Idiopathic Palmar Fasciitis with Polyarthritis Syndrome

A 31-yr-old Korean woman was presented with a 4-month history of bilateral hand swelling and stiffness. On clinical examination, she had a painful synovitis of both hands, wrists, knees and ankles. The radiologic and histological examinations confirmed it with palmar fasciitis and polyarthritis syndrome (PFPAS). PFPAS is an uncommon disorder characterized by progressive flexion contractures of both hands, inflammatory fasciitis, fibrosis, and a generalized inflammatory arthritis. Although most reported cases of PFPAS have been associated with various malignancies, our patient have not been associated with malignancy during 24 months follow up period from her first symptom onset. Her symptoms were improved with moderate dose of corticosteroid and she is currently taking prednisone 5 mg daily without any evidence for internal malignancy. We present here in a young Korean patient with idiopathic PFPAS who was successfully treated with administration of corticosteroid.

Key Words: Fasciitis, Arthritis, Malignancy

INTRODUCTION

Palmar fasciitis and polyarthritis syndrome (PFPAS) is an uncommon disorder characterized by progressive flexion contractures of both hands, inflammatory fasciitis, fibrosis, and a generalized inflammatory arthritis (1, 2). The course might be rapidly progressive and result to a loss of function of the upper extremity due to contractures and pain. In 1982, Medsgen et al. (1) described PFPAS as a disease entity, which should be differentiated from reflex sympathetic dystrophy (RSD) and Dupuytren’s contracture. More than 40 cases have since been reported, but the exact etiology of PFPAS is still not known. Most of them have been suggested associations with various malignancies (3, 4) and only a few reports were not associated with malignancy (5, 6).

We present herein a young woman with PFPAS in the absence of an associated internal malignancy.

CASE REPORT

A 31-yr-old Korean woman presented in February, 2004, with 4-month history of bilateral hand swelling and stiffness. On clinical examination, she had a painful synovitis of both hands, wrists, knees and ankles. The radiologic and histological examinations confirmed it with palmar fasciitis and polyarthritis syndrome (PFPAS). PFPAS is an uncommon disorder characterized by progressive flexion contractures of both hands, inflammatory fasciitis, fibrosis, and a generalized inflammatory arthritis. Although most reported cases of PFPAS have been associated with various malignancies, our patient have not been associated with malignancy during 24 months follow up period from her first symptom onset. Her symptoms were improved with moderate dose of corticosteroid and she is currently taking prednisone 5 mg daily without any evidence for internal malignancy. We present here in a young Korean patient with idiopathic PFPAS who was successfully treated with administration of corticosteroid.

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Idiopathic Palmar Fasciitis and Polyarthritis Syndrome

The underlying skeletal muscle showed well preserved myocytes, mild interstitial edema, and fibrosis. Marked interstitial infiltration, mainly of T lymphocytes and focal infiltration of B lymphocytes with scattered plasma cells, were observed on immunohistochemical stainings. There was no evidence of perifascicular atrophy, necrosis, granuloma or leukocytoclastic vasculitis.

Because of her unusual clinical features, she was screened for malignant disease. We performed endoscopic and radiologic examinations for internal organ to find out the presence of malignancy but those were negative. Examinations for the breast, thyroid, and pelvis showed no abnormal findings. Tumor markers including CA125, CA19-9, AFP, and CEA were each in normal ranges.

The patient started on prednisone 30 mg daily. Her symptoms such as arthralgia and bilateral hand swelling with stiffness had gradually improved over 2 months and the dosage of prednisone was tapered gradually. MRI study which performed 6 months after taking medication revealed no inflammatory reactions in both fascia and muscles of the forearm. Follow up bone scan also showed no increased uptake in all the joints. Twenty-four months after her first symptom onset, she is currently taking prednisone 5 mg daily without evidence for internal malignancy in radiological and serologic tests.

