CASE REPORTS

Refer to: Gomberg R, Hamm P, Martin A: Mucocutaneous lymph node syndrome (Kawasaki disease) in an adult. West J Med 135:406-408, Nov 1981

ABBREVIATIONS USED IN TEXT

CNS = central nervous system
ECG = electrocardiogram
MCLNS = mucocutaneous lymph node syndrome

Mucocutaneous Lymph Node Syndrome (Kawasaki Disease) in an Adult

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MUCOCUTANEOUS LYMPH NODE SYNDROME (MCLNS), also known as Kawasaki disease, is an acute disease characterized by exanthem, persistent fever, mucous membrane reddening, enlarged cervical lymph nodes, periungual desquamation, and multiple organ or system complications. Until very recently,[1-4] mucocutaneous lymph node syndrome was considered a disease of infants and young children.[5]

Diagnostic criteria established by the MCLNS study group are based on Kawasaki's original detailed description.[5] They are (1) fever of five or more days duration, (2) bilateral reddening of the ocular conjunctiva, (3) erythema, dryness and fissuring of the lips and tongue, (4) reddening of the oropharynx, (5) reddening and indurative edema of the hands and feet and periungual desquamation, (6) generalized exanthem (appearing scarlatiniform) and (7) acute swelling of the anterior cervical lymph nodes.[6] Prolonged fever accompanied by four of the other criteria and excluding other similar diseases establishes the diagnosis. This includes negative cultures (viral, rickettsial and bacterial) and lack of response to antibiotic drugs.

Thus, MCLNS is a clinical diagnosis of exclusion. The syndrome becomes apparent over time and as other possible diagnoses are discarded. The cause remains unknown although there are autopsy data linking MCLNS to infantile periarteritis nodosa.

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Report of a Case

A 22-year-old man reported that he had generalized ache, stomach cramps, rash and difficulty in breathing. Fever began on the first day of the illness. The patient was seen by one of the authors the same day and treatment with penicillin, the antispasmodic drug Donnatal and meprobamate was begun. Because of continuing fever and symptoms he was seen in the emergency room of La Habra Community Hospital two days later. Temperature was 40.4°C (104.7°F); there was a generalized exanthem (maculopapular), edema and erythema of the pharynx; the tongue was beefy red and coated, and generalized aching had persisted. The patient was admitted to hospital at the time of the emergency room visit.

A throat culture was negative for *Streptococcus* and *Staphylococcus,* six blood cultures were negative and a stool culture showed no enteric pathogens, with *Escherichia coli* as the predominant organism. Despite therapy with multiple antibiotic drugs, fever persisted and symptoms grew worse as characterized by nausea and diarrhea; joint pains; swollen, reddened gums; dry, cracked, peeling lips; bilateral ocular conjunctivitis; reddening and induration of the palms; strawberry tongue; pharyngitis and cervical adenopathy; gastrointestinal and liver involvement; persistent fever (ten days), and desquamation of skin (late onset), including the periungual areas of the fingers.

Laboratory Data

Serial leukocyte counts were as follows: day one, 9,200 leukocytes with a differential of 78

percent polymorphonuclear neutrophils, 10 percent band neutrophils, 6 percent lymphocytes, 5 percent monocytes, 1 percent basophils and adequate platelets; day two, 8,900 leukocytes with 73 percent polymorphonuclear neutrophils, 21 percent band neutrophils, 4 percent lymphocytes, 1 percent monocytes, 1 percent eosinophils and adequate platelets; day three, 8,500 leukocytes with 47 percent polymorphonuclear neutrophils, 35 percent band neutrophils, 6 percent lymphocytes, 8 percent monocytes and 4 percent eosinophils and adequate platelets; day four, 6,300 leukocytes with 43 percent polymorphonuclear neutrophils; 41 percent band neutrophils, 5 percent lymphocytes, 6 percent monocytes, 5 percent eosinophils and adequate platelets; day eight, 10,500 leukocytes with 75 percent polymorphonuclear neutrophils, 12 percent lymphocytes, 12 percent monocytes, 1 percent eosinophils and 516,000 platelets. Hemoglobin and hematocrit values were stable. The sedimentation rate was 42 mm per hour on day seven in hospital. Analysis of urine was positive for bilirubin (2+) and urobilinogen (8 Erlich units). Follow-up analysis of urine was negative for urobilinogen and positive for bilirubin. Two heterophile tests were negative. Antistreptolysin O titer was negative, C-reactive protein was positive (1:5), liver enzyme levels remained abnormal throughout the clinical course along with prolonged prothrombin time and elevated bilirubin levels (2.6 to 4.9 mg per dl). Hepatitis antigen studies were negative and cytomegalovirus titer was negative as was VDRL. Cold and febrile agglutinin tests were negative; blood urea nitrogen levels remained normal, and direct and indirect Coombs' tests were negative. An electrocardiogram (ECG) showed nonspecific ST wave elevations. The patient was discharged afebrile, with abating symptoms but still icteric, on a regimen of 1,200 mg (four 5-grain tablets) of aspirin four times a day.[7] Because MCLNS was not suspected until late in the clinical course, earlier sedimentation rates and platelet counts were not done.

