ASSOCIATION OF PYODERMA GANGRENOSUM WITH RHEUMATOID ARTHRITIS

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A 30-year-old female presented with recurrent leg ulcers of 3-year duration. History revealed that she also suffers from pain and stiffness in the small joints of hands and feet for the past 4 years. Rheumatoid factor was strongly positive in the serum. Association of the two is interesting.

Key words : Pyoderma gangrenosum, Rheumatoid arthritis, Association.

Pyoderma gangrenosum is a rare skin disorder of unknown etiology occurring mainly in patients with ulcerative colitis,[1] granulomatous colitis,[2] diverticulitis,[2] chronic active hepatitis,[3] leukemia,[4] paraproteinemias,[5] and rheumatoid arthritis.[2,3,6] Reports of pyoderma gangrenosum in rheumatoid arthritis are very rare.[6] We report such an association.

Case Report

A 30-year-old female was developing recurrent painful leg ulcers for 3-years. History revealed that the ulcers would begin as tiny red lesions which would rapidly change into bullae, full of pus. These bullae would soon rupture and form an acutely painful ulcer with marked central necrosis. The ulcers would rapidly increase in size by involving the adjacent normal skin and soon a progressively spreading large area of skin ulceration would be formed having a central purulent region and an inflamed border of bluish red skin. Healing in the ulcers would begin slowly from the edge and be complete in 6-8 weeks time leaving a thin atrophic scar. The patient had 6 such episodes in the past. All were in the winter season and all lasted on an average 2-3 months. The present ulcer at the time of reporting was of three weeks duration. A biopsy from the margin of the ulcer resulted in a large area of leg ulceration which healed in eight weeks time. During the past four years, the patient had also suffered from pain and stiffness in the small joints of the hands and feet which was maximum in the morning and disappeared in the evening.

Examination revealed a single, ragged, deep ulcer 10×8 cm in size on the right lower leg having purulent necrotic floor and an irregular boggy border with a halo of bluish red skin (Fig. 1). The ulcer and the surrounding skin were extremely tender. Multiple, atrophic, thin scars of varying sizes due to the past leg ulcerations were present on the front and back of both lower legs. Joint examination showed tenderness and fusiform swelling of metacar-

Fig. 1. Pyoderma gangrenosum.

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pophalyngeal and proximal interphalyngeal joints of both hands. Deformity or deviation of joints was not seen. Systemic examination was normal.

Routine investigations of urine and stools were normal and blood showed a polymorphonuclear leucocytosis and raised ESR. Levels of immunoglobulins and complement were normal and tests of ANF, LE cell and cryoglobulin were negative in the sera. Rheumatoid factor was positive in high titre. Pus culture grew a heavy growth of *Staphylococcus aureus*. Skin biopsy showed non-specific inflammatory changes around the blood vessels in the upper dermis with a dense infiltration of polymorphs, lymphocytes and plasma cells.

A 2-week course of dapsone 100 mg/day was given with no benefit. Systemic antibiotics did not also bring any improvement. Steroids in 40 mg daily dosage brought rapid relief in symptoms and the dose was withdrawn gradually in 6-weeks time.

Comments

Fifty per cent cases of pyoderma gangrenosum are associated with an underlying systemic disease.[5] In our case it was found to be rheumatoid arthritis. Other causes of leg ulcer and polyarthritis were carefully ruled out by detailed clinical and laboratory investigations. Its association in rheumatoid arthritis is rare and to the best of our knowledge is being reported for the first time in India.

No other skin manifestation of rheumatoid arthritis was seen. The cause of pyoderma gangrenosum in rheumatoid arthritis is unknown. It is considered by some to be a type of Schwartzman reaction—a non-immunological mechanism, though others propose an immunological injury mechanism.[6] No abnormality of immunoglobulins or complement levels were detected in our case.

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