GFFCC Expanded International Cooperation

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### TABLE OF CONTENTS

#### Original Studies

- **Penile Cancer in India: A Clinicoepidemiological Study**
  
  M. Pahwa, M. Girotra, A. Rautela, R. Abraham  
  
  - Page 07

- **Gastric Cancer: A Retrospective Analysis from AIIMS, New Delhi**
  
  R. Hadi, B.K. Mohanti, S. Pathy, G.K. Ragh, N.K. Shukla, S.V.S. Deo, A. Sharma, V. Raina  
  
  - Page 11

#### Review Articles

- **Intensity Modulated Radiotherapy (IMRT) In Head and Neck Cancers – An Overview**
  
  C.M. Nutting  
  
  - Page 17

- **Adult T-Cell Leukemia/Lymphoma**
  
  K.I. Rasul, Z.A. Barwari  
  
  - Page 27

#### Case Reports

- **Adrenocortical Tumors in Children: A Kuwait Experience**
  
  R. Mittal, D. G. Ramadan, N. M. Khalifa, S. O. Khalifa, Z. Mazidi, M. Zaki  
  
  - Page 38

- **Limb Sparing Surgery in Soft Tissue Sarcoma of Extremities: An Indian Perspective**
  
  R.V. Bhargavan, P. Kumar, K.C. Kothari  
  
  - Page 47

- **Mixed Germ Cell Tumor of Ovary and Clitoromegaly in Swyer’s Syndrome: A Case Report**
  
  S. Aminimoghaddam, B. Mokri, F. Mahmoodzadeh  
  
  - Page 55

- **Palmar Fasciitis and Arthritis Syndrome Associated With Metastatic Ovarian Cancer: A Paraneoplastic Syndrome**
  
  I.K. Nahar and M. S. Al-Rajhi  
  
  - Page 59

- **Trichilemmal Pilar Tumor of the Scalp: A Case Report**
  
  K. Al Saleh, H.S. Hooda, H. El-Wakiel, R. Safwat, A. Bedair, W. Eskaf  
  
  - Page 62

- **Carcinosarcoma of Renal Pelvis with Immunohistochemical Correlation**
  
  S.D. Deshmukh, V.L. Gaopande, D.P. Pande, G.S. Pathak, B.K. Kulkarni  
  
  - Page 65

- **5-Flourouracil Cardiotoxicity – An Elusive Cardiopathy: Case Report**
  
  G. M. Bhat, M. H. Mir, H. I. Showkat, B. Kasanna, F. Bagdadi, A. H. Sarmast, S. Qadri  
  
  - Page 70

- **An Unusual Variant of Prostatic Adenocarcinoma with Metastasis to Testis: A Case Report**
  
  K. R. Anilta, T. Somanathan, A. Mathews, K. Jayasree  
  
  - Page 73

- **Mammary Fibromatosis in a Male Breast**
  
  N. Al-Saleh, T. Amir, I. N. Shaf  
  
  - Page 77

- **Primary Isolated Extramedullary Plasmacytoma of Mesentry: A Rare Case Report**
  
  R. Galhotra, K. Saggar, K. Gupta, P. Singh  
  
  - Page 81

#### Feature Article

- **Balsam Organization for rehabilitation and support for cancer patients and their families**
  
  - Page 85

#### Conference Highlights /Scientific Contribution

- **Conference Highlights – 1st Palliative Care Conference in Kuwait**
  
  - Page 86

- **News Notes**
  
  - Page 89

- **Advertisements**
  
  - Page 91

- **Scientific events in the GCC and the Arab World for the 2nd Semester of 2012**
  
  - Page 92
Abstract

Palmar fasciitis and polyarthritis syndrome (PFPA) is an uncommon syndrome that affects predominantly elderly women and characterized by symmetrical polyarthritis followed by flexion contracture of the hands. It is usually associated with a metastatic malignant neoplasm, and therefore implies a poor prognosis. We report a case of a 54-year old woman presented with palmar fasciitis and polyarthritis six months before the diagnosis of a metastatic adenocarcinoma of the ovary. Surgical excision of the tumor and adjuvant chemotherapy caused remission of the polyarthritis.

Keywords
Palmar fasciitis and arthritis, paraneoplastic syndrome, polyarthritis ovarian cancer

Case report

A 54-year-old housewife and a mother of five adults presented to the rheumatology outpatient clinic with five months history of bilateral progressive pain of the small joints of the hands associated with swelling and prolonged early morning stiffness. She had no constitutional symptoms. Her past medical history includes hypertension for seven years and hyperlipidemia for nine years. She had no psoriasis nor did any of her family members and had not used oral contraceptive agents. Her system review revealed no history of Raynaud’s phenomenon or dysphagia. She is a non-smoker. Physical examination revealed tenderness and swelling of her left 2nd, 5th and right 1st, 2nd, 3rd and 5th metacarpophalangeal joints, right 1st Interphalangeal joint, left 2nd, 3rd and right 2nd and 5th proximal interphalangeal (PIP) joints. She had sclerodactyly-like skin thickening of all fingers with flexion deformities, and positive prayer’s sign. Her immunological screen revealed a negative antinuclear antibody, a negative rheumatoid factor, and a normal C-reactive protein level. Radiography of the hands showed asymmetric erosions over DIPs and PIPs with no juxtaarticular osteoporosis. One month later, she developed progressive flexion contracture of the fingers and had experienced an abdominal discomfort and a pelvic mass was felt on gynecological examination. She underwent an exploratory laparotomy with total hysterectomy and bilateral salpingo-oophorectomy. Microscopy of the right and left ovaries revealed papillary serous adenocarcinoma (Figure 1). Staging of the tumor showed grade III which implies peritoneal metastasis outside the pelvis with or without retroperitoneal lymph nodes, and with or without superficial liver metastases. Perioperatively, the patient had received cycles...
of adjuvant chemotherapy with paclitaxel and carboplatin. Six months later, her pain and swelling had gradually resolved with some loosening of skin over fingers, but flexion contractures have persisted. A repeat radiological examination of hand showed progression of articular erosions and joint space narrowing (Figure 2).