Posthospital Course

The patient has remained asymptomatic with follow-up ECG and biplanar echocardiogram showing no abnormalities. Aspirin therapy has been continued at a reduced dosage, and long-term echocardiographic follow-up is anticipated. A convalescent titer for leptospirosis was negative.

Discussion

This case and other cases in adults reported in the United States literature[1-4] show the need for family practitioners and internists to be as alert to MCLNS as pediatricians have become. Aside from the prolonged illness and associated morbidity, MCLNS has grave implications for heart and coronary artery disease occurring in the later phases—often after discharge from hospital. The cardiac lesions usually appear in one of two forms: arrhythmia with possible sudden death[8] or coronary artery arteritis with or without formation of microaneurysms.[9] The arteritis may lead to myocardial infarction or an aneurysm may rupture. Most deaths occur between days 11 and 50 of the disease. The pediatric literature shows 1 percent to 3 percent mortality secondary to cardiac complications and up to 30 percent cardiac involvement in patients with MCLNS.[8,9]

Other major systems involved are genitourological (renal), adrenal and central nervous system (CNS). CNS symptoms include irritability, meningismus and positive lumbar puncture. Cerebrospinal fluid may show 25 to 100 leukocytes per cu mm. The predominant cells are lymphocytes. Protein and glucose levels in cerebrospinal fluid remain within normal limits. Many of these complications seem related to arteritis.[10]

Therapy is aimed at preventing arteritis and cardiac complications. Acetylsalicylic acid (aspirin) in standard therapeutic dosages (80 to 100 mg per kg of body weight per day) has been effective as an antiinflammatory agent and tends to inhibit platelet aggregation. Thrombocytosis is a fairly uniform laboratory finding in MCLNS. The incidence of microaneurysm formation and coronary artery arteritis is lowest in patients treated with aspirin. Aspirin malabsorption and the need for extra-high dosage has been reported,[7] and serum levels should be followed carefully to avoid overdosage.

Corticosteroids appear contraindicated. A controlled study showed increased microaneurysm formation in patients taking corticosteroids. Aneurysms developed in 65 percent of these patients as opposed to 11 percent of those taking aspirin.[11] In some patients microaneurysms resolve spontaneously as evidenced by multiplane echocardiography; however, others have persistent lesions and may eventually require coronary artery bypass operations.

It is anticipated that MCLNS will be recognized

far more frequently in adult patients as articles such as this appear in the literature. It is essential that family practitioners and internists be able to recognize and treat MCLNS appropriately.

Summary

This is one of the rare reports in the United States literature of an adult with mucocutaneous lymph node syndrome (Kawasaki disease), an increasingly common pediatric problem. It is anticipated that MCLNS will be reported with increasing frequency in the adult population as family practitioners and internists become more familiar with this syndrome. MCLNS is of significance because of a 1 percent to 3 percent mortality resulting from cardiac complications of coronary artery arteritis, arrhythmias, coronary artery microaneurysm formation and prolonged morbidity.

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Refer to: Rittenberg GM, Korn JH, Schabel SI, et al: Rapid osteolysis in pancreatic carcinoma. West J Med 135: 408-411, Nov 1981

Rapid Osteolysis in Pancreatic Carcinoma

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THE BONY MANIFESTATIONS of pancreatitis are usually related to ischemia. Circulating lipolytic and proteolytic enzymes may produce endothelial damage and vascular compression, which may occlude vessels with resultant ischemia and infarction.[1-6] When vascular compromise occurs in the shaft of bones, medullary infarcts are produced; when it occurs at the end, aseptic necrosis of articular surfaces results. We present a case of rare bony manifestations of pancreatic disease related to ischemia, diagnosed as rapid osteolysis in pancreatic carcinoma.

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Report of a Case

A 63-year-old black man, a chronic alcoholic, was admitted to hospital with pain and swelling of four days' duration in his right foot, accompanied by fever and shaking chills. There was no history of trauma or preceding arthritis. His right foot and ankle were swollen and tender and two nodules, 1.5 by 1.5 cm in size, were palpable on the right achilles tendon. Initial laboratory studies gave the following values: hemoglobin 9 grams, leukocyte count 9,600 per cu mm, albumin 2.2 grams per dl, serum aspartate aminotransferase 48 IU (normal less than 50 IU), alkaline phosphatase 500 IU (normal 30 to 85 IU), bilirubin 2.4 mg per dl (1.6 per dl direct), serum amylase 18,350 IU (normal 60 to 300 IU) and serum lipase 20.7 IU (normal 0.7 to 1.0 IU).

The right third metacarpophalangeal joint, initially slightly swollen, became exquisitely tender and erythematous the day after admission. Initial roentgenograms (Figure 1A), aside from soft tissue swelling, showed no abnormalities. A week after admission, however, substantial destruction was noted in the distal end of the third metacarpal and in the fifth midphalanx (Figure 1B). Fluid aspirated from the joint contained no organisms on