Radiologic Features of Acne Fulminans

Hagit Peleg MD1, Benjamin Koslowski MD1, Nurit Hiller MD2 and Samuel N. Heyman MD1

Departments of 1 Medicine and 2 Radiology, Hadassah-Hebrew University Medical Center, Jerusalem, Israel

KEY WORDS: acne vulgaris, osteomyelitis, SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis), isotretinoin

A 17 year old male, previously healthy, was diagnosed with acne and treated with isotretinoin 60 mg/day. He did not receive any other medications. During the following 2 months he became bedridden due to progressive weakness, fever, weight loss, night sweats and severe arthralgia. His physical examination was remarkable for severe nodulocystic acne [Figure 1], fever, and diffuse skeletal and joint pain on passive motion, especially involving the hip and sacroiliac joints, with pronounced tenderness over both clavicles. Laboratory evaluation revealed leukocytosis, increased C-reactive protein and mildly disturbed liver function tests. Plain X-rays disclosed irregular cortical erosions of the right humeral mid-shaft with peri-osteal reaction [Figure 2A] and obliteration of the sacro-iliac joints [Figure 2B]. Technetium-99 bone scan [Figure 2C] revealed scattered enhanced uptake, most intense at the right mid-shaft humerus, median head of the right clavicle, left acromion and mid-sternum. Computed tomography showed bilateral axial lymphadenopathy and several osteolytic lesions, most prominent at the proximal humerus, with medullary involvement, cortical erosions and periosteal response, which on magnetic resonance imaging appeared with intense T2 and reduced T1 signals, undergoing enhancement with gadolinium [Figure 2D]. Repeated biopsies excluded bacterial infection and malignancies, leading to a diagnosis of acne fulminans.

Acne fulminans is a rare form of acne vulgaris associated with systemic symptoms such as fever, malaise, arthralgia and elevated erythrocyte sedimentation rate and C-reactive protein [1]. Aseptic osteolytic bone lesions such as those described here have been reported. The predominant locations are in the clavicle, sternum and long bones of the extremities [2]. It may also be a part of the spectrum of SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis) [3]. Its pathogenesis remains speculative and may be an immunologically mediated type III or IV hypersensitivity reaction to Propionibacterium acnes antigens [4]. Isotretinoin has been suspected to precipitate acne fulminans by neutrophil pro-inflammatory priming [5] or by increasing the pilosebaceous duct epithelium fragility and thereby increasing immune response and neutrophil contact with P. acnes antigens. Elevated blood levels of testosterone may also play a role in...
this disease which primarily affects male adolescents [1].

Treatment options based on uncontrolled series include high dose non-steroidal anti-inflammatory drugs, steroids alone or in combination with isotretinoin, antibiotics or anti-tumor necrosis factor-alpha interventions. Our patient was treated with NSAIDs, steroids and tetracycline with prompt resolution of symptoms and normalization of inflammatory indices, but was lost to follow-up.

Correspondence:
Dr. S. Heyman
Dept. of Medicine, Hadassah-Hebrew University Medical Center, Mt. Scopus, P.O. Box 24035, Jerusalem 91240, Israel
Fax: (972-2) 584-4526
email: heyman@cc.huji.ac.il

References

Cell competition is a tumor suppressor mechanism in the thymus

Cell competition is an emerging principle underlying selection for cellular fitness during development and disease. Competition may be relevant for cancer, but an experimental link between defects in competition and tumorigenesis is elusive. In the thymus, T lymphocytes develop from precursors that are constantly replaced by bone marrow-derived progenitors. Martins et al. show that in mice this turnover is regulated by natural cell competition between ‘young’ bone marrow-derived and ‘old’ thymus-resident progenitors that, although genetically identical, execute differential gene expression programs. Disruption of cell competition leads to progenitor self-renewal, up-regulation of Hmga1, transformation, and T cell acute lymphoblastic leukemia (T-ALL) resembling the human disease in pathology, genomic lesions, leukemia-associated transcripts, and activating mutations in Notch1. Hence, cell competition is a tumor suppressor mechanism in the thymus. Failure to select fit progenitors through cell competition may explain leukemia in X-linked severe combined immune deficiency patients who showed thymus-autonomous T cell development after therapy with gene-corrected autologous progenitors.

Nature 2014; 509: 465
Eitan Israeli

The pathology of severe schistosomiasis

Schistosomiasis-causing blood flukes infect hundreds of millions of people in tropical regions, but the occurrence of pathology is highly variable, with 5 to 10% of infections becoming severe. Likewise, schistosome infections take very different courses in different strains of mice, a phenomenon that relates to their relative ability to generate lymphocytes classified as CD4+ T helper 17 (TH17) cells. Sick children with blood flukes have also been found to have higher percentages of CD4+ TH17 cells. Ponichtera and colleagues have now discovered that the antigen-presenting dendritic cells of a mouse strain that develops severe hepatic granulomatous responses to schistosome eggs have a many times greater expression of a C-type lectin receptor called CD209a (a homolog of human ICAM-3-grabbing nonintegrin) on their cell surfaces as compared with a mouse strain that shows little pathology. CD209a is essential for the induction of the cytokines interleukin-1β and interleukin-23 that stimulate CD4+ TH17 cell development. Possibly the pathology of severe schistosomiasis is caused by elevated CD209a levels in some people sensitizing recognition of the fucose-rich glycans that coat the parasites’ egg surface.

J Immunol 2014; 10.4049/immunol.1400121
Eitan Israeli

“Just as appetite comes by eating so work brings inspiration”

Igor Stravinsky (1882-1971), Russian (and later, a naturalized French and American) composer, pianist and conductor, widely considered one of the most important and influential composers of the 20th century. Stravinsky’s compositional career was notable for its stylistic diversity. He first achieved international fame with three ballets commissioned by the impresario Sergei Diaghilev and first performed in Paris by Diaghilev’s Ballets Russes: The Firebird, Petrushka and The Rite of Spring.