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ACNE FULMINANS ASSOCIATED WITH REACTIVE POLYARTHRITIS: REPORT OF A CASE AND REVIEW OF THE LITERATURE

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ABSTRACT

We describe a 16 year old boy with acne fulminans associated with axial and peripheral polyarthritis. The patient's clinical course and therapy with isotretinoin, prednisolone and oxytetracycline are described. A possible association between the presence of HLA-B27 antigen and reactive arthritis with acne fulminans in this case is evaluated. A review of the literature is included.

Keywords: Acne fulminans, reactive arthritis, HLA-B27 antigen

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INTRODUCTION

Acne fulminans is an uncommon acute type of acne, associated with fever, weight loss and papulopustular lesions that are highly inflamed, tender and eventually ulcerative. These lesions occur on the face, chest, back and upper extremities. This devastating type of acne is one of the most scarring dermatalogic disorders and occurs almost exclusively in boys.[1,2] In 1975, Plowing and Kligman clearly separated this disease from acne conglobata and coined the term acne fulminans.[3] Musculoskeletal abnormalities including arthralgias and arthritis may occur in association with ance fulminans. In Stathan et al's study symptoms of joint pain occurred in twenty-three out of thirty-two cases reviewed. Nevertheless, reports of associated arthritis were very rare. Destructive arthritis is reported by Hunter et al. as a case report.[6] Hault et al. reported a case with acne fulminans and systemic manifestations and musculoskeletal pain associated with osteolytic lesions.[7] Piazza and Giuntareported a case with lytic bone lesions and polyarthritis associated with acne fulminans.[8] Leukocytosis, anemia and an elevated erythrocyte sedimentation rate (ESR) are typical laboratory findings in acne fulminans.[9] To our knowledge so far 55 cases have been reported. Herein we report a 16 year old boy with acne fulminans and with reactive polyarthritis involving the right knee, left sacroiliac and left metatarsophalangeal (MTP) joints associated with severe tenderness of the left costochondral junctions. The patient was treated with diclofenac and promptly recovered. Acne fulminans was effectively treated with isotretinoin, prednisolone and oxytetracycline.

CASE REPORT

A 16 year old white male presented with mild facial acne. 11 months later these lesions flared and new ones appeared on his face, neck, shoulder, chest and back. One month afterwards he developed recurrent chills, fever and axial skeletal pain with diffuse arthralgias, most marked in the right knee. Upon admission his physical examination revealed the following: Temperature 38.9°C, pulse rate 90 beats per minute, respiration rate 20 per minute, blood pressure 130/70 mmHg, markedly inflamed, tender pustules and ulcers were present on the face, neck, shoulders, chest and back (Fig. 1). The nodulocystic and ulcerative lesions were deep and most were extremely tender (Fig. 2). Tenderness was so severe at the left costochondral junctions that full respiratory movements were limited. The right knee joint was swollen and tender and showed slight flexion contracture and effusion. There was tenderness on the left sacroiliac joint. The left metatarsophalangeal (MTP) joints were tender and swollen. There was tenderness on the right ischial tuberosity. Except for minor cervical adenopathy,

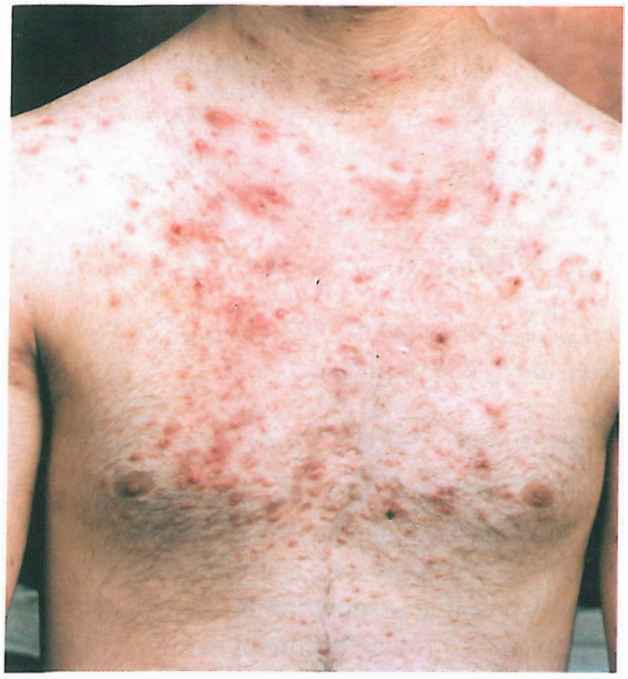

Fig. 1. Lesions of acne fulminans on the chest.

