treatment support a primary cause for the deformity.

Case 1 did have a diagnosis of bilateral metatarsus adductus at birth and was casted with reported initial correction that deteriorated with development. In this case casting might have played a role in the deformity. In case 2 corrective shoes were applied to a unilateral deformity that progressively became worse with age despite the treatment. It is doubtful that this deformity was caused by the corrective therapy.

The delayed and progressive nature of the deformity in both patients is remarkable. Both patients were treated with conservative bracing, and the initially supple deformities later progressed. The progression of the deformity and the transition from a relatively supple deformity to a fixed deformity is consistent with the natural history of skew foot as described by several authors.6,7 In both cases operative intervention has become necessary9,11 and may be more successful if performed early, as advocated by Bleck.10 In cases in which the uncorrected deformity becomes inflexible and severe, the option for hindfoot arthrodesis and forefoot realignment may be a reasonable surgical alternative at the time of skeletal maturity.

REFERENCES


EDITORIAL DISCUSSION

ORTHOPEDICS: Is first web space widening pathognomonic for this condition?

Scully and Ferguson: The condition of metatarsus adductovarus, as described by McCormick and Blount and subsequently by others, is based on the relationship of the midfoot to the fore- and the hindfoot. The deformity is described as hindfoot valgus, lateral navicular subluxation, and marked forefoot adduction. In both cases reported here, the severe forefoot supination that is associated with the forefoot adduction limits the ability to evaluate the intermetatarsal angle radiographically.

ORTHOPEDICS: Is the Z-foot a cosmetic or functional problem?

Scully and Ferguson: In the current report both individuals exhibited a significantly delayed ambulation at 34 and 42 months. These individuals displayed multiple manifestations of a genetic syndrome and had documented deletions of chromosome 15. Whether the delay in ambulation was attributable to the functional limitations due to the skew foot deformity or was of a more complex etiology associated with abnormal neuromuscular development is not entirely clear. The children walked with an ataxic gait pattern before surgery and the first case continued to do so after a period of convalescence.

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RECURRENT CERVICAL
SUBLUXATIONS IN A PATIENT WITH GOUT AND ENDSTAGE RENAL DISEASE

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The inflammatory condition gout or gutta ("a drop" in Latin) represents a spectrum of pathologic conditions resulting from the deposition of monosodium urate crystals in bodily tissues when the body is unable to accommodate uric acid overproduction or undersecretion.1,2 The association of gouty arthritis afflicting the spine has been mentioned sporadically in the literature.3-10 To our knowledge there are only five cases of tophaceous involvement of the spine resulting in neurologic cord compression,3,7,8 and only one case of true tophus involvement of the cervical spine with subsequent subluxation.11
Fig 1: A sagittal T2-weighted MRI showing bone erosive changes to vertebral bodies C2 through C6 with obvious scalloping seen in the posterior body of C5 consistent with urate crystal bony deposition.

Reported is a rare case of a 67-year-old man with severe systemic gouty disease and renal failure who experienced multiple episodes of cervical subluxations due to soft tissue and bony erosion from gouty tophus deposition. This case is reported to alert the physician that a patient with severe systemic gout involvement may be at risk for neurologic deficit due to gouty involvement of the cervical spine with resultant subluxation.

CASE REPORT

A 67-year-old man had endstage renal disease for more than 10 years due to urate nephropathy. He initially presented in 1986 with weakness in all extremities, upper greater than lower, after a minor traumatic event. The patient was given a diagnosis of central cord syndrome in the setting of severe cervical spondylosis. He underwent an anterior cervical decompression with fusion at C4-5 and had a full neurologic recovery. At 1-year follow up, the patient again presented with weakness in all extremities; plain radiographs showed a retrolisthesis of C5 on C6. The patient subsequently underwent a posterior cervical fusion with wiring and autogenous iliac bone graft from C4 to C6 with an uneventful recovery. Two months prior to his most recent admission in January 1989, the patient had noted gradual return of weakness in both upper and lower extremities. He was unable to walk with assistance of a cane, become wheelchair bound, and was unable to feed himself. The patient was being treated by hemodialysis for urate nephropathy and was on colchicine.

Physical examination at this time was significant for a cachectic appearing black man with large soft tissue nodules (3 cm × 4 cm) over both olecranon bursas, and numerous nodules in the subcutaneous tissues on the extensor surface of his forearms, Achilles tendons, and both eyelids. The neurologic examination was significant for bilateral decreased sensation to light touch in the C5-C8 dermatomes. Motor examination, according to the standards adopted by the American and British Academies of Orthopaedic Surgeons, was graded as 2/5 in C5, 3/5 in C6-C8, 4/5 in L2, and 3/5 in L3 through S1 bilaterally. Reflexes were not exaggerated, but Babinski tests were positive bilaterally. The laboratory examination on admission revealed the following values: hemoglobin 13.4 g/dL (14-18), hematocrit 40.8% (40% to 54%), BUN 35 mg/dL (10-20), creatinine 5.6 mg/dL (<1.5), calcium 10.2 (9-10.5), PO4 3.0 mg/dL (3-4.5), and uric acid 6.6 (2.5-8 mg/dL).

