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Eosinophilic fasciitis: demographics, disease pattern and response to treatment: report of 12 cases and review of the literature

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Abstract

Background Eosinophilic fasciitis is a rare scleroderma-like illness. The clinical spectrum of the disease has evolved since its initial description.

Methods We identified all patients diagnosed with eosinophilic fasciitis over the past 10 years at our scleroderma clinic. Demographics, disease pattern, serologies, tissue pathology and response to treatment were all recorded.

Results Twelve patients with eosinophilic fasciitis were identified in our clinic over the past 10 years. The mean age at diagnosis was 49.8 ± 9.8 years, with nine female and three male patients. The first symptoms were noticed at an average of 8.8 ± 6.1 months before diagnosis. The mean initial absolute peripheral blood eosinophil count was 1188 ± 1059 cells/L. Two patients had a monoclonal gammopathy, and two had positive ANA titers. All patients received corticosteroids, 10 of whom received the equivalent dose of > 20 mg/day of prednisone for more than a month. Five patients received hydroxychloroquine, two received methotrexate, one received cyclosporine, one received topical tacrolimus, and one received sulfasalazine. At a mean follow up of 17.6 months (range 2–94 months), 8 patients had a good response to treatment, 2 patients had no effect, and 2 patients had a poor response to treatment.

Conclusion High dose corticosteroid treatment lasting longer than a month with

or without an immunosuppressive agent helped most patients with eosinophilic fasciitis, best results seen in those patients who were initiated treatment early on after their first symptoms.

Introduction

Eosinophilic fasciitis is a scleroderma-like syndrome first described in 1974 by Shulman in patients with diffuse fasciitis and eosinophilia.¹ The clinical spectrum of this disease has broadened since its first description, as more patients are recognized and diagnosed. Eosinophilic fasciitis is a rare disorder with varying clinical presentations, making its clinical definition challenging.^{2,3}

Patients with eosinophilic fasciitis typically report swelling and induration of the arms and legs with skin thickening. The usual stages of the disease progress from edema of the extremities, to peau d'orange with hyperpigmentation, to woody induration with skin tightness. Localized morphea, defined as inflammation localized to the reticular dermis and superficial panniculus, has been reported. Synovitis and contractures may also be present. Some patients have presented with rapidly progressive muscle weakness with associated pain and stiffness of the extremities.² A variety of extracutaneous manifestations have been reported including arthritic, pulmonary, hematologic, and neoplastic.⁴⁻⁶ The diagnosis of eosinophilic fasciitis may be delayed, as it has overlapping features of other diseases. Scleroderma, polymyositis, hypereosinophilic syndrome, and Churg-Strauss vasculitis may all be in the differential diagnosis for a patient presenting with eosinophilic fasciitis.

Diagnosis is made by a full thickness skin to muscle biopsy, showing inflammation and thickening of collagen bundles in the superficial muscle fascia with infiltration of lymphocytes and plasma cells. Eosinophils are occasionally seen on biopsy but are not necessary to make the diagnosis. More recent -studies have been conducted using MRI for the diagnosis of eosinophilic fasciitis.⁷⁻⁹ In a retrospective study involving 6 patients, MRI was able to detect fascial thickening and signal abnormalities in patients with eosinophilic fasciitis at the time of diagnosis⁷. Laboratory analysis for eosinophilic fasciitis typically reveals an elevated sedimentation rate, peripheral eosinophilia, and hypergammaglobulinemia. The degree of eosinophilia does not correlate with disease severity and laboratory results are not helpful in following disease activity.² Some patients have shown normalizing laboratory results with persistent clinical evidence of eosinophilic fasciitis.^{2,7}

The etiology of eosinophilic fasciitis is uncertain. Reports of extreme physical exertion as well as ingestion of certain pharmaceuticals such as l-tryptophan and statins have been suggested to have a pathogenic role.^{2,10-12} Other potential triggers that have been suggested are trauma, arthropod bites and borreliosis.¹³⁻¹⁷

No clear consensus exists regarding the demographics of eosinophilic fasciitis. The mean age of onset has consistently been found to be between 40 and 50 years of age with a wide range reported from early childhood to the elderly.^{2,3} It remains unclear whether race and family history are risk factors in developing eosinophilic fasciitis.