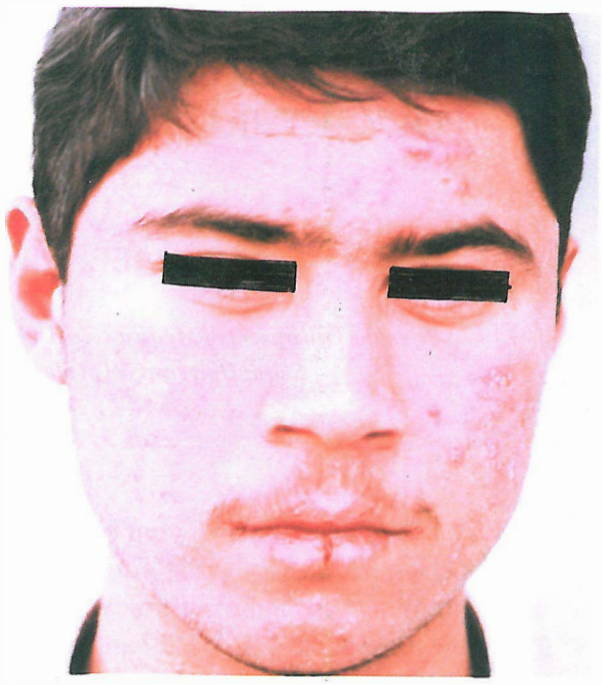

Fig. 2. Appearance of acne fulminans lesions after treatment.

the general physical examination was otherwise normal. Laboratory investigations showed a white blood count of 17300/mm³, with 78% polymorphonuclear neutrophils, 17% lymphocytes, 3% monocytes and 2% eosinophils. The platelet count was 820,000/mm³, and the erythrocyte sedimentation rate (ESR) was 76 mm/hour (Westergren). C-reactive protein was positive, and antinuclear antibody was positive with a titer of 1/80 and a speckled pattern. HLA typing revealed the presence of B27 antigen. Urinalysis showed 1-2 red blood cells per high power field. Total blood protein was 8 gm%, with an albumin level of 4.6 gm%. The serum calcium was 9.2 mg/dl, and phosphorus was 5 mg/dl. Blood urea nitrogen was 11.5 mg/dl with a serum creatinine of 0.5 mg/dl. HBsAg and HIV were negative. Liver function tests were normal. The C_3 level was 1.04 g/L and the C_4 level was 0.4 gm/L (normal range in our laboratory). Anti-streptolysin O was 166 and febrile agglutination tests were negative for brucella and typhoid fever. Specimen cultures from skin lesions were negative and blood cultures were sterile. Radiographs of the right knee showed soft tissue swelling and evidence of effusion. Sacroiliac joint radiographs were nomal. The chest X-ray was also normal. Treatment with isotretinoin, prednisolone and oxytetracycline was started and proved to be effective.

DISCUSSION

Acne fulminans is an acute febrile illness with extensive ulcerating and inflammatory lesions affecting the back, chest and face. The healed lesions show considerable granulation tissue.[8] Weight loss is a prominent feature of this disease. Laboratory abnormalities include anemia, leukocytosis and high ESR. ANA and rheumatoid factor are negative. The incidence of HLA-B27 antigen does not seem to be significantly increased. Males are much more frequently affected than females. Piazza et al. mentioned that all patients are young males with an average age of 15.[8] The pathogenesis of this disorder is unknown. Skin and blood cultures are inconclusive.[10] Hypersensitivity is speculated,[11] as decreased delayed hypersensivity responses have been reported.[12] Some authors postulate an immune complex mechanism and decreased serum complement levels.[8,13,14,15,16] Others have been unable to confirm these observations. Acne fulminans can be complicated by a systemic inflammatory arthropathy.[8] In the reported cases by Davis et al, the majority had arthralgia of large joints i.e, hips, knees and shoulders.[10] Objective arthritis was demonstrated in the sacroiliac joints, hips, knees and ankles.[10] In our patient, examination disclosed active synovitis involving the right knee joint, left metatarsophalangeal (MTP) joints and tenderness of the left sacroiliac and left costochondral junctions. The arthropathy associated with acne fulminans is believed to be slight and characterized by normal radiological findings.[16] In our patient X-ray of the left foot and right knee joints demonstrated soft tissue swelling and effusion. In Ellis et al's study in five out of six patients histocompatibility antigen HLA-B27 was absent.[16] In Davis et al's study radiological evidence of sacroiliitis was found in one patient who did not possess HLA-B27.[10] In our case with signs of