A plain lateral radiograph showed a 25% anterior subluxation of C2 on C3 with extensive erosive changes between C3 and C6. A sagittal T2-weighted MRI revealed entire vertebral body erosive changes, including posterior elements with both anterior and posterior spinal cord compression between C2 and C6 (Fig 1). An axial CT scan was performed at the level of C6, again revealing pan vertebral body destruction with obvious involvement of both facet joints (Fig 2).

The cervical spine was reduced with Gardner-Wells tong traction followed by a posterior cervical fusion from C2-C6 (Fig 3). On exposure of the cervical posterior elements a white chalky substance characteristic of a tophaceous deposit was noted involving the posterior elements with obvious destruction of the facet joints bilaterally between C2 through C6. Postoperatively, the patient was treated with halo immobilization and did well with a complete grade improvement in all muscle groups. At 12-month follow up, the patient was ambulatory with a walker and able to feed himself. His individual muscle strengths remained essentially unchanged until his death due to renal nephropathy 15 months after his last surgical procedure.

At autopsy several gouty tophus specimens were studied histologically. These included lesions in bilateral hands, elbows, and kidneys, the last also displaying end-stage changes of chronic uric acid nephropathy and nephro lithiasis. More specifically, in formalin-fixed tissues, the renal tubules were dilated by homogeneous, pink amorphous material, with concomitant atrophy and hyalinization of surrounding structures. The interstitium revealed a classical granulomatous response to urate: dense infiltration by lymphocytes, macrophages, fibroblasts, and foreign body giant cells. With alcohol fixation, the amorphous precipitants were revealed as closely packed sheets of 5 µm to 20 µm long needle-shaped crystals that were doubly refractive on polaroscopic examination (Figs 4-5).
DISCUSSION

Gout is an inflammatory process characterized by a deposition of monosodium urate crystals in bodily tissues causing the spectrum of severe articular or periarticular inflammation, tophaceous crystalline deposits in numerous tissues, and severe renal disease. Gout is known primarily for its predilection for the appendicular musculoskeletal system with soft tissue manifestations such as symmetric polyarthritis, swelling of peripheral joints, and subcutaneous nodules in para-articular areas. Lichtenstein et al. noted other unusual sites of involvement in a series of 11 necropsied cases including the penis, cornea, eyelids, myocardium, great vessels, tongue, and vocal cords. In comparison, there are relatively few examples reported of gouty involvement of the spine. The first mention of spinal involvement was by Bauer and Klemperer with pathologic confirmation by Lichtenstein et al. Hall and Selin discussed the findings of an autopsy of a 51-year-old man with a history of gouty symptoms but without back pain who, at autopsy, was found to have deposits of uric acid crystals in the posterior capsules, ligaments, and articular surface of the lumbar spine. Uric acid deposits have also been noted in the vertebral bodies, facet joints, and discs with extensive destruction. Diffuse idiopathic skeletal hyperostosis (DISH) has been noted in association with gout.

Nervous system involvement in association with gout is rare. Previous reports include median nerve compression by uric acid deposits, as well as quadriparesis or paraparesis due to an epidural tophus. Spinal involvement that is severe enough to cause subluxation is extremely rare, with only one previously mentioned upper cervical subluxation in a gouty patient.

Radiographic findings in gout began initially with nonspecific evidence of para-articular soft tissue swelling with preservation of joint spaces.
With chronic involvement articular cartilage thinning occurs with subchondral sclerosis and cystic degeneration with erosive changes and even ankylosis. Constanz and Bluestone described the possibility of an extensive bony lytic lesion (pseudotumor) due to a tophus causing expansion of the cortex with new bony formation.

There is the possibility of an associated destructive spondylarthropathy due to hydroxyapatite crystal deposition, aluminum toxicity, amyloidosis, infection, or hyperparathyroidism in a patient with gouty spondylarthropathy. The patient with hyperparathyroidism should have abnormal serum calcium, phosphorus, and parathyroid hormone levels. Our patient presented with a long history of hemodialysis that has been associated with severe narrowing of the cervical intervertebral disc space and endplate erosion, and the absence of osteophytes. Secondary hyperparathyroidism may be present in patients with renal failure on hemodialysis with an associated characteristic destructive spondylarthropathy. Our patient had the chalky white substance at surgery that was morphologically characteristic of gout, with infection ruled out by intraoperative negative cultures.

In summary, this case illustrates the