The mainstay of therapy for eosinophilic fasciitis has largely been corticosteroid therapy, although multiple drug regimens have been tried. High-dose corticosteroids have shown some efficacy, although steroid-refractory cases have been documented. Cyclosporine, hydroxychloroquine, cimetidine, azathioprine, and d-penicillamine have all been used for the treatment of eosinophilic fasciitis.^{2,3,18,19} These drugs showed variable results and were often used in steroid-refractory cases. Recent work has also suggested the potential use of antitumor necrosis alpha inhibitors, such as infliximab, to manage resistant cases not responsive to typical therapy.²⁰ Evaluating the response to treatment can be complicated since the natural history of the disease can involve spontaneous remission². Lastly photochemotherapy has also been used with some success.^{21,22}

Sample size remains a limiting factor in studying eosinophilic fasciitis. The disease is rare and the incidence is not known. More data is needed to determine the etiology of the disease and its demographic distribution. More information needs to be gained in the treatment of the disease, particularly in steroid-refractory cases. In this study, we attempt to gain more information regarding demographics, risk factors, disease patterns and response to treatment in 12 patients who presented to our scleroderma clinic over a 10 years span. Our goal is to help clarify these parameters of eosinophilic fasciitis in which there have been few answers due to the small patient population.

Methods

Employing a retrospective chart review, we looked at all patients who were seen at our scleroderma clinic within a 10 year span that had a diagnosis of eosinophilic fasciitis. A total of 12 patients were identified with eosinophilic fasciitis during this time period. These patients were diagnosed clinically by

rheumatologists at the scleroderma center of our institution. All patients were referred to the scleroderma center for clinical presentations of a scleroderma-like illness.

We compiled a database including the demographics, duration and quality of presenting symptoms, laboratory results and imaging results for each patient. Data regarding treatment regimens, follow-up, and clinical response were also collected, as well as family, social and past medical history.

The history of each patient's illness at the time of presentation was documented and included the amount of skin involved, the time of onset of symptoms, the presence of Raynaud's phenomenon, contractures, carpal tunnel, and arthritis. Any correlation of disease symptoms to strenuous exercise, medication use or other potential triggers was documented. At the initial visit, patients were examined to assess the distribution, extent, and quality (morphea, p'eu d'orange, induration) of skin involvement. Patients were also examined for extracutaneous manifestations of eosinophilic fasciitis.

Laboratory analysis included complete blood count with leukocyte differential, serum protein electrophoresis, rheumatoid factor, antinuclear antibody, creatine phosphokinase, aldolase, antitopoisomerase I and anticentromere antibodies, sedimentation rate, and C-reactive protein. The results of other studies including EMGs, MRIs, 2D echocardiograms, and pulmonary function tests were documented when available. Ten patients underwent full thickness skin to muscle biopsy to confirm the diagnosis of eosinophilic fasciitis and all slides were reviewed by a rheumatologist at our clinic.

Treatment was initiated as clinically necessary and each patient's drug regimen was documented as well as any clinical change found at follow up visits. Response to therapy was recorded as good when there was more than 40% of skin regression based on the total body surface involvement and the modified Rodnan skin score, poor when there was continued skin progression, or no effect if the skin findings remained the same or had less than 40% skin regression.

Results

A total of 12 patients were identified with eosinophilic fasciitis at the scleroderma center of our institution. Of these, nine were female and three were male. The mean age at presentation was 49.8 ± 9.8 years of age. Ten of the patients identified themselves as Caucasian and two as African American. The average time of diagnosis from the first symptoms of eosinophilic fasciitis was 8.8 ± 6.1

months (Table 1). For patients with a poor outcome this was 12.5 ± 7.2 months while for patients with a good outcome this was 7 ± 4.5 months.

All patients had been given other initial diagnoses based on signs that eventually led to the diagnosis of eosinophilic fasciitis. The patients studied had visited an average of 2.4 physicians, ranging from one to seven, before being referred to our clinic and diagnosed with eosinophilic fasciitis. Six patients had an initial diagnosis of tendonitis. Of the remaining patients, one was diagnosed with Lyme disease, one with fibromyalgia, one with intertigo, and one as a skin manifestation of pregnancy. One patient was diagnosed with both tendonitis and carpal tunnel syndrome. Only four patients related that their symptoms appeared after vigorous exercise. None of our patients reported using products containing l-tryptophan or statins. None of our patients had any recent trauma.

