Manifestation of X-Linked Agammaglobulinemia

We report a case of recurrent pneumococcal arthritis as the presenting manifestation of this condition [3].

*Streptococcus pneumoniae* is an infrequent cause of septic arthritis and seldom infects the joints of children older than 2 years. In one series of 22 children with pneumococcal arthritis, the age of the oldest patient was 23 months [6]. We report a case of recurrent pneumococcal septic arthritis as the presenting manifestation of X-linked agammaglobulinemia in a school-aged child.

A 5-year-old boy presented to the emergency department with right knee pain of 12 hours’ duration, swelling, erythema, and refusal to bear weight on his right leg. He had been well until the previous evening, when he experienced right knee pain after a trivial bump against a table. He was afebrile and had no other symptoms.

The patient’s past medical history was notable for the presence of septic arthritis of the right knee 2 years previously. At that time, he experienced swelling and tenderness of the knee after bumping into a swing set. Cultures of blood and synovial fluid specimens grew *S. pneumoniae*. After undergoing incision and drainage of the right knee, he received ceftriaxone and rifampin for 21 days and had a complete recovery. The patient had no other hospitalizations and showed no history of recurrent sin-

References


Recurrent Pneumococcal Arthritis as the Presenting Manifestation of X-Linked Agammaglobulinemia

Pneumococcal arthritis in children older than 24 months is unusual and can suggest underlying immunodeficiency. We report a case of recurrent pneumococcal arthritis as the presenting manifestation of X-linked agammaglobulinemia.

X-linked agammaglobulinemia is characterized by a profound deficiency of circulating B cells and serum immunoglobulin that results from a defect in the gene encoding Bruton’s tyrosine kinase (Btk) [1, 2]. Patients with this immunodeficiency typically present during the first year of life with recurrent bacterial infections, most commonly with otitis media, pneumonia, gastrointestinal infections, or pyoderma [3, 4]. Mono- or oligoarticular arthritis in patients with X-linked agammaglobuli-
opulmonary or skin infections. There was no family history of sickle cell anemia or unusual susceptibility to infections.

In the emergency department, the patient was afebrile, and his right knee was warm, swollen, erythematous, and tender and had limited range of motion. The skin overlying his right knee was intact, and he would not bear weight on his right leg. Examination did not reveal other musculoskeletal abnormalities. Radiographs of the right knee revealed a joint effusion without bony abnormality. His peripheral WBC count was 21,900 cells/µL (68% leukocytes), and his erythrocyte sedimentation rate was 36 mm/h. Arthrocentesis involved the removal of 25 mL of sero-sanguinous synovial fluid that revealed gram-positive diplococci on Gram’s stain and that yielded S. pneumoniae on sheep blood agar culture plates. The patient underwent incision and drainage of his right knee and was treated with iv ceftriaxone and clindamycin for 21 days; he had a complete recovery.

Additional laboratory studies were undertaken to evaluate the child for conditions that would predispose to pneumococcal septic arthritis. The results of screening for sickle cell anemia and of HIV ELISA were negative, and a total hemolytic complement level was normal. Quantitative immunoglobulin studies showed an IgG level of 123 mg/dL (normal range, 633–1280 mg/dL), an IgM level of 31 mg/dL (normal range, 48–207 mg/dL), and an IgA level below the limits of detection. Flow cytometry analysis showed normal T cell counts and an absence of CD19-positive B cells. Findings of CD19-positive B cell levels of 2% in the peripheral circulation and a marked reduction in concentrations of IgG, IgM, and IgA in a patient with recurrent bacterial infections suggested the diagnosis of X-linked agammaglobulinemia [7]. Intravenous immunoglobulin therapy was started [8, 9], and genetic studies were performed, which revealed a 3-bp deletion in exon 15 of the Btk gene and thus confirmed the diagnosis of X-linked agammaglobulinemia.

The serotypes of the pneumococcal isolates were compared by means of the Quellung reaction with rabbit polyclonal pneumococcal typing antisera. The S. pneumoniae isolate recovered from synovial fluid obtained from the patient’s right knee in 1997 was of serotype 14, whereas the isolate recovered from the same site in 1999 was of serotype 6. These data demonstrate that the 2 episodes of septic arthritis represent separate infections, rather than a persistent infection, in this immunocompromised child.

Recurrent pneumococcal septic arthritis in an otherwise healthy child is an unusual presentation of X-linked agammaglobulinemia. It is of special note that our patient had pneumococcal joint infections at 3 years and 5 years of age. When pneumococcus is isolated from the synovial fluid of a child older than 24 months, further studies to identify a predisposing condition should be considered [10].

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References


Human Granulocytic Ehrlichiosis Presenting as Facial Diplegia in a 42-Year-Old Woman

Neurologic manifestations of human ehrlichiosis are unusual and have been described almost exclusively in human monocytic ehrlichiosis associated with Ehrlichia chaffeensis. We report here a case of a previously healthy 42-year-old woman who developed bilateral facial nerve palsies in association with infection by the agent of human granulocytic ehrlichiosis (aoHGE). The diagnosis was made by specific polymerase chain reaction amplification of aoHGE sequences from samples of the patient’s blood and cerebrospinal fluid (CSF), as well as propagation of aoHGE in culture of HL60 cells inoculated with the pa-