DISCUSSION

Palmar fasciitis is a rare condition in which the patient develops rapidly progressive contractures of both hands and arthritis of the wrists and larger joints. The underlying immunological mechanisms have not been defined so far and may include the activation of certain factors with proinflammatory activities, transforming growth factor β or connective tissue

Fig. 1. Hands of the patient. It shows the digital flexion contractures and the thickened palmar fascia. The MCP, and PIP joints were tender, and swollen.

Fig. 2. T1WI gadolinium enhanced MRI of the left forearm. It shows diffuse enhancements in the fascia, extensor and flexor tendons and some muscles.

(Fig. 3). The underlying skeletal muscle showed well preserved myocytes, mild interstitial edema, and fibrosis. Marked interstitial infiltration, mainly of T lymphocytes and focal infiltration of B lymphocytes with scattered plasma cells, were observed on immunohistochemical stainings. There was no evidence of perifascicular atrophy, necrosis, granuloma or leukocytoclastic vasculitis.

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growth factor (7). Medsger et al. (1) suggested that a fibroblast proliferative factor might be secreted by the tumor. They were unsuccessful, however, in their attempt to show a proliferative factor in the laboratory. The majority of affected patients have been women, which suggests that the female hormonal state may predispose to this syndrome as is the case in most autoimmune diseases.

Most of reported cases have been suggested associations with neoplasms, most commonly in elderly patients, preceding or accompanying the diagnosis of carcinoma of the ovary. They followed by neoplasms of the breast, pancreas, lung, stomach, and endometrium. Table 1 summarizes the published cases of PFPAS associated with malignancies. The rheumatic symptoms may precede detection of the tumor by 1-23 months. They have confirmed improvement in rheumatic symptoms after chemotherapy but it is unclear whether this is a result of tumor necrosis, immunomodulation, or anti-inflammatory effects.

Some PFPAS patients without apparent neoplasms have been described in previous reports. Seaman et al. (6) reported seven patients undergoing antituberculosis therapy, who developed palmar fasciitis within 4 months of the addition of the drug ethionamide and the fasciitis resolved when ethionamide was discontinued. Until now, only one idiopathic PFPAS of 75-yr-old white woman was reported by Laszlo et al. (5) in 1995. However, they could not exclude the possi-

| Author (Ref.) | Year | Age (yr)/Sex | Associated malignancy | Effect of treatment on manifestations |
|---------------|------|--------------|-----------------------|-------------------------------------|
| Medsger et al. (1) | 1982 | 65/F | Ovary | No improvement |
| | | 50/F | Ovary | No improvement |
| | | 62/F | Ovary | Improved with chemotherapy & CS |
| | | 58/F | Ovary | No improvement |
| Baron (2) | 1982 | 65/F | Bladder, Lung, Colon | Improved with CS |
| Baer et al. (8) | 1983 | 62/F | Pancreas | No improvement |
| Shiel et al. (9) | 1985 | 86/F | Lung | No improvement |
| | | 75/F | Ovary | Improved with CS |
| Pfinsgraff et al. (3) | 1986 | 66/F | CML | Improved with CS |
| | | 59/F | Pancreas | Improved with Penicillamine |
| | | 66/F | Squamous cell carcinoma | No improvement |
| | | 71/F | Adenocarcinoma | No improvement |
| | | 54/M | Hodgkin | Improved with chemotherapy |
| Valverde-Carcia et al. (10) | 1987 | 55/F | Breast | Improved with chemotherapy & CS |
| Caron et al. (11) | 1989 | 74/F | Thyroid plasmacytoma | No improvement |
| Willemsen et al. (12) | 1991 | 54/F | Coelomic epithelial | Improved with CS |
| Van den Bergh et al. (13) | 1991 | 59/M | Prostate | No improvement |
| Strobel et al. (14) | 1992 | 59/F | Ovary | Not mentioned |
| Leslie (15) | 1992 | 55/F | Uterine cervix | No improvement |
| Laszlo et al. (5) | 1995 | 75/F | Idiopathic | Improved with CS |
| Viniker et al. (16) | 1996 | 25/F | Ovary | No improvement |
| Dhole et al. (17) | 1997 | 66/F | Breast | Improved with chemotherapy |
| Saxman et al. (18) | 1997 | 54/F | Breast | Not mentioned |
| Wright et al. (19) | 1997 | 57/F | Ovary | Not mentioned |
| Grados et al. (20) | 1998 | 59/F | Renal pelvis | No improvement |
| | | 92/F | Uterine | No improvement |
| Eekhoff et al. (21) | 1998 | 71/M | Large cell carcinoma | Not mentioned |
| Matteen (22) | 1998 | 69/F | Ovary | No improvement |
| Enomoto et al. (4) | 2000 | 44/M | Stomach | Improved with operation |
| Kase et al. (23) | 2000 | 54/F | SCC of cervix | Improved with chemotherapy |
| Roman et al. (24) | 2001 | 45/M | Hepatocellular carcinoma | Improved with CS |
| Docquier et al. (25) | 2002 | 74/F | Uterine adenocarcinoma | No improvement |
| Virk et al. (26) | 2002 | 69/M | Gastroesophageal carcinoma | Improved with CS |
| Rammeh et al. (27) | 2003 | 37/M | Multiple myeloma | Not mentioned |
| Martorell et al. (28) | 2004 | 62/F | Ovary | Improved with chemotherapy |
| | | 70/F | Ovary | No improvement |
| | | 73/F | Ovary | No improvement |
| | | 51/F | Ovary | No improvement |
| Denschlag et al. (29) | 2004 | 73/F | Fallopian tube | Improved with chemotherapy |
| Giannakopoulos (30) | 2005 | 67/F | Pancreas | Improved with chemotherapy |
| Present case | 2005 | 31/F | Idiopathic | Improved with CS |