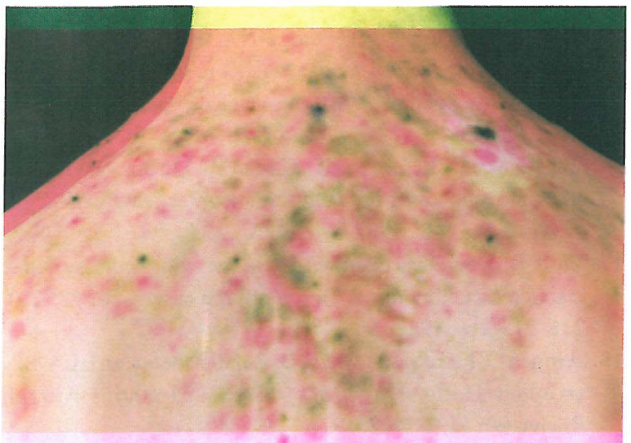

Fig. 3. Appearance of skin 12 months after treatment. Widespread residual keloids remain on the upper part of the back.

sacroiliitis, HLA-B27 was present without any radiological evidence of sacroiliitis. The presence of B27 antigen could not be fortuitous in our patient, especially since clinical signs of sacroilitis were present. On the other hand the question of seronegative spondyloarthropathy was not a viable one because of the dramatic response of acute polyarthritis to diclofenac without recurrence during the course of his acne fulminans. Reactive arthritis has been described in patients with nonarticular infections including *Yersinia entercolitica*,[18] and Reiter's syndrome after nongonococcal urethritis,[19] and there is also evidence to suggest that enteric infection may be involved in the pathogenesis of ankylosing spondylitis.[20] In these disorders, polyarthritis follows infection, but is often associated with the presence of HLA-B27 antigen in the affected individual. Several patients are reported with exacerbations of arthritis associated with activity and flare up of skin lesions. The presence of HLA-B27 antigen in our patient supports the possibility of an association, as reactive polyarthritis developed one month after the skin lesions. Conversely, improvement of polyarthritis followed therapy for skin lesions. Decreased serum complement levels is postulated by some authors.[13] In our patient C_3 and C_4 levels were in the normal range. ANA is reported as negative in some of the reported cases.[8,10] The significance of positive antinuclear antibody (ANA) at a titer of 1/80 in our case is uncertain. Thrombocytosis has been previously reported in two cases.[9,17] In our patient the platelet count was 820,000/mm^3. Antibiotic treatment alone is usually ineffective in acne fulminans. A favorable response, with the regression of systemic symptoms and cutaneous lesions was seen with the administration of adrenal steroids (prednisolone 15-50 mg daily) for systemic effects, combined with debridement of the ulcerations and systemic antibiotics. Our patient was initially treated with 60 mg prednisolone daily, and the dose was tapered to 7.5 mg over a 4-6 week period, plus one gm/day oxytetracycline. Diclofenac was administered as 75 mg daily for a short course. As an out-patient he continued to improve and after 2 months showed improvement of arthritis and progressive healing of skin lesions after 20 months. The patient's platelet count, hemoglobin level, ESR and white blood count returned to normal, but skin lesions recurred six months later, along with right knee joint arthritis. A daily regimen of prednisolone, 40 mg, isotretinoin 80 mg and oxytetracycline 1gm was started. Prednisolone therapy was tapered to 7.5 mg daily and continued for 3 months. Isotretinoin was reduced to 40 mg daily after 2 months and discontinued 2 months later. The skin lesions improved dramatically at the end of 12 months (Fig. 2). 12 months later, the skin remained clear, but residual keloids were present on the site of skin lesions (Fig. 3). Occasional bilateral knee joint arthralgia associated with vague low back pain is still present.

To summarize, several reports describe an association of acne fulminans and joint disease. Our experience with our patient reinforces this concept. The arthropathy may range from a reactive phenomenon to a chronic, suppurative cutaneous disease, analogous to that of Reiter's disease or inflammatory bowel disease. The presence of HLA-B27 antigen in some cases including ours is of considerable interest. A role for immune complex disease has been suggested in the arthropathy associated with acne fulminans. Further research may indicate how this arthrocutaneous disorder should properly be categorized with respect to the reactive arthritides. The arthropathy associated with acne fulminans is believed to be silent and characterized by normal radiological findings.

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