**Discussion**

We presented a case of a middle-aged woman who developed a paraneoplastic syndrome, in the form of erosive polyarthritis with flexion contractures six months before a malignant ovarian tumor was discovered. The overall clinical presentation in this case is consistent with the syndrome of palmar fasciitis and polyarthritis (PFPA).

Palmar fasciitis and polyarthritis syndrome (PFPA) is an uncommon syndrome, which primarily affects elderly women. It was first described by Medsger and colleagues in 1982 (1) in six patients with metastatic ovarian neoplasm. PFPA had also been reported in association with other types of neoplasm including chronic lymphocytic leukemia, pancreatic adenocarcinoma, squamous cell carcinoma of the lung, chondrosarcoma, and Hodgkin’s disease(2,3). The syndrome generally presents with morning stiffness and arthralgia of multiple joints, mostly the metacarpophalangeal and interphalangeal joints. Prominent palmar changes include local warmth, erythema, and diffuse swelling. It may occur concomitantly or precedes the diagnosis of a malignant neoplasm by several months(4). The laboratory results are usually unremarkable with the absence of an acute phase response. X-ray of the joints generally shows no abnormalities (2). The pathophysiological mechanism of the PFPA syndrome is unknown(5). Autoimmune etiology had been suggested by the observation of deposits of immunoglobulin in the fascial tissue of affected patients(4). It had also been suggested that female hormonal state may predisposes to this syndrome, given its predominance in women. Indeed, serum estrogen level was found to be increased in several cases of PFPA(1, 2). Treatment with steroids, non-steroidal anti-inflammatory drugs, or hand therapy has little effect, whereas successful treatment of the underlying tumor can relieve the symptoms (2, 4).

The clinical manifestations of PFPA syndrome often appear before the tumor is clinically evident, which leads to poor prognosis since the malignant tumor often is advanced (6). The differential diagnosis for PFPA syndrome includes rheumatoid arthritis, Dupuytren’s contracture, scleroderma, eosinophilic fasciitis, and reflex sympathetic dystrophy(2).

Paraneoplastic rheumatic disorders are those cancer-associated rheumatic syndromes that occur at a distance from the primary tumor or metastases and are induced by the cancer through hormones, immunoglobulins or other humoral mediators(5). Other rheumatic disorders which are associated with cancer include pulmonary hypertrophic osteoarthropathy, polymyalgia rheumatica, rheumatoid-like arthritis, scleroderma, dermatomyositis and polymyositis, relapsing polychondritis and antiphospholipid syndrome(7). A thorough history and physical examination should be undertaken in all patients presenting with a rheumatic syndrome associated with neoplasm or with progressive and unusual musculoskeletal manifestations. If a clue to occult malignancy is present then an extensive search for the presence of malignancy should be undertaken.
Recognizing a paraneoplastic syndrome is important for many considerations. First, it simply represents the first clue to an existing neoplasm and therefore, it may be detected at an earlier stage where intervention is possible. Secondly, detecting a neoplasm at an early stage may also improve or cure the paraneoplastic rheumatic disease and may therefore, limit or avoid associated disabilities.

Our patient fits into the literature description of the syndrome. She presented with polyarthritis and later developed swelling of the palm and fingers with skin thickening and rapidly progressing flexion contractures of the hands. This had closely preceded the diagnosis of a metastatic ovarian carcinoma. The clue to diagnosis in this patient was the development of progressive flexion contractures. Although her skin thickening and presence of DIP joint erosions might suggest a different diagnosis such as scleroderma and psoriatic arthritis, respectively. The absence of Raynaud’s phenomenon and the lack of psoriatic patches do not favor these diagnoses. Our case is unique in that the patient developed progressive polyarthritis with joint erosions and joint space narrowing over relatively a short period of disease onset. Erosive arthritis in association with PFPA has not been described in the literature. On the other hand, erosive osteoarthritis, polyarthritis, and polyarthralgia had been reported following chemotherapy for ovarian cancer, particularly bevacizumab, although our patient did not take this drug\(^8\). In our case, a pelvic mass was recognized during pelvic examination which stresses the importance of a full examination for patients with progressive or unusual rheumatic symptoms. Following resection of the tumor and appropriate chemotherapy, our patient’s symptoms on polyarthritis, skin thickening and fingers flexion function were reversed. However, her hand contractures persisted.

Our case emphasizes the importance of recognizing palmar fasciitis and polyarthritis syndrome for the early diagnosis and treatment of an occult ovarian carcinoma. We recommend a thorough neoplastic work-up including gynecological examination for any woman presenting with a sudden onset of unexplained hand pain, inflammatory fasciitis with or without digital contractures.

References