Upon questioning, five patients admitted to being smokers: two had a 10 pack year history, one had a 20 pack year history, and two had a 30 pack year history. Past medical history revealed two patients with past diagnoses of cancer, one breast and the other chronic lymphocytic leukemia. Half of the patients reported a family history of cancer in a first degree relative. Of these patients, four had a family history of breast cancer, one ovarian cancer, and one a cardiac cancer.

Cutaneous manifestations

All patients presented with skin induration (Fig. 1), while seven patients displayed peau d'orange and three patients had morphea. The average affected total body surface area was $20.1 \pm 17.9\%$. The average skin tightness score based on the Modified Rodnan Skin Score, which is typically used in scleroderma, was 11.1 ± 7.4 out of a possible maximal score of 51. Eleven patients had lower extremity involvement. Upper extremity involvement was seen in nine patients. Of these patients, upper extremity involvement affected only the hand in one, only the forearm in two, and both forearm and hand in the remaining six (Table 1).

Extracutaneous manifestations

Of the extracutaneous manifestations described in the literature, our patients displayed joint contractures, inflammatory arthritis, and carpal tunnel syndrome. No patients had experienced Raynaud's phenomenon or had nailfold capillaroscopy changes. Of the extracutaneous manifestations, joint contractures were the most common, affecting eight patients. Of these patients, four involved the wrists, three involved the knee, three involved the elbow, two involved the ankle, two involved the hand, and one involved the shoulder. Five patients were documented as having inflammatory arthritis affecting the feet, wrists, and knees. Only two patients had documented carpal tunnel syndrome.

Laboratory analysis

Leukocyte differential was recorded for each patient and hypereosinophilia was found in 10 patients. Two patients had eosinophil levels within normal limits. The mean initial peripheral eosinophil count was $12 \pm 10.7\%$ (absolute eosinophil count 1188 ± 1059 cells/L, with a range from 49 to 4059 cells/L). The mean absolute eosinophil count in the patients who were diagnosed within 6 months of initial symptoms was 1762 cells/L as compared to 784 cells/L in the patients who were diagnosed more than 6 months after their initial symptoms though this did not reach statistical significance ($P = 0.12$).

Serum protein electrophoresis was documented in all patients, two of which had a monoclonal gammopathy. One patient had an elevated IgG lambda, and the other an IgA kappa light chain. Rheumatoid factor was only positive in one out of the 12 patients tested. Antinuclear antibodies were found in two patients, one with a titer of 1 : 80, in a speckled pattern, and one with a titer of 1 : 40 in a homogenous pattern. Muscle enzymes documented in all patients were normal. All patients had a negative antitopoisomerase I and anticentromere antibodies.

EMG studies were performed in six patients and were abnormal in only one patient with bilateral carpal tunnel syndrome. Four patients underwent MRI studies with only one patient showing findings suggestive of eosinophilic fasciitis. This patient showed evidence of disease activity in the right forearm on T2 weighted MRI images (Fig. 2). Results for 2D echocardiography were reported for nine patients, all of which had normal ejection fractions. Right systolic ventricular pressure (RSVP) ranged from 20 mmHg to 40 mmHg in these patients.

Ten patients underwent full thickness skin to muscle biopsy and histopathology of eosinophilic fasciitis was documented including thickened collagen bundles in the deep reticular dermis (Figs 3–5).

Seven patients had documented pulmonary function tests, including forced expiratory volume (FEV1), forced vital capacity (FVC), total lung capacity (TLC), and diffusing capacity (DLCO). The mean FEV1 was 89.5%, ranging from 64 to 118% of predicted, the mean FVC was 93.2% with a range of 71–106% of predicted, the mean TLC was 98.6%, ranging from 75 to 121% of predicted and the mean DLCO was 105.9% and ranged from 76 to 147% of predicted.

Treatment

All patients received corticosteroids with 10 patients receiving the equivalent to prednisone > 20 mg/day as an initial dose, while other medications included

hydroxychloroquine, topical tacrolimus, methotrexate, sulfasalazine and cyclosporine. (Table 2). At a mean follow-up of 17.6 months (range 2– 94 months) eight patients had documented marked clinical improvement of cutaneous and extracutaneous manifestations and were categorized as having a “good” response to therapy. The clinical course for two patients was documented as progressive, with quantitative and qualitative progression of the skin manifestations, and were categorized as having a “poor” response to therapy. The remaining two patients had no change in their presenting cutaneous and extracutaneous manifestations after pharmacotherapy, and were labeled as having “no effect” in response to treatment (Table 2).

