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MSUS ACADEMY

NEWSLETTER

Eosinophilic Fasciitis: A Rare Mimicker of Systemic Sclerosis

presented by
Seth VanDerVeer

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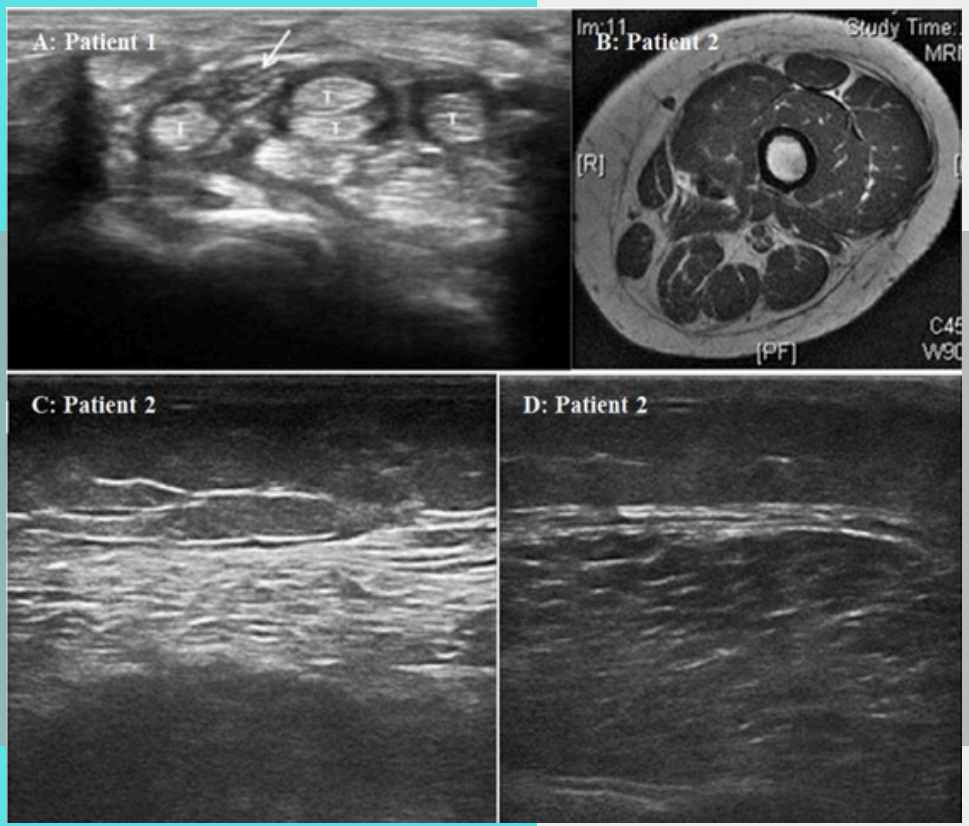
Eosinophilic fasciitis (EF) is a rare condition characterized by inflammation, thickening, and sclerosis of the skeletal muscle fascia. It can present with a variety of symptoms including induration and peau d'orange changes of the skin, stiffness in the distal limbs, arthralgias, myalgias, and joint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

https://pmc.ncbi.nlm.nih.gov/articles/PMC10902094/



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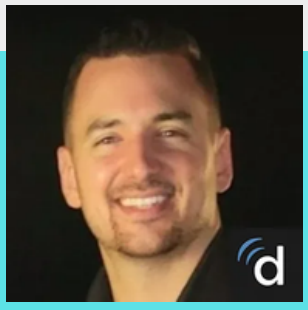
Interview with
Seth VanDerVeer



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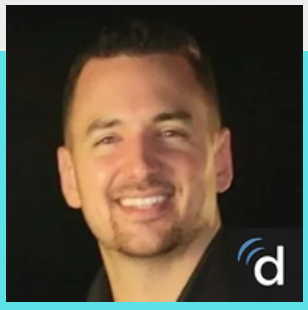
MSUS Academy: Shulman syndrome is a rare disease and a mimicker of systemic sclerosis. What is your opinion regarding the value of ultrasound in diagnostics, compared to clinical examination and MRI (https://pubmed.ncbi.nlm.nih.gov/37340516/)?

