Reply

Sir—The original version of our manuscript [1] included the reference to vincristine that Dr. Ena and colleagues refer to [2], but that reference was deleted during revision of the manuscript because of space constraints. Vincristine remains an unproven, albeit potentially useful, therapy for thrombocytopenia. Prospective studies are needed to determine what role this agent will play in the treatment of thrombocytopenia.

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References

Recurrent Erysipelas or Erysipeloid Rash?

Sir—The photo quiz in the June 1995 issue of Clinical Infectious Diseases [1] reminded us of a case with a very similar presentation in which our differential diagnosis included familial Mediterranean fever (FMF) and familial Hibernian fever (FHF) [2, 3]. Our patient, a Dutch male who was born in 1948, had a history of episodes of fever, erysipelas-like rash over the buttocks, and edema of the genitals (figure 1) and signs of acute-phase response (raised erythrocyte sedimentation rate and neutrophilia). The disease started at the age of 14 and occurred 3–8 times per year [4]. The patient’s family history revealed that his father, who had died, may also have had recurrent erysipelas-like lesions.

Since the patient had not responded to treatment with penicillin (for presumed erysipelas) or to prophylaxis with colchicine (for FMF), we considered the possibility of FHF in our differential diagnosis; in this disease, skin lesions are associated with painful erythema of the arms, trunk, and legs [3]. In 1987, however, he experienced a severe attack. At this time the patient had exanthem and a high fever (temperature of ≥40°C) as well as painful shoulders and knees. He was treated with prednisolone and azathioprine for presumed FHF and/or vasculitis. His condition did not improve, and he was transferred to the Academic Medical Center in Amsterdam with a diagnosis of polymyositis.

On admission to the hospital, he had exanthem and his right upper arm was swollen; in addition, he had severely restricted movements of his shoulder joints in all directions. The erythrocyte sedimentation rate was 110 mm/h, and the WBC count was 18.6 × 10^9/L. A roentgenogram of his shoulders showed severe joint destruction. An echogram revealed an abscess in his right upper arm; pus was obtained by puncture of the abscess, and culture yielded group A β-hemolytic streptococci. The patient responded to a 14-day course of iv penicillin therapy (12 million units per day).

The patient relapsed 1 month after his discharge from the hospital; he was again treated with iv penicillin, which was followed by oral penicillin. He did not have any attacks for 4 months, after which the antibiotic therapy was stopped. He relapsed again and prophylaxis with penicillin was started. He did not have any attacks for at least 1 year. In retrospect, it is not unlikely that the treatment with prednisolone and azathioprine resulted in streptococcal sepsis and abscess formation; the last two conditions led us to believe that β-hemolytic streptococci caused all his attacks, and the diagnosis had to be changed to recurrent erysipelas.

With regard to Dr. Jones’ case, we believe that the diagnosis of erysipelas should be considered, especially since the clinical presentation was atypical for FMF and the patient did not respond to colchicine; according to published reports, colchicine prevents attacks in 64% of patients with FMF and markedly reduces the frequency and severity of attacks in 31% of these patients [5, 6].

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Progressive Multifocal Leukoencephalopathy as an AIDS-Defining Condition in a Patient with Human Immunodeficiency Virus Infection

SIR—We read with interest the recent article by Fong et al. [1] describing progressive multifocal leukoencephalopathy (PML) in patients with AIDS. They found that the mean (±SE) CD4+ cell count at the time of presentation or diagnosis was 85 ± 82/mm³ and that only two of 28 patients had CD4+ cell counts of >200/mm³. Also significant was the difference in survival time between patients with CD4+ cell counts of <90/mm³ and those with CD4+ cell counts of >90/mm³ at the time of presentation. We report an additional case in which PML was an AIDS-defining condition in a patient with HIV infection, who had a CD4+ cell count of 198/mm³ at the time of diagnosis.

A 30-year-old, HIV-infected female was transferred to our service for evaluation of progressive neurological complications, including right-sided hemiparesis. One week before admission, she was hospitalized twice for fever, malaise, and decreased level of consciousness. Dysarthria had progressed to mutism 2 days before, and she was unable to clear copious oropharyngeal secretions.

The patient's condition had been diagnosed as AIDS-related complex 5 years earlier, and she had begun treatment with zidovudine that was followed by didanosine therapy and prophylaxis with trimethoprim-sulfamethoxazole. Candida esophagitis had been suspected, and no opportunistic infections had been previously identified. Her medical history included hepatitis B and C, pelvic inflammatory disease, anogenital condylomata, maxillary sinusitis, pneumonia, motor aphasia, intravenous drug abuse, prostitution, and multiple suicide attempts.

A physical examination revealed a cachectic woman who was responsive only to pain. Her temperature was 38.9°C, and her pupils were dilated bilaterally 3–4 mm and were reactive. Mild nuchal rigidity and right hemiparesis were observed, as were a contracted right hand, a flaccid right lower extremity, and a positive right Babinski’s sign. Coarse rhonchi were present bilaterally.

The findings of laboratory studies indicated markedly elevated results of liver function tests as well as a moderate amount of blood and a high number of bacteria in the urine. A drug screen was negative. Findings on a chest roentgenograph were unremarkable. Cultures for fungi, viruses, and bacteria were negative. CT of the head (with contrast) demonstrated focal ischemic areas in the left parietal lobe. MRI revealed multiple areas of abnormal signal on T-2 weighting that involved the brain stem and paraventricular white matter (figure 1); it also showed severe maxillary sinusitis. An electroencephalogram (EEG) depicted generalized slow frequency, which was consistent with mild, diffuse encephalopathy. An EEG to measure auditory evoked response also revealed abnormalities.

Examination of CSF specimens did not reveal any abnormalities. Cultures for fungal, viral, and bacterial pathogens, including anaerobes and acid-fast bacilli, were negative. A VDRL (venereal disease research laboratory) test was nonreactive, and PCR for cytomegalovirus (CMV) was negative. The CD4+ cell count was 198/mm³ with a CD4/CD8 ratio of 0.5. Cultures for Mycobacterium avium complex, a test for serum cryptococcal antigen, anduffy coat stain and culture for CMV were negative. Serologies for human T-cell lymphotropic virus type 1 and 2 and for CMV were negative. Immunological tests...