Discussion

Eosinophilic fasciitis is a rare disease characterized by progressive skin thickening. Patients report initial swelling and cutaneous thickening, usually involving the extremities that can progress to peau d’orange and then induration.² Extra-cutaneous manifestations include, joint contractures, inflammatory arthritis, restrictive lung disease and pleural effusions.²³ Monoclonal gammopathy and multiple myeloma have been at times associated with eosinophilic fasciitis.⁵

Currently, full thickness skin to muscle biopsy is the accepted gold standard for diagnosing eosinophilic fasciitis. Biopsy typically reveals thickening of the dermis due to dense deposition of collagen while the fascia has a lymphocytic and eosinophilic infiltrate, especially early on in the disease. The inflammatory infiltrate may involve the panniculus and muscle and can lead to fibrinoid necrosis (Figs 3–5). Several studies comparing MRI to biopsy have been published showing MRI as a useful aid in the diagnosis of eosinophilic fasciitis as well as helpful in following a patient’s response to therapy. Both biopsy and MRI show evidence of fascial thickening during clinically active disease.^{7–9,24}

Although eosinophilic fasciitis is a rare disease, its clinical manifestations can mimic other syndromes. Scleroderma patients present with similar cutaneous involvement, making distinction between these two diseases difficult, this further complicated by the fact that the treatment of these diseases are similar. Patients with early isolated skin involvement may present to a dermatology practice for a suspected rash, limiting the cases seen in rheumatology clinics to those patients who have extensive skin disease that mimics scleroderma. This in turn suggests that cases seen in a dermatology clinic may have more limited skin involvement as compared to those patients who are seen in scleroderma clinics. The clinical picture of eosinophilic fasciitis may present similarly as a variety of other diseases as evidenced by the range of initial diagnoses that were given to the patients presented in this study.

Of the 12 patients identified with eosinophilic fasciitis, 75% were female. Some studies acknowledge that eosinophilic fasciitis is more common in males² however, other studies in addition to ours have shown a female predominance.³ The mean age of our patients was 49.8 ± 9.8 years of age which is consistent with the mean age reported in other studies.²

The etiology of eosinophilic fasciitis remains unclear. In our study, only four patients reported a correlation between symptom onset and intense physical exertion. Other studies have documented larger percentages (24 out of 52, and 4 out of 6) of patients with possible exercise induced eosinophilic fasciitis.^{2,8} None of our patients reported using products containing l-tryptophan or statins. None of our patients had any recent trauma, while only one patient was previously diagnosed with lyme disease.

Cutaneous manifestations of eosinophilic fasciitis have been described as progressive, with the earliest stage being edema of the extremities, followed by peau d'orange with hyperpigmentation, and finally induration.² Interestingly, all 12 patients in our study had documented skin induration at the time of presentation. Only seven patients were documented as having peau d'orange. This may represent interobserver error. It is also possible that the patients who were seen in our clinic presented later in their disease and therefore displayed the most advanced stage of cutaneous involvement. Eleven out of 12 patients had lower extremity involvement, indicating that this is a commonly affected site. Although fewer patients had upper extremity involvement, it was still a site of disease in 75% of patients. In our study patients more commonly had involvement of both the hand and forearm, however, isolated hand and isolated forearm involvement was also documented.

Joint contractures were present in two-thirds of the patients studied, with wrist being the most commonly involved joint, affecting half of the patients. However, several joints were reported to be affected including the ankle, elbow, hand, knee and shoulder. Inflammatory arthritis was seen in less than half of the patients studied. Both large and small joints were affected, more specifically the hand, knee, and wrist.

Hypereosinophilia was documented in the majority of our patients, however, two patients maintained normal eosinophil counts. This is consistent with the previous literature, which indicated that peripheral eosinophilia is not necessary to make the diagnosis of eosinophilic fasciitis and does not correlate with the clinical severity of the disease.² While the majority of patients did have laboratory

data indicating hypereosinophilia, only three patients had leukocytosis.

