Skeletal Radiol (2009) 38:729–730
DOI 10.1007/s00256-009-0695-9

TEST YOURSELF: ANSWER

Bilateral tight swollen legs: diagnosis and discussion

Dorota Linda · Ashwyn Rajagopalan · Samih Salama · Nader Khalidi · Srinivasan Harish

Published online: 22 April 2009
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Diagnosis

Diagnosis is eosinophilic fasciitis.

Discussion

Eosinophilic fasciitis (EF) was first described by Shulman [1] in 1974 as an autoimmune syndrome characterized by diffuse fasciitis with hyperglobulinemia and eosinophilia. EF is a relatively rare disease with just over 200 cases reported in the literature. EF possesses striking clinical similarities to scleroderma and inflammatory myopathies, but, unlike scleroderma, visceral involvement and Raynaud's phenomenon are classically absent. Our patient was initially diagnosed as scleroderma but was subsequently shown not to satisfy the clinical criteria for the diagnosis.

The case presentation can be found at doi:10.1007/s00256-009-0694-x.

D. Linda · S. Harish (✉)
Department of Diagnostic Imaging,
St. Joseph's Healthcare and McMaster University,
Hamilton, Ontario, Canada L8N 2A6
e-mail: sriniharish@gmail.com

A. Rajagopalan · S. Salama
Department of Pathology,
St. Joseph's Healthcare and McMaster University,
Hamilton, Ontario, Canada

N. Khalidi
Department of Rheumatology,
St. Joseph's Healthcare and McMaster University,
Hamilton, Ontario, Canada

Patients most frequently present with inflammatory swelling and induration of the arms or legs. The hands and feet are less often implicated. There may be skin hyperpigmentation and thickening, with a peau d'orange appearance (Fig. 1). Muscle weakness, extremity stiffness, and joint contractures can occur [1].

Characteristic laboratory features of EF include peripheral eosinophilia, hypergammaglobulinemia, and elevated erythrocyte sedimentation rate. Laboratory analysis in our patient revealed a negative serum antinuclear antibody, an elevated eosinophil count, and a normal creatine kinase.

EF may occur after new strenuous exercise (as was the case in our patient), ingestion of L-tryptophan, environmental exposure to various chemical substances, and with the use of simvastatin.

Magnetic resonance imaging (MRI) demonstrates characteristic findings including skin thickening (white arrows in Fig. 2a–c), fascial edema (blue arrows in 2b), and contrast enhancement of the fasciae (blue arrows in Fig. 2b–c). MRI is also beneficial in guiding the choice of biopsy site [2, 3]. MRI can also sometimes reveal edema and enhancement, as in our case (yellow arrows in Fig. 2a), in muscle tissue adjacent to fascia [2, 3]. The muscle edema seen on MRI in our case was not proven at histology, as a different part of the fascia was biopsied. MRI is useful to serve as a marker for disease activity, identifying disease recurrence and response to therapy [3]. The MRI is quite characteristic for EF, but fascial abnormalities have been noted in other conditions such as graft-versus-host disease and deep and generalized morphea [4]. Definitive diagnosis can only be established by full-thickness skin-to-muscle biopsy [2]. Histologically, the disease is characterized by fibrous or fibromyxoid inflammation of the superficial muscle fascia with lymphocytes and plasma cells (Fig. 3a and arrowheads

in Fig. 3b). Eosinophils may be infrequent (yellow arrow in Fig. 3b) and may not be seen in every histological section. Perivascular inflammation can be seen in the dermis (arrows in Fig. 3c). Patients generally respond well to corticosteroid treatment, highlighting the need for early diagnosis and treatment [1]. Our patient was started on oral steroids and methotrexate with moderate improvement in symptoms and MRI features at a 4-month follow-up.

In conclusion, MRI demonstrates characteristic findings in EF and is useful in the diagnosis and management of the same.

References

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