SV: For disorders of skin thickening, the clinical history and physical exam are of paramount importance. When approaching these conditions, I feel it is most beneficial to first identify if you are dealing with systemic sclerosis or a scleroderma mimic. Key diagnostic clues for a scleroderma mimic include absence of Raynaud's phenomenon with normal nailfold capillaries, atypical distribution of skin thickening with sparing of the hands, feet, and face, and lack of characteristic organ involvement seen in systemic sclerosis. For Shulman syndrome (eosinophilic fasciitis (EF)) specifically, two characteristic clinical features include the "peau d'orange" or pseudo-cellulite appearance of the skin with a woody, firm texture and longitudinal skin depressions along the path of superficial veins, known as the groove sign. Although full-thickness skin biopsy remains the diagnostic gold standard, the literature reflects an evolving clinical practice favoring increased reliance on MRI to confirm the diagnosis without an invasive procedure. When compared against ultrasound, MRI is preferable for diagnosis because it offers a higher sensitivity and specificity and provides a comprehensive assessment of disease extent in a single scan. MRI in EF demonstrates symmetric thickening and hyperintensity of the superficial and deep muscle fascia on T2-weighted and STIR sequences with strong enhancement after IV contrast administration, findings which also correlate well with disease activity and can be used to monitor response to treatment. However, musculoskeletal ultrasound provides complementary diagnostic value in several situations and is particularly useful when MRI is unavailable or contraindicated. Importantly, the most reliable distinguishing feature between systemic sclerosis and EF is the depth of tissue involvement, dermal/subcutaneous in SSc versus fascial in EF, which is readily visualized on ultrasound. Ultrasound can demonstrate the characteristic findings of EF including fascial thickening, disorganization of myofibrils adjacent to the superficial fascia, fascial edema, and subcutaneous edema in a rapid, point-of-care assessment. In addition, ultrasound is particularly valuable for identifying an optimal site for skin biopsy and monitoring treatment response. Furthermore, conventional ultrasound can be combined with shear-wave elastography (SWE) for additional objective data. Through quantitative assessment of tissue elasticity, it can provide an effective means of tracking therapeutic response without the need for repeated invasive biopsies or costly MRIs. This is an important consideration in EF because the condition often requires prolonged treatment with combination immunosuppressive therapy.



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MSUS Academy: Can you highlight the diagnostic use of ultrasound for patients with erythema nodosum (https://onlinelibrary.wiley.com/doi/full/10.1111/srt.13112)?

SV: Erythema nodosum (EN) is known as the most common cause of septal panniculitis and presents as erythematous, tender and indurated nodules. On visual inspection, they appear as ill-defined areas of erythema, but palpation will reveal tender, deep-seated nodules in the subcutis. When ultrasound is performed using high-frequency probes, it can visualize subcutaneous inflammation and help identify the characteristic septal pattern of panniculitis in EN. In the literature, the sonographic findings been described as a "jigsaw" pattern with marked hypoechogenicity of the subcutaneous fat lobules and intervening thickened, hypoechoic septa with positive Doppler signal. However, these findings are not specific to EN and can be seen in many other vasculitic and non-vasculitic septal panniculitides, such as cutaneous polyarteritis nodosa, Behcet's disease, necrobiosis lipoidica profunda, deep morphea, lipodermatosclerosis, and subcutaneous granuloma annulare, to name a few. Furthermore, although EN is classically described as a septal panniculitis, a diagnostic dilemma arises in early, atypical, or recurrent cases (the cases most likely to be biopsied) where EN can display features falling outside of the typical septal pattern including lobular panniculitis, vasculitis, and even suppuration. Identification of these additional features further expand the differential to include various other types of panniculitis, such as lupus, pancreatic, infectious, and alpha-1-antitrypsin. Because of the clinical heterogeneity of EN as well as the sizable differential for panniculitis, ultrasound has a limited role in the diagnosis of EN and serves mainly as a supplementary tool to visualize subcutaneous inflammation rather than a definitive diagnostic method. Thus, in my view, a more appropriate potential role for ultrasound use in EN would be monitoring response to treatment after the diagnosis is confirmed histologically. Ultimately, the diagnosis of EN remains predominantly clinical with histopathologic confirmation via deep incisional or excisional biopsy when needed.

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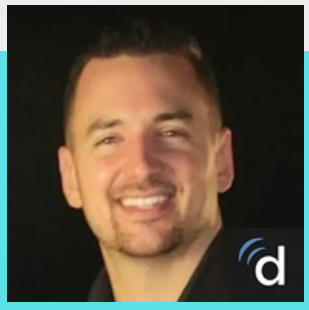

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MSUS Academy: Now on to another topic. What is your opinion and experience with synovial biopsies in everyday rheumatology practice?

