CRYPTOCOCCUS NEOFORMANS
OSTEOMYELITIS OF THE CLAVICLE

Donna Magid, MD
Brian Smith, BS

Cryptococcal musculoskeletal infections are often associated with immunosuppression and/or inoculating local trauma. An unusual case of apparently isolated, rapidly progressive, clavicular osteomyelitis was seen in a woman who denied any previous trauma and with only marginal, if any, immunocompromise. Plain films, scintigraphy, and computed tomography (CT) were used to direct her diagnostic and therapeutic course.

CASE REPORT

A 54-year-old woman with diabetes mellitus presented with a 1 month history of progressive left shoulder pain and loss of range of motion associated with an enlarging clavicular mass. Initial consultations elsewhere did not localize a source for her discomfort, and she was started on nonsteroidal antiinflammatory medication with some subsequent pain relief. However, the soft tissue mass continued to enlarge, and she presented to our institution.

On physical examination, a 10 cm × 12 cm tender, firm mass was visible and palpable over the proximal aspect of the left clavicle. Range of motion of the left shoulder was markedly limited secondary to pain. Physical and neurological examination was otherwise unremarkable. Laboratory values were notable for an alkaline phosphatase of 465, and a normal leucocyte count and differential. She was afebrile.

Initial radiographs of the chest 3 weeks earlier had shown a subtle, discrete lytic lesion just lateral to the clavicular head (Fig 1A). At second presentation, this had expanded and enlarged, with ill-defined soft tissue density also increasing in the interim (Fig 1B). CT of the neck and thorax was obtained, revealing a 6 cm inflammatory chest wall mass centered around the left clavicular head with focal lytic destruction of the medial aspect of this bone (Fig 2). A Te99M methylene diphosphonate bone scan demonstrated increased uptake in the medial one third of the clavicle with a small focal photopenic area.

Fine needle aspirate of the clavicular soft tissue mass was performed. Cytopathology (Fig 3) confirmed an inflammatory process with numerous multinucleated histiocytes in association with both intracellular and extracellular organisms, morphologically consistent with Cryptococcus. Histochemical staining demonstrated the very characteristic mucicarmine positive mucopolysaccharide capsule of Cryptococcus. Budding encapsulated yeast forms were plainly seen on both Papanicolaou and Diff-quick smears. Although aspirate cultures failed to recover the organism, latex agglutination for serum cryptococcal antigen was positive at 1:256. The patient was begun on IV amphotericin B and oral 5-flucytosine for synergy to preclude possible disseminated cryptococcal infection. Blood cultures remained negative as did CSF cell count and culture and HIV tests. Immunologic and S-PEP evaluations were completed and found to be consistent with chronic infection. The patient was appropriately reactive to an anergy panel screening for common antigens.

Clavicular resection or curettage was considered by surgical consult but deferred, as secondary damage to mediastinal vessels, brachial plexus, or thoracic cavity was thought probable if extensive debridement was undertaken. Once adequate clinical response to therapy was confirmed, a Hickman catheter was
Fig 2: Transaxial CT scan through the clavicle. Lung windows (WW 1650, WC-450) show aggressive endosteal erosion of the anterior cortex and destruction of the posterior cortex (A, arrowhead). Soft tissue windows show the tissue-density medullary matrix in continuity with an overlying deep anterior soft tissue mass (arrows), with a less extensive retroclavicular soft tissue inflammatory response effacing fascial tissue planes. There is anterior local cellulitis and skin thickening of the chest wall (B).

Fig 2A.

Fig 2B.

placed and the patient was discharged for home therapy on amphotericin B. Follow-up CT 10 months after presentation confirmed ongoing clavicular healing and decreased soft tissue mass.

DISCUSSION

Cryptococcal infection was infrequently diagnosed prior to the mid 1980s, but has increased steadily in the era of AIDS. Up to 9% of AIDS-defined patients have been reported with prior or present evidence of cryptococcal infection in the United States. Prior to 1984, half of all cryptococcal infections occurred in the presence of other immunocompromising states such as sarcoidosis, lymphoma, leukemia, tuberculosis, diabetes mellitus (as with this patient), and iatrogenic immunosuppression due to steroid treatment or chemotherapy.

The organism Cryptococcus is a nonmycelial budding yeast that inhabits soil after deposition via pigeon droppings. Inactive forms can reactivate years after exposure, if the host immune mechanisms have been compromised or lost. Skeletal involvement is rare in non-AIDS patients, occurring in only 5% to 10% of cases with disseminated or isolated cryptococcal infection. Of this percentage less than one half (2.5% to 5%) may have isolated osteomyelitis (vs documented disseminated disease with subsequent hematogenous bone involvement). Almost every major bone has been reported as susceptible to cryptococcal infection, with the vertebrae, pelvis, and ribs commonly involved.

