed
9	6 mo	forearms/not performed	thickening and ↓ density of SF fascia	Not performed	35 sessions (~9 wks)/50 sessions (~14 wks)	F/U 1: near normal skin, ↓ in thickness and increase in density of fascia F/U 2: normal fascial thickness	not performed
Patient 1	3 mo	forearm/not performed	thickened skin/SC tissue/ fascia, ↑ muscle signal/AN, hypoechoic halo around flexor tendons, MN enlargement	Not performed	6 wk	↓ thickness of the skin, SC tissue, and fascia, ↓ CSA of flexor tendons, hypoechoic halo, and MN	not performed
Patient 2	several years	arm, leg/leg	thickened skin/SC tissue, ↑ EG and thickness of fascia and muscle most prominent at VL	edema of SC tissue, SF and deep fascia	7 mo	↓ thickness of the skin, SC tissue, and fascia	not performed
Patient 3	unknown	forearm/forearm	thickened skin/SC tissue, and fascia, edema around flexor tendons, ↑ finger flexor muscle signal	abnormal signal of SC tissue	N/A	not performed	not performed

Abbreviations: US (ultrasound), MRI (magnetic resonance imaging), SC (subcutaneous), FR (flexor retinaculum), R (right), L (left), EG (echogenicity), AN (anisotropy), MN (median nerve), VL (vastus lateralis), ECU (extensor carpi ulnaris), SF (superficial), US (ultrasound), F/U (follow up), CSA (cross-sectional area).