Serum protein electrophoresis was performed in each patient, with two patients having a monoclonal gammopathy. Abnormal serum protein electrophoresis results have been previously reported in the literature.⁵

MRI has been studied as an alternative means by which to diagnose eosinophilic fasciitis as well as to follow the efficacy of treatment. The findings are those of fascial thickening, with increased uptake on fluid-sensitive sequences and enhancement after IV contrast administration. MRI T2 weighted images showing fascial signal hyperintensity describe disease activity while T1 weighted images showing fascial thickening describe disease chronicity.⁷⁻⁹ While diagnosis is only definitive by full-thickness skin to muscle biopsy the potential use of MRI for diagnosis appears promising. In our sample, only four patients underwent MRI at the time of diagnosis, with only one patient showing MRI evidence of fascial thickening. Clinically, this patient had induration of his right forearm, visualized on MRI, and his lower extremities, as well as peau d'orange of his axilla. Interestingly, this patient had the highest eosinophilia count (41%) and believed his symptoms were induced by exercise. Unfortunately, this patient did not undergo a full thickness skin to muscle biopsy so we were unable to correlate the MRI findings to tissue histopathology.

While eosinophilic fasciitis is a rare disease, its cutaneous manifestations present similarly to other diseases frequently encountered in rheumatology. This raises the possibility that the prevalence of eosinophilic fasciitis is higher than has been previously documented, with some patients being misdiagnosed. It is therefore important to develop a clinical picture of eosinophilic fasciitis in order to differentiate it from other syndromes, as well as provide optimal therapy and counseling to the patient.

Systemic corticosteroids are the mainstay of therapy with a better response in those patients who present early and potentially have a more inflammatory rather than fibrotic skin lesion. Typical course of therapy is a dose of prednisone > 20 mg/day tapered over many months as the disease improves. Other therapies have been used either for corticosteroid sparing or for patients not responding to this form of therapy. Hydroxychloroquine² histamine 2 antagonists^{3,19,25,26} photochemotherapy^{21,22} immunosuppressive agents¹⁸ and anti-TNF-alpha inhibitors²⁰ have all been used. Physical therapy should also be initiated early on to limit joint contractures and maintain mobility.

This study presents clinical, historical, and laboratory information of 12 patients

with eosinophilic fasciitis. While more data is necessary in order to determine the etiology, clinical course, and response to treatment of the disease, we feel that the information these 12 patients have provided is a useful addition to the limited data that has been published on eosinophilic fasciitis.

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TABLES & FIGURES

Table 1

Eosinophilic Fasciitis patient demographics

Characteristics	Values
Age	49.8 ± 9.8 years old
Gender	9 female/3 male
Race	10 Caucasian/2 African American
Time from first symptoms to diagnosis	8.8 ± 6.1 months
Exercise induced	4/12
Skin induration	12/12
Peau d'orange	7/12
Hands	7/12
Forearm	8/12
Lower extremity	11/12
Total body surface involvement	20.1 ± 17.9%
Modified Rodnan skin score (maximum score = 51)	11.1 ± 7.4/51
Joint contractures	2 hand/4 wrist/3 elbow/2 knee/2 ankle/1 shoulder
Inflammatory arthritis	5/12
Malignancy	2/12 (chronic lymphocytic leukemia, breast cancer)

Table 2**Eosinophilic Fasciitis treatment response**

Patient	Treatment	Follow up	Response
1	Hydroxychloroquine 200 mg bid, methylprednisolone 20 mg/day with slow taper over 4 months	13 months	Good
2	Tacrolimus (topical), hydroxychloroquine 200 mg bid, prednisone 60 mg/day with slow taper over 3 months	4 months	No effect
3	Hydroxychloroquine 200 mg bid, Prednisone 60 mg/day with slow taper over 2 months	2 months	No effect
4	Sulfasalazine 500 mg bid, methotrexate 20 mg/week, Prednisone 10 mg/day for 6 months	18 months	Good
5	Cyclosporine 75 mg bid for 10 months, then methotrexate 25 mg/week for 12 months, prednisone 60 mg/day tapered over 2 months.	30 months	Good
6	Methotrexate 20 mg/week for 25 months, hydroxychloroquine 200 mg bid for 40 months, Prednisone 20 mg used early in diagnosis on and off for 24 months	94 months	Good, recurred 2 year later
7	Hydroxychloroquine 200 mg bid for 10 months, Prednisone 60 mg/day tapered over 6 months	13 months	Good
8	Prednisone 30 mg/day tapered over 12 months	12 months	Poor
9	Prednisone 30 mg/day tapered over 4 months	4 months	Good
10	Prednisone 40 mg/day tapered over 4 months	4 months	Good
11	Prednisone 60 mg/day tapered over 4 months	4 months	Good
12	Dexamethasone 6 mg/day tapered over 12 months	12 months	Poor

