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Case Report

Sonographic Features of Eosinophilic Fasciitis with Asymmetrical Symptoms: A Case Report

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Eosinophilic fasciitis is a rare rheumatological disorder, with the clinical presentations of myalgia, symmetrical skin induration, and peripheral eosinophilia. Because of overlapping symptoms with other rheumatological disorders, early diagnosis of eosinophilic fasciitis is difficult. The confirmatory diagnosis relies on full-thickness skin biopsy. Imaging studies are also used for evaluation and diagnosis. Among them, high-resolution sonography has the advantage of high accessibility over other imaging modalities in detecting musculoskeletal disorder, but the sonographic features of eosinophilic fasciitis are rarely reported. This study reported a 24-year-old woman who was diagnosed as eosinophilic fasciitis with rare presentation of asymmetric involvement. She had left wrist/hand contracture and a tightness sensation at the right calf and trunk. Diagnosis was confirmed by a full-thickness skin biopsy 1.5 years after initial symptoms. The ultrasound showed marked thickening of the tendon sheaths of the flexor tendons with reduced echogenecity at the level of metacarpal bones in a transverse view. Thickening of the superficial fascia was also present at the right gastrocnemius. After treatment with prednisolone for 8 months, the wrist and hand contracture improved and the sonographic findings normalized. (Tw J Phys Med Rehabil 2012; 40(2): 91 - 95)

Key Words: eosinophilic fasciitis, sonography, rheumatological disorder

INTRODUCTION

Eosinophilic fasciitis, first described by Shulman in 1974, is a rare scleroderma-like disorder presented with diffuse fasciitis and eosinophilia. Clinical features include swelling and rapidly progressive thickening of the involved skin with woody induration and peau d'orange change. Laboratory data reveals peripheral eosinophilia, an elevated erythrocyte sediment rate (ESR), and hypergammaglobulinemia.[1,2] If untreated, it would lead to collagenization and thickening of the inflammed structures, cutaneous fibrosis, and joint contractures, and therefore, impair the patient's functional performance and daily activity. Different from systematic scleroderma, eosinophilic fasciitis primarily involves the fascia, panniculus, and muscle.[3] The most commonly involved body parts include arms and legs with hands/feet, arms and legs only, and trunk.[2] Unilateral symptoms of limbs are rare.[4] Although the etiology of eosinophilic fasciitis

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is still unknown, reports have suggested possible causes, such as strenuous exercise, trauma, and infection with Borrelia Burgdorferi.[4] Additionally, rare report related statin use to eosinophilic fasciitis or intake of L-tryptophan to eosinophilia-myalgia syndrome.[3]

Confirmatory diagnosis of eosinophilic fasciitis is established by a full-thickness (epidermis to muscle) biopsy, which shows thickening of the fascia and infiltration of inflammatory cells, including lymphocytes, plasma cells, and eosinophils. In advanced stage, there is fascia fibrosis, hyalinization of collagen, and sclerosis of the reticular dermis.[5] Image studies such as magnetic resonance image (MRI) and sonography have been used to guide the diagnosis.[6-8] Comparatively, sonography has the advantage of low-cost and easy accessibility in evaluating soft tissue pathology; however, the sonographic feature of eosinophilic fasciitis was rarely reported.[8] This report presented a case of eosinophilic fasciitis with a rare presentation of asymmetrical symptoms. Sonographic findings alone with their correlation with clinical improvement were reported.

CASE REPORT

A 24-year-old woman presented to our clinic for progressive flexion contracture of the left hand and wrist for more than 1 year. Initially, she noticed some erythematous nodules and hyperpigmentation over her right upper arm approximately 18 months before the visit. It was diagnosed as local morphea, and a local corticosteroid injection was given at a local hospital. Nevertheless, skin tightness and stiffness at the back, left arm and right calf developed insidiously later. This symptom was most significant at the left distal forearm to the hand, where painful indurations at the flexor side prevented her from making a fist or fully grasping because of limited joint movement. Laboratory data from a second visit to the dermatologist showed peripheral eosinophilia (20 %). She was treated for scleroderma and received 10 mg of prednisolone daily for several weeks. Skin tightness slightly improved, but limited range of motion at the left hand and wrist persisted. Therefore, she discontinued the medication for 2 months.

The woman had been working as a vet in a zoo for 1 year and denied any history of major systematic diseases, such as diabetes, hypertension, thyroid disease, renal disease, or hepatic disease. She did not engage in regular exercise and denied participating in strenuous exercise before the onset of symptoms. She was not under any medication or intake products containing L-tryptophan or statins.