Isolated cryptococcal osteomyelitis without traumatic inoculation is rare and is a diagnosis of exclusion in the absence of other organ involvement. In this case no other sites of infection were confirmed. No history of local trauma could be elicited, and HIV screen was negative.

Presentation is indolent; fever is rare and white blood cell count is normal or only slightly elevated in all groups. Serum cryptococcal antigen may remain negative even when infection is proven by culture.

Radiographically, bone lesions are geographically osteolytic with variable definition of margin and with periosteal reaction or marginal sclerosis only infrequently present. Local soft tissue involvement often accompanies the bone infection. Adjacent joints may be involved.

Differential diagnosis includes other infective lesions (coccidiomycosis, tuberculosis, actinomycosis, blastomycosis) or neoplastic disease.

Fig 3: Fine needle aspiration of inflammatory soft tissue mass, mucicarmine stain. Mucicarmine positive capsules are seen in both extra- and intracellular encapsulated yeast forms. The organisms are associated with inflammatory cells, including multinucleated histiocytes.
Radioisotope scanning may show increased tracer uptake at the site of involvement.

CT evaluation has proven valuable in characterizing the location and extent of lytic destruction and soft tissue changes in cryptococcal osteomyelitis. In this case, CT confirmed the extent of bone destruction and guided aspiration for diagnosis. This study also demonstrated the extent of retroclavicular soft tissue changes, suggesting that definitive debridement could be threatening to underlying mediastinal and thoracic structures. However, surgical treatment is usually preferred.

REFERENCES

From the Department of Orthopedic Surgery and the Russell H. Morgan Department of Radiology and Radiological Sciences, The Johns Hopkins Medical Institutions, Baltimore, Md.

Reprint requests: Donna Magid, MD, Dept of Orthopedic Surgery, The Johns Hopkins Medical Institutions, Baltimore, MD 21205.

LEGG-CALVE-PERTHES DISEASE AND SLIPPING OF THE CAPITAL FEMORAL EPIPHYSIS IN THE SAME CHILD

Mark J. Ruoff, MD*
Edward P. Schwentker, MD†

Slipped capital femoral epiphysis (SCFE) occurring concurrently with Legg-Calve-Pertkhes (LCP) disease is a rare occurrence, and only five reported cases exist in the literature. Marklehm1 and Crawford2 reported individual cases and Graziano et al3 reported a series of three cases seen over 18 years. Each of these cases included a slip on the contralateral side from the hip affected by Perthes disease. Ipsilateral slip current with Perthes disease has been alluded to in the literature,4 but has not been formally reported.

We report two cases of slipped capital femoral epiphysis coexistent with Perthes disease, one on the contralateral side and one on the ipsilateral side.

CASE REPORTS

Case 1. A 14-year-old boy presented with a longstanding history of left hip pain and a limp. He had decreased hip abduction with a Trendelenburg sign, and radiographs were significant for severe Perthes disease with coxa magna and greater trochanteric overgrowth. He underwent cheliectomy and greater trochanteric advancement from which he recovered uneventfully with a good result.

Two years later, at age 16, he complained of the insidious onset of pain in the right hip, and radiographs were significant for a grade II SCFE with chronic changes (Fig 1). He was treated with in situ percutaneous pinning. Postoperatively he began partial weight bearing on the right side, and by 12 weeks was full weight bearing. Healing of the slipped epiphysis was complete.

Case 2. A 13-year-old boy with Down's syndrome presented complaining of pain in his left hip and thigh for 1 year. Radiographs were significant for severe Perthes with coxa magna and plana. He was placed in a Toronto brace for 1 year with minimal relief. At that point there was approximately 25% head protrusion, a Catterall classification of IV, 20° of abduction, and a femoral head in the reossification phase. He underwent cheliectomy of the left hip. Postoperatively he was placed in Buck's traction for 3 weeks, was started on partial weight bearing, and then advanced to full weight bearing by 6 weeks.

He did well until 1 year 4 months postoperatively, when he fell from a bed and complained of pain in his left hip and knee. Radiographs were significant for slipping of the capital femoral epiphysis on the left side (Fig 2). He was treated with Buck's traction for 3 days and underwent percutaneous pinning. Six years later he was pain free with full flexion and extension, and 5° of internal and external rotation with 40° of abduction.

DISCUSSION

The coexistence of Perthes disease and SCFE is an uncommon entity, as evidenced by the small number of cases reported. Crawford2 calculated that the incidence of both occurring in the same child is .71/3.41/12.000.000. He believed that the simultaneous occurrence is purely by chance and are independent events, with no similarities between cases to suggest a link between the Perthes disease and slipping of the capital femoral epiphysis.