Case Report

Paraneoplastic Palmar Fasciitis and Polyarthritis Syndrome

G Santra

Abstract

Palmar fasciitis and polyarthritis syndrome is a rare paraneoplastic syndrome that has been reported mainly for ovarian cancers. It is thought to be a tumour-associated autoimmune disorder. It is associated with polyarthritis and rapid flexion contractures of hands with palmar nodules due to palmar fasciitis. Similar paraneoplastic features in gastric cancers have seldom been reported. Here, I am reporting a case of paraneoplastic syndrome of metastatic gastric adenocarcinoma with polyarthritis of hands and contractures of multiple joints with asymmetric clubbing (right hand more than left hand) and hyperpigmentation over back of hands and fingers. Hyperpigmentation and asymmetric clubbing have not been reported in literature in palmar fasciitis and polyarthritis syndrome.

INTRODUCTION

Palmar fasciitis and polyarthritis syndrome is a rare paraneoplastic syndrome. It has been reported mainly for ovarian cancers. Similar paraneoplastic features in gastric cancers have seldom been reported. Here, I am reporting a case of palmar fasciitis and polyarthritis syndrome due to metastatic gastric adenocarcinoma.

CASE REPORT

A 37-years old housewife presented with cachexia and recurrent projectile vomiting of one month duration. Vomiting followed immediately after taking food or water. It contained old food particles. She also complained of something moving in the abdomen with colicky abdominal pain for same duration. She was constipated. There was one episode of melaena three weeks back. One and half months back she was admitted to the hospital with anorexia and weakness and received four units of blood transfusion.

On enquiry, she gave six months history of pain and swelling of small joints of hands, followed by development of rapidly progressive flexion contractures of right hand and to a lesser extent of the left hand (Figs. 1 & 2). There was gradual blackening of dorsal of hands and fingers. The patient had restricted movements and pain in the left knee. She had limitation of abduction of both arms and restricted movements in both elbow joints due to contractures. Her proximal and distal interphalangeal (PIP and DIP) and metacarpophalangeal (MCP) joints were flexed with limitation of movements. Flexion contractures were more on right PIP joints (~90°). She had palpable thickening of palmar fascia with nodules that were present bilaterally but more on right side. She had higher degree of clubbing of right hand (2°) than the left (1°) and hyperpigmentation over back of hands and fingers. Her hands were mildly edematous with no evidence of telangiectasia, digital pitting or systemic sclerosis. Sensory examination and sweat pattern were normal in both hands. She denied any history of Raynaud's phenomenon or trauma to the hands and had no history of alcohol or phenytoin intake. She was afebrile and had no history of contact with tuberculosis or exposure to STDs.

The patient was cachectic with severe pallor and bilateral pedal oedema. Pulse was 88/minute, regular. Blood pressure was 116/78 mmHG. Respiration was normal. Jaundice, cyanosis and lymphadenopathies were absent. Neck vein was not engorged. Abdomen was soft. Flanks were full. Abdominal tenderness was not present. Fluid thrill was absent but shifting dullness was positive. She had an ill-defined lump in epigastric region. Liver was palpable and firm with irregular surface. Breast, thyroid and pelvis examinations were normal. Respiratory, cardiovascular and neurological systems were normal.

Her complete blood counts revealed severe microcytic hypochromic anaemia (Hb 5 gm%). eSR was 110mm/1st hour. Total and differential white cell counts were normal. Her blood sugar, urea and creatinine levels were normal. LFT was normal except raised ALP level (480 U/L). Rheumatoid factor and ANF were negative. T3, T4 and TSH levels were normal. Stool for occult blood was positive. Upper GI endoscopy showed deformed distal part of antrum with a mass and grossly narrowed pyloric channel. X-ray of hands showed periarticular osteopenia without any erosions.
USG abdomen showed multiple secondaries in liver and intraabdominal lymph nodes. Moderate ascites was present. Gall bladder had multiple calculi but no evidence of malignancy. Ovaries were normal. Gastric mucosal biopsy was noncontributory due to sampling error. A repeat biopsy was not possible. FNAC from a liver lesion suggested metastatic gastric adenocarcinoma (Figs. 3 & 4). A diagnosis of paraneoplastic palmar fasciitis and polyarthritis syndrome due to metastatic gastric adenocarcinoma was made.