A physical examination showed a retractile flexed posture of the left hand, particularly the index and middle finger, accompanied by subcutaneous thickening and painful indurations at the left palm and the distal forearm with palpation. Subcutaneous thickening at both calves, thighs, and back were also manifested. She had no Raynaud's phenomenon, joint swelling, or hand numbness. Her muscle tone, deep tendon reflexes, and muscle strength of the four limbs were normal.

Laboratory data revealed normal white blood cell count (7070/mm^3), marked peripheral eosinophilia (29%), a slightly elevated ESR (first hour: 15 mm/h), but normal C-reactive protein (CRP). She had a negative antinuclear antibody (ANA) and anti-ENA antibodies, and normal C3 and C4 complement levels. Serum protein electrophoresis also showed polyclonal gammopathy. A biopsy of the left distal forearm revealed thickened and homogenized collagen bundles in the reticular dermis and fascial fibrosis. However, there was no apparent eosinophilic or lymphocytic infiltrate in the dermo-subcutis junction and no thickened collagen bundles in the papillary dermis. The diagnosis of eosinophilic fasciitis was made.

Sonographic examination of the left palm, wrist, and both calves was performed by the Siemens ACUSON S2000 high-resolution sonography system with a 7-14 MHz linear transducer. The overlying subcutaneous tissue was mildly thickened with intact flexor tendons at the level of pisiform and scaphoid bones, and a mild hypoechoic tissue surrounding the tendons (Figure 1A). There was no visible flow on the power Doppler (Figure 1B). Moreover, there was marked thickening of the tendon sheaths of the flexor tendons with reduced echogenicity of tendons at the level of metacarpal bones in the transverse view (Figure 2). Thickening of the superficial fascia of the right medial gastrocnemius was also recorded (Figure 3).

Prednisolone 30 mg per day and hydroxychloroquine sulfate 400 mg per day were prescribed. Within 2 months, the peripheral eosinophilia lowered to normal range (0.2 %). Splinting for the left hand/wrist was given and a home program for gentle stretching exercise was

instructed. Eight months later, the tightness sensation of the calves subsided and subcutaneous thickening and indurations at the left palm and distal forearm improved. The woman was able to extend her left fingers fully with the wrist in neutral position, but not in the extended position. The dosage of prednisolone was lowered to 5 mg per day 8 months later. Meanwhile, sonography showed a normal tendon sheath of flexor tendons at the left wrist and the superficial fascia at the right medial gastrocnemius muscle (Figures 4A, 4B), compatible with clinical improvement.

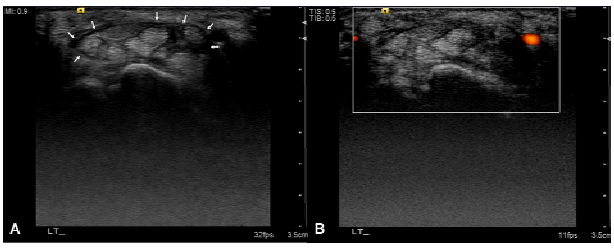

Figure 1 (A) Transverse sonogram of the left carpal tunnel showing mild thickening of the subcutaneous tissue and a mild hypoechoic tissue surrounding the tendons at the level of pisiform and scaphoid bones. (B) Power Doppler sonogram showing no increased vascularity of the peritendinous tissue.

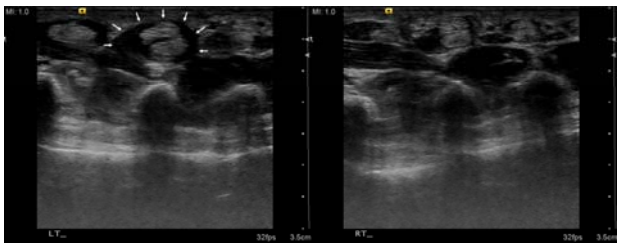

Figure 2. Transverse sonogram of the left palm showing thickening and reduced echogenecity of the peritendinous area surrounding the flexor tendons at the level of metacarpal bones. The right side is relatively normal.

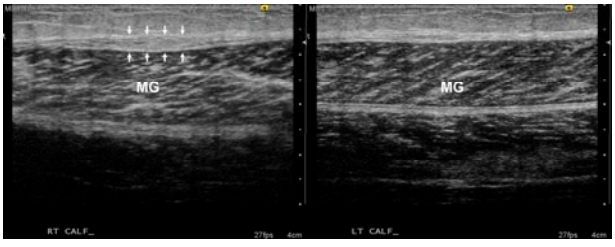

Figure 3. Sonogram at middle calf showing significant thickening of the superficial fascia at right side in comparison with left side (thickness 3.4 mm, *vs.* 1.9 mm). MG: medial gastrocnemius.