Figure 1

Eosinophilic fasciitis related skin induration of the leg



Figure 2

**T2 weighted MRI of the forearm in a patient with eosinophilic fasciitis.
*Arrows point to inflammatory changes in the fascia.**

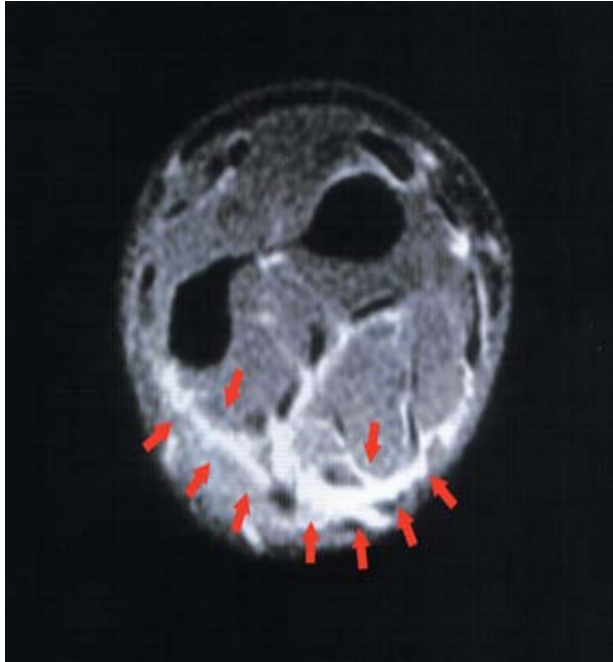


Figure 3

Fibrinoid necrosis and lymphocytic infiltration of the fascia (H&E x 100)

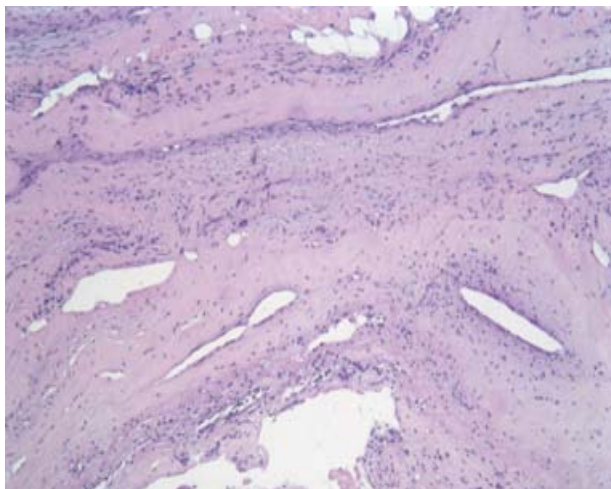


Figure 4

Cellular accumulation of lymphocytes and eosinophils in the fascia with extension into the panniculus (H&E x 200)

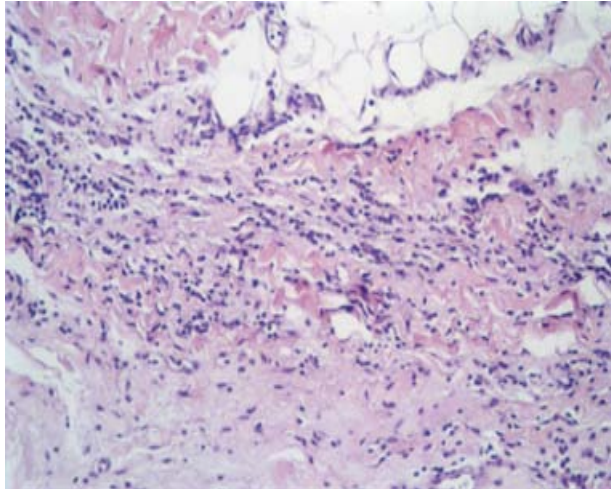


Figure 5

Cellular accumulation of lymphocytes and eosinophils in the fascia (H&E x 400)