**DISCUSSION**

Musculoskeletal problems associated with malignancy include i) arthropathies, ii) muscular disorders including dermatomyositis and polymyositis, iii) scleroderma, panniculitis and fasciitis, iv) vasculitides, and v) miscellaneous rheumatic syndromes. Paraneoplastic rheumatic syndromes can provide the hints for earlier diagnosis of occult malignancy.1

Palmar fasciitis and polyarthritis syndrome (PFPAS) is an uncommon paraneoplastic musculoskeletal presentation, first described in 1982 by Medsger et al2 in six patients with ovarian carcinoma. More than 40 cases have since been reported, mainly ovarian cancers, followed by lung, pancreas, colon, cervix and hematolymphatic malignancies. The syndrome usually develops in patients with advanced or metastatic cancers. Idiopathic or antituberculosis drug (ethionamide) induced PFPAS has also been reported. PFPAS usually develops in females aged over 55 years. Enomoto et al described a male patient of advanced gastric cancer with PFPAS.3 Carli et al also reported a case remarkably similar to PFPAS in a 76-year-old man, subsequently diagnosed to have gastric carcinoma with colonic metastasis. After resection of tumours, finger and thumb contractures and pain were lessened.4

PFPAS may have an association with autoimmune reactions as suggested by deposits of IgG in palmar fascia and presence of low titer antinuclear antibodies or elevated serum levels of soluble interleukin-2 receptor (SIL-2R) in some patients. Histologic examination of involved tissues reveals extensive fibrosis with increased fibroblast and mononuclear cell infiltration. There is no evidence of
collagen deposition like that seen in scleroderma.

PFPAS is characterized by polyarthritis and rapid flexion contractures of hands with palmar nodules due to palmar fasciitis. The MCP and PIP joints are most commonly affected, but other affected joints include the elbows, wrists, knees, ankles and feet. PFPAS is associated with nonerosive form of arthropathy with periarticular osteopenia. Planter fasciitis can occur with lower extremity involvement. Carpal tunnel syndrome is a rare association.

The differential diagnosis of PFPAS includes scleroderma, complex regional pain syndrome (CRPS), Dupuytren's contracture, eosinophilic fasciitis and shoulder-hand syndrome. The absence of Raynaud's phenomenon, normal nail fold capillary examination, lack of specific autoantibodies and rapid progression of clinical features help to exclude it from scleroderma. Although initially thought to be an atypical variant of CRPS, the more aggressive course, bilateral presentation and strong association with malignancy suggest PFPAS as a distinct entity. Dupuytren's contracture is characterized by nodular thickening and contraction of the palmar fascia, usually affecting the ulnar side of both hands. The arthritis and synovitis and rapid progression of clinical features makes Dupuytren's contracture an unlikely diagnosis. In my patient contractures involved other joints also. Eosinophilic fasciitis is characterized by painful erythematous swelling of the extremities, accompanied by rapid weight gain, fever and myalgia. Eosinophilia in peripheral blood and less commonly in the affected tissue is prominent in acute stage and hands and fingers are usually spared. The shoulder-hand syndrome, a variant of CRPS, is much milder than PFPAS. This syndrome is most often described with ovarian cancer or with lung cancer localized to superior sulcus (Pancoast tumour) infiltrating the stellate ganglion or brachial plexus. Pain in the shoulder with loss of motion may result in adhesive capsulitis, and the hand of the involved side becomes puffy and stiff with vasomotor instability.

The presence of PFPAS portends a poor prognosis as it typically manifests after tumour metastasis. The response to NSAIDs, corticosteroids and physical therapy is variable. Successful removal of the underlying tumour may result in dramatic clinical improvement of the affected extremities.

In the present case, finger and multiple other joint contractures with preceding history of polyarthritis of hands were present. Other features included higher degree of clubbing of right hand (2°) than the left (1°) and hyperpigmentation over back of hands and fingers. Asymmetric clubbing and hyperpigmentation have not been reported in literature in PFPAS. Paraneoplastic cutaneous manifestations may rarely include pigmentary disorders including diffuse melanotic macules and papules with visceral adenocarcinoma.

**References**


**Announcement**

6th Annual Conference of Indian Thyroid Society – ITSCON 2009 will be held on 7 & 8th March 2009 at Hyderabad.

For registration & Abstract Submission please contact : Dr. Rakesh Sahay, Organizing Secretary ITSCON 2009, I/c Professor of Endocrinology, Department of Endocrinology, 2nd Floor, Golden Jubilee Block, Osmania General Hospital, Afzalgunj, Hyderabad.

Phone +91-9849597507, itscon2009@gmail.com, sahayrk@gmail.com