SV: Synovial biopsy plays a crucial diagnostic role in the rheumatology space. The main indication for acquiring one is to evaluate a chronic monoarthritis lasting > 6 weeks, for which the most important diagnosis to rule out is infection. If an infection has been ongoing for this duration or longer without significant joint destruction, you are often dealing with a more indolent organism such as a fungus or mycobacterium. During my fellowship training and beyond, I have already seen a handful of cases of wrist monoarthritis resulting from non-tuberculous mycobacterium in patients who enjoy gardening. However, it is worth noting that several autoimmune rheumatic diseases can present initially as a monoarthritis. The most common of these include spondyloarthritis (psoriatic arthritis, IBD-associated, reactive arthritis) and rheumatoid arthritis but can also include systemic lupus erythematosus and Behcet's disease. As a result, it is also common to be left with a nondiagnostic biopsy because synovial biopsy alone cannot reliably differentiate between systemic connective tissue diseases. In this setting, the practitioner is left to symptomatic management and continued surveillance for new or evolving symptoms.

MSUS Academy: Finally, do you have any advice for young colleagues in training using musculoskeletal ultrasound?

SV: In my opinion, musculoskeletal ultrasound can be broken down into three fundamental components: anatomy, pathology, and technical skills. My suggestion is to always focus on anatomy first. When you are learning ultrasound, make it your goal to identify all the anatomical structures on the screen in each of the standard views for each joint. Jon Jacobson's Fundamentals of Musculoskeletal Ultrasound is a great source for this, which provides you with sample scanning protocols and structures of interest for each region. Once you have a framework, I would recommend scanning as much as possible. As you scan, ensure that you evaluate the structures of interest completely in orthogonal views and learn to follow these structures proximally and distally. In the early days, I would focus more on asymptomatic people (i.e. yourself, peers, family members), which will help you develop a strong foundation in anatomy and appreciate the range of normal findings that can be seen. By investing in this foundation early, you will gain much more knowledge from any structured teaching that you pursue in the future. As you continue to scan patients and gain more experience, shift your focus to pathology and regularly consult ultrasound atlases for reference. Throughout your journey, seek mentorship, participate in ultrasound organizations, and take every opportunity to advance your skills. Pursue courses and conferences with as much hands-on scanning as possible and connect with the world's foremost experts on social media platforms. Embrace the concept of lifelong learning and always remember --- even a master knows that he or she is always a student.

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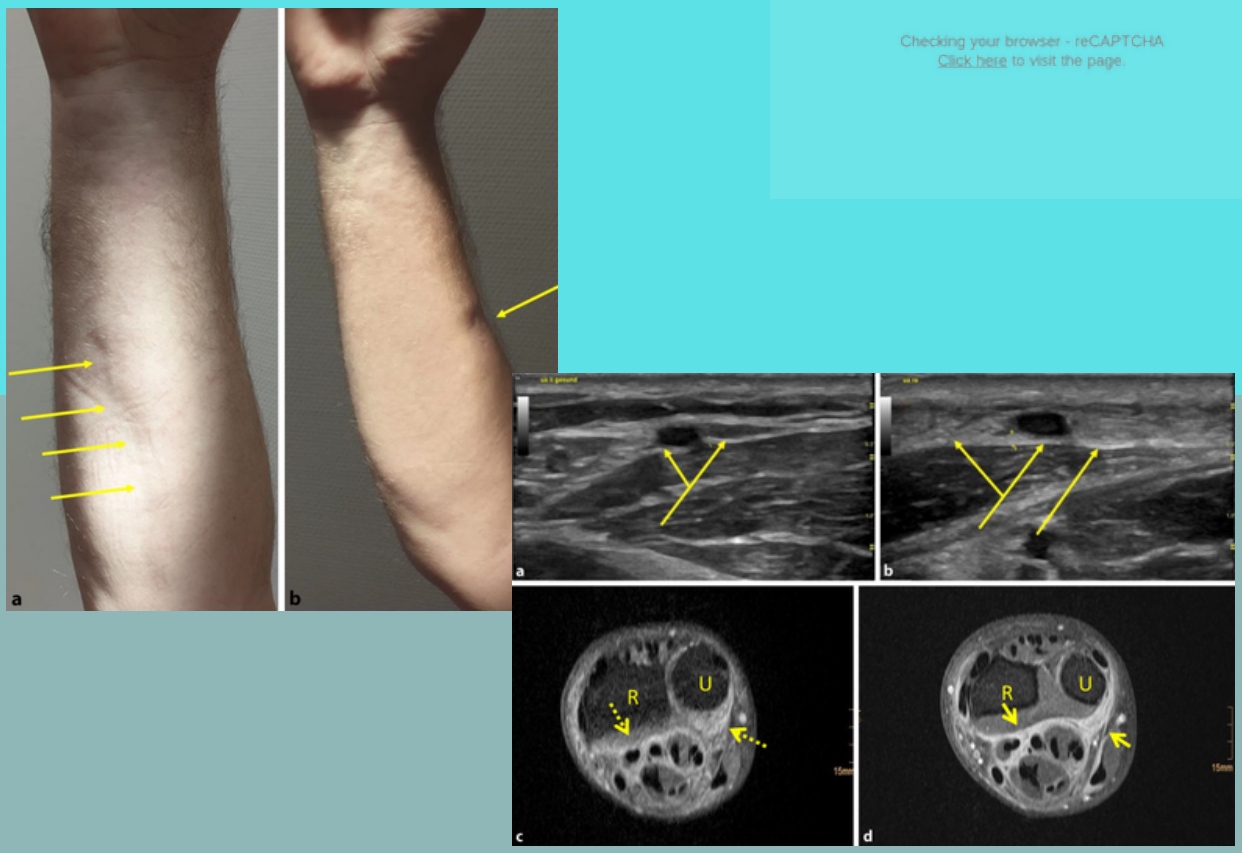
Publication of the month