CS, Corticosteroid; CML, chronic myelogenous leukemia; SCC, squamous cell cancer.
manifestations of PFPAS are similar to RSD but is characterized by contractures of the hands. The musculoskeletal symptoms with internal malignancy did not respond to steroid treatment. A high dose of corticosteroid, while most PFPAS patients associated with malignancy did not respond to steroid treatment.

The differential diagnosis includes other conditions associated with contractures of the hands. The musculoskeletal manifestations of PFPAS are similar to RSD but is characterized by its considerably more aggressive nature and diffuse articular involvements. Most cases of RSD have a preceding noxious event and symptoms of vasomotor disturbances. Bone scanning of the patient with RSD usually shows an asymmetry between affected and non-affected limbs. Our patient, however, did not show these clinical features. In addition, the pathological finding of our patient was far from that of RSD characterized by muscle changes consisting of type I fiber atrophy, an increase in lipofuscin and capillary abnormalities with thickening of the basement membrane (31).

Dupuytren's contracture is a relatively common condition characterized by nodular thickening and contraction of the palmar fascia. Its manifestation is quite similar with that of PFPAS, but it usually affects the ulnar side of both hands and it is more common in aged men. The findings of our patient differed from the disorder in that the fasciitis was extending to the forearms and apparent polyarthritis was present. Pathologically, the early lesion of Dupuytren's disease is characterized by marked fibroplastic proliferation, vascular hyperplasia and clusters of macrophages and T lymphocytes. This is followed by dense, disorderly collagen deposition with thickening of the palmar fascia and nodule formation (32). These are different from the pathological findings of PFPAS.

Eosinophilic fasciitis is characterized by painful and erythematous swelling of the extremities, accompanied by rapid weight gain, fever and myalgia. Eosinophilia in peripheral blood, and somewhat less commonly in affected tissue, is prominent at the acute stage (33). In this disorder, however, the hands are usually spared. Our patient had no eosinophilia in peripheral blood or eosinophilic infiltration in the tissue.

In summary, we have presented a young Korean patient with idiopathic PFPAS who was successfully treated with administration of corticosteroid. Therefore, we suggest that close search for underlying malignancy and follow ups are essential to PFPAS patient but in a case not associated with malignancy like our case, administration of steroid might be an effective option for treatment.

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