Arthritis and Hypergammaglobulinemic Purpura in Hypersensitivity Pneumonitis

Hendrika Maria Oosterkamp, MD, Hans van der Pijl, MD, Jan Derksen, MD, PhD, Luuk N.A. Willems, MD, PhD, Paul H.E.M de Meijer, MD, PhD, Leiden University Hospital, Leiden, the Netherlands

In hypersensitivity pneumonitis or extrinsic allergic alveolitis, both systemic and respiratory symptoms may occur. Two patients are reported with systemic manifestations of this disease that have not been described previously.

CASE REPORTS

Case I

A 51-year-old man was admitted because of dyspnea, thoracic pain, dry cough, and weight loss of 1 month duration. He had been holding parakeets for 40 years. Physical examination revealed dorsalbasal pulmonary rales, palpable purpura on the legs, and swelling of both knees and the right ankle.

Besides an erythrocyte sedimentation rate (ESR) of 80 mm/h, standard laboratory tests were normal. The serum protein concentration was elevated (85 g/L) due to hypergammaglobulinemia with raised immunoglobulin A (IgA) (13.3 g/L) and IgG (44.9 g/L). There were circulating immune complexes and complement activation. Antinuclear antibody and rheumatoid factor were negative. The arterial blood gas was unremarkable.

Chest roentgenogram demonstrated fine reticulonodular shadows throughout both lungs. Antibodies to parakeet antigen were strongly positive. Pulmonary function tests revealed decreased volumes and decreased diffusion capacity. The bronchoalveolar lavage (BAL) fluid showed 60% lymphocytes with a CD4/CD8 ratio of 0.4. In the transbronchial biopsies, interstitial pneumonitis was seen and biopsies of the purpura revealed a leukocytoclastic vasculitis.

The diagnosis of bird-breeder's lung was made. After cessation of the exposure and initiation of prednisone therapy, clinical improvement was noted.

DISCUSSION

Besides the well-known features of hypersensitivity pneumonitis, the 2 patients reported here had additional symptoms. Both patients exhibited hypergammaglobulinemia and purpura on the legs, while the first patient also exhibited a perivascular infiltrate and developed oligoarthritis and the second patient also exhibited a leukocytoclastic vasculitis.

Although hypergammaglobulinemia is a well-known feature of hypersensitivity pneumonitis, the development of purpura has not been mentioned before. Waldenstroem described a syndrome consisting of recurrent purpura, hypergammaglobulinemia, elevated ESR, and mild anemia. The purpura seen in hypergammaglobulinemia are petechial, scattered, and usually on the legs and are brought on by standing, walking, tight-fitting garments, and heat. The purpura are often accompanied by arthralgia, low-grade fever, and edema of the lower extremities.

Hypergammaglobulinemic purpura (HGP) is divided in a primary type and a secondary type associated with an underlying disease. Laboratory abnormalities in HGP are elevated gamma globulins, elevated ESR, and mild anemia. Rheumatoid factor, antinuclear antibody, or cryoglobulinemia may be present. Inflammation of the superficial dermal blood vessels, with a polymorphonuclear leukocyte infiltrate in the acute phase, followed by a mononuclear infiltrate is found. Red blood cell extravasation, leukocytoclasia, and blood vessel damage have been described—from arteriolar necrosis to fibrinoid degeneration.

Olmstead et al found IgM and C3 deposition in blood vessel walls and elevated levels of IgM and IgG immune complexes, which supports the concept that HGP is mediated by immune complexes. Because of the clinical and laboratory abnormalities in our patients, we think the pathogenesis of the purpura is identical to that described for HGP.

Several concomitant phenomena (eg, arthralgia) have been reported in hypersensitivity pneumonitis.
A recent case report mentions myositis and arthritis in 1 patient with this disease. In our opinion, however, the clinical features described do not justify the diagnosis of arthritis in this patient.

To our knowledge, our patient, therefore, is the first described with frank arthritis. The immunologic process in the pathogenesis of HGP may also be the explanation for the development of arthritis in hyper-sensitivity pneumonitis. We suggest HGP and arthritis in our patients resulted from an immunologic process in which hypergammaglobulinemia and high levels of circulating immune complexes lead to deposition of immune complexes and complement with subsequent inflammation in skin vessels and joints.

REFERENCES

Manuscript submitted August 17, 1995 and accepted in revised form October 31, 1995.

Septic Olecranon Bursitis in Patients With Chronic Obstructive Pulmonary Disease

Raymond J. Enzenauer, MD, LTC, MC, Jerry L. Pluss, DO, FCCP, LTC, MC, Fitzsimons Army Medical Center, Aurora, Colorado

Septic olecranon bursitis is seen in patients with frequent elbow trauma, especially middle-aged men involved in manual labor. While immunocompromised patients with alcoholism or diabetes may be at increased risk for septic olecranon bursitis, large series do not include chronic obstructive pulmonary disease (COPD) as a risk factor for disease.

We report a 74-year-old woman with severe COPD and septic olecranon bursitis. The forward leaning posture in patients with severe COPD may be a risk factor predisposing to olecranon bursal infection.

CASE REPORT
A 74-year-old woman with severe oxygen-dependent COPD was admitted with 1 week of increasing pain, swelling, and warmth of the left forearm. Erythema spread proximally to the olecranon bursa. The patient stated that she had been increasingly leaning on her forearms and elbows to provide ventilatory support. Medications included prednisone (10 to 20 mg daily), oral theophylline, and three mini-dose inhalers.

Her temperature was 98.6°F. Extremity examination revealed warm, erythema, and swelling of the left mid-forearm to the elbow. The olecranon bursa was distended, warm, and tender, with no visible skin wound or laceration. Complete blood count was remarkable for an elevated white blood cell (WBC) count of 18.2 x 10^9/L with 88% polymorphonuclear leukocytes. Left olecranon bursa aspiration revealed a bursal fluid WBC of 11.4 x 10^9/L. Bursal fluid Gram's stain showed rare intracellular gram-positive cocci with culture positive for Staphylococcus aureus. Treatment with intravenous nafcillin sodium (Unipen, Wyeth-Ayerst Laboratories, Philadelphia, Pennsylvania) resulted in prompt symptom improvement.

Patients with severe COPD frequently lean forward, bracing themselves on their forearms. A forward, "sustained posture increases the capacity for sustained hyperventilation. Such a posture, with its concomitant sustained pressure over the olecranon bursae, could also predispose patients with severe COPD to septic olecranon bursitis.

DISCUSSION
The case presented is clearly atypical demographically. Septic bursitis is a disease of middle-aged manual laborers. Mean age is 47 years, with up to 100% of patients being men. Typically patients with severe COPD are older, with no significant male predominance. Other factors, such as uremia, diabetes, and alcoholism may increase host susceptibility to bursal infection. Corticosteroid therapy may increase the risk of septic olecranon bursitis in patients without COPD. Accidental trauma, be it direct, repetitive minor trauma or constant pressure, appears to be the common denominator.

Hemorrhagic olecranon bursitis, or "dialysis elbow" has been reported in uremic patients as a result of prolonged pressure against the olecranon process during dialysis. Local or distant skin disruption may not be present in 42%. Even when visible skin wounds are not apparent, the avenue of bacterial introduction is presumed to be through the skin. A high index of suspicion should prompt bursal aspiration when the diagnosis of septic olecranon bursitis is entertained. Bursal fluid WBC counts in septic bursitis are lower than those in septic arthritis. Bacteria may be seen on Gram's stain smears in only 65% of culture-positive bursal fluids. S aureus is the infecting organism in the majority of cases.