Link

Imaging of eosinophilic fasciitis in ultrasound and MR

Abstract

Eosinophilic fasciitis (EF, also known as Shulman syndrome) is an uncommon connective tissue disease characterized by inflammatory thickening of the fasciae as well as swelling and hardening of the skin. It mostly affects the lower extremities. Swollen and indurated skin, together with the groove sign, are typical clinical signs. So far, biopsy evidence of inflammation and thickening of the fascia has been the gold standard for diagnosis. Magnetic resonance imaging (MRI) is mentioned in the literature as an alternative method for confirming the diagnosis. We present a case of asymmetric EF in a 54-year-old German male. He came with painful induration of the right forearm, with a characteristic groove sign and limitation of motion of the right hand. The blood count revealed eosinophilia with 0.57 G/l or 9.6% (normal: 0.05–0.5 G/l and 0.5–5.5%), ANA and ENA were negative. The diagnosis was confirmed histologically and we were able to detect a thickened fascia in MRI and ultrasound imaging. The EF also appeared in the left lateral malleolus during the course of the illness. Treatment was carried out with prednisolone and methotrexate.



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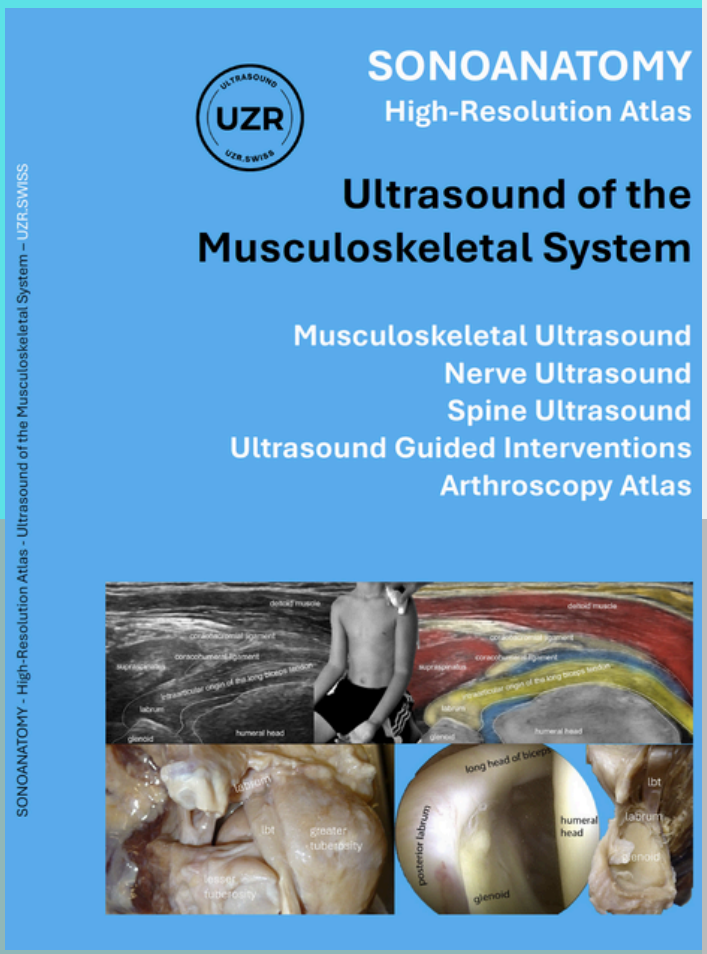

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