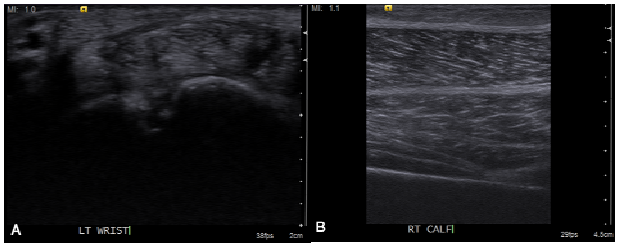

Figure 4. Ultrasound performed after 8 months showing: (A) Normal tendon sheath of flexor tendons at the left wrist. (B) Normal superficial fascia (thickness 1.4 mm) at the right medial gastrocnemius.

DISCUSSION

This work reported the sonographic features of a case of eosinophilic fasciitis and portrayed her clinical improvement and sonographic change after treatment. The results are important for future evaluation and treatment of similar cases.

Classical features of eosinophilic fasciitis include initial swelling and rapidly progressive thickening of the involved skin with woody induration and peau d'orange change.[2,3] The most commonly involved body parts are arms, hands, legs, and feet. Clinical presentations of eosinophilic fasciitis are usually symmetrical.[4] Initially, the reported case was treated as a localized scleroderma. Early diagnosis has been difficult because of overlapping symptoms of scleroderma and eosinophilic fasciitis. The mean duration from the first symptoms to diagnosis might be as long as 8.8 and 13 months in two case series.[2,3] However, the differential diagnosis is important because of their different disease courses. Scleroderma is typically presented with Raynaud's phenomenon and involves epidermis and dermis on the skin biopsy.[2] In contrast, eosinophilic fasciitis primarily involves the fascia, panniculus, and muscle.[3] For this case, a biopsy was performed before visiting our hospital, but the diagnosis was not conclusive, probably due to no full-thickness biopsy obtained. Lack of a definite diagnosis and incomplete treatment may be responsible for the progressive flexor contracture of her left hand in the early course.

Both MRI, and less frequently, sonography have been used to assist the diagnosis of eosinophilic fasciitis.[6-8] The typical findings of MRI are high signal intensity within the fascia in T2-weighted sequences and enhancement after contrast administration in T1-weighted

sequences.[6-8,9] In a study of Chan et al, sonography disclosed thickening of the flexor retinaculum at wrists, thickening of the skin, and subcutaneous tissue. A cuff of hypoechoic tissue also evidenced around the flexor tendons in the carpal tunnels, suggesting soft tissue swelling of the tendon sheath.[8] Pillen et al also presented a case using sonography to detect thickened fascia of the forearm.[9] In the current case, sonography showed similar findings including hypoechoic thickening of the tendon sheath of the left palm and thickened fascia of the right calf. It indicated the presence of tenosynovitis and fasciitis. These findings differed from that of localized scleroderma, in which sonography showed hyperechoic thinning and increased vascularity of the subcutaneous tissue.[10]

Systemic corticosteroid therapy is the mainstay of treatment and the response is more effective for an early inflammatory stage than an advanced fibrotic stage.[3] The current case significantly improved in laboratory abnormalities and clinical symptoms after an 8-month treatment with an oral corticosteroid. Sonography also showed parallel improvement, including thinning of the tendon sheath and the superficial fascia at affected sites.

In conclusion, sonography is a useful tool in evaluating the change of soft tissue and fascia, and helps in assessment of therapeutic response in case with eosinophilic fasciitis.

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非對稱性嗜伊紅性筋膜炎的超音波表現：病例報告

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嗜伊紅性筋膜炎(eosinophilic fasciitis)是一種相當少見的風濕免疫疾病，臨床表現有肌肉疼痛、對稱性手腳局部皮膚硬化、以及周圍血液嗜伊紅性白血球數量上升。因爲此疾病與其他免疫風濕性疾病之症狀多所重複，因此，早期診斷不易，確診需倚賴全皮層皮膚切片檢查。影像學檢查也用於協助評估診斷，而超音波對於軟組織具高解像力、且取得方便，讓它在診斷嗜伊紅性筋膜炎的可用性不輸於核磁共振造影，然而，過去極少文獻提出嗜伊紅性筋膜炎的超音波影像表現。本文報告一位 24 歲女性，出現少見的非對稱性症狀，包括左手腕/手部攣縮以及右小腿與軀幹的緊繃，在初始症狀發生後一年半，經全皮層皮膚切片病理診斷爲嗜伊紅性筋膜炎。超音波檢查顯示左手掌腱鞘明顯增厚、回音減少，右側腓腸肌的表淺筋膜增厚。經過類固醇治療八個月，左側手腕/手部攣縮改善，超音波的表現也趨於正常。（台灣復健醫誌 2012；40(2)：91 - 95）

關鍵詞：嗜伊紅性筋膜炎（eosinophilic fasciitis），超音波（sonography），風濕免疫疾病（rheumatological disorder）

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