Eosinophilic Fasciitis And Associated Late Emerging Hodgkin Lymphoma

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Abstracts: Eosinophilic fasciitis is often seen between the ages of 40-50, and is characterized with skin thickening, and etiopathogenesis is still unclear. Patients diagnosed as eosinophilic fasciitis have accompanying hematologic diseases in less than 10%. In cases reported previously in the literature with a diagnosis of eosinophilic fasciitis, accompanying hematological malignancies and solid organ malignancies were within the first 5 years after diagnosis. We here present, in this case 58 years old male who diagnosed hodgkin lymphoma after 21 years of eosinophilic fasciitis. [Somak N. NJIRM 2015; 6(2):105-107]

Key Words: Eosinophilic fasciitis, Hodgkin lymphoma

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Introduction: Eosinophilic fasciitis has been described first time as a new syndrome by Shulman, characterised with peripheral eosinophilia, elevated erythrocyte sedimentation rate (ESR), hypergammaglobulinemia and scleroderma-like skin changes. Eosinophilic fasciitis has been reported in association with hematologic malignancies such as multiple myeloma, T-cell lymphoma, non-Hodgkin lymphoma, Hodgkin lymphoma besides breast cancer and colorectal malignancies. The diagnosis of a hematological malignancy in previously reported cases of eosinophilic fasciitis has been shown in the first five-years of follow-up. As far as it can be achieved in the literature, no case of a hematologic malignancy were seen 20 years after the diagnosis of eosinophilic fasciitis. We here present a case of Hodgkin lymphoma emerging 21 years after the diagnosis of eosinophilic fasciitis.

Case: A 58 year old male patient had fever for 1.5 months, only at evening first but continuing all day for a week, night sweats and swelling in his right inguinal region. On physical examination, his fever was 39 °C. He had 2 cm hepatosplenomegaly. In the right inguinal region firm, fixed conglomerate lymph nodes of about 3x3 cm in size were detected. A nodular lesion of 2x2 cm in diameter in the left anterior tibia was present. In the patient's history; it was learned that he was followed for 21 years with a diagnosis of eosinophilic fasciitis. Other system examination was normal. On the laboratory results, WBC was 2400/μl, eosinophil rate was 0.7%, and neutrophils were 62.8 %, lymphocytes 27.2 % and the hemoglobin 10 g / dL, platelet count was 126000/μl. Routine biochemistry tests were as follows: ALT 108 U / L, AST 143 U / L, ALP 982 U / L, GGT 974 U / L, LDH 891 U / L, albumin 2.83 g / dL, total protein 6.81 g / dL. Erythrocyte sedimentation rate was 87 mm / h, CRP was 124.6 mg / l, respectively. Antinuclear antibodies (ANA) and rheumatoid factor (RF) were negative. No organism grew in blood and urine cultures taken during periods of fever. Ig G was increased as 29045 g / I, IgM and IgA were within normal limits.

On the superficial ultrasonography of the right inguinal region, multiple conglomerate lymph nodes were observed, the largest was 23 mm in size in short axis. In abdominal ultrasonography, the liver was 200 mm, the spleen was 192 mm in size, and there were conglomerate lymph nodes 30 x 17 mm in size in the portal hilus. A right inguinal lymph node excisional biopsy was performed. Biopsy of the lesion revealed Hodgkin lymphoma in the type of nodular sclerosis. Pre-treatment positron emission tomography and computed tomography (PET-CT) examinations were performed and the patient was staged as 4SB. ABVD (adriamycin - bleomycin - vinblastine - dacarbazine) treatment protocol was initiated. The patient is still in hematology clinic for follow up and treatment.

Discussion: Eosinophilic fasciitis is often seen between the ages of 40-50, involves the skin, especially in the lower extremities, and is characterized with skin thickening. In the etiopathogenesis, denatured fatty acids, drugs, infectious agents, and trauma are blamed as a cause, but it is still unclear. Cutaneous edema and thickening of the skin are usually first lesions, but may progress to induration and peau d'orange appearance. Extracutaneous findings may be joint
contractures, inflammatory arthritis, carpal tunnel syndrome, Dupuytren’s contracture, pericarditis, pleural effusion, restrictive lung disease and proteinuria \(^1,5,16\).

Laboratory findings may include peripheral eosinophilia, hypergamaglobunemia, and increased erythrocyte sedimentation rate. Eosinophilia does not correlate with the degree and severity of the disease. In a series of 12 cases with a diagnosis of eosinophilic fascitis, eosinophil levels were in the normal range in two patients. In the same series, IgG lambda was increased in a patient, Ig Kappa light chain in another, while there were monoclonal gammopathy in two patients \(^16\). In our case, there were increases in the levels of IgG and IgM at the time of the diagnosis of eosinophilic fascitis.

The definite diagnosis of eosinophilic fasciitis is established based on the inflammation, thickening collagen fibers in the superficial muscle fascia infiltrated by lymphocytes and plasma cells in full-thickness tissue biopsy. Although sometimes eosinophils are seen, they are not required for a diagnostic biopsy \(^16\). Currently, international diagnostic criteria for eosinophilic fasciitis has not been established yet.

Patients diagnosed as eosinophilic fasciitis have accompanying hematologic diseases in less than 10 % \(^3-10\). It is difficult to determine whether eosinophilic fasciitis is a trigger or a paraneoplastic syndrome of the hematologic malignancy \(^1\). Most cases of malignancy and eosinophilic fasciitis demonstrate the properties of paraneoplastic syndromes. In cases reported previously in the literature with a diagnosis of eosinophilic fascitis, accompanying hematological malignancies and solid organ malignancies were within the first 5 years after diagnosis. Rodat et al. reported the development of Hodgkin lymphoma in a 70 year old female patient 3 years after the diagnosis of eosinophilic fasciitis \(^10\); and Micheals RM, reported a 53 year old female patient with the development of Hodgkin lymphoma after 3.5 years \(^9\). In our case, the diagnosis of Hodgkin’s lymphoma after 21 years of eosinophilic fasciitis history is the first in the literature as far as we can reach, and this is remarkable.

Today, the pathogenesis of eosinophilic fasciitis and associated lymphoma have not been clearly demonstrated yet. In cases of eosinophilic fasciitis associated T -cell lymphoma, the potential role of IL-5 in T-cell production has been implicated \(^1\). At the same time, a random association of lymphoma and idiopathic eosinophilic fasciitis could not have been excluded as well. Therefore, any definite conclusion has not been reached about the predisposition of this association. In addition, the age of the previously reported two cases were different from each other, our case was in the late period, and these make it difficult to guess the emergence of lymphoma with an age-related associaton.

**Conclusion:** In patients with a diagnosis of eosinophilic fasciitis, it should be kept in mind that hematological and solid organ malignancies can be seen after a period of approximately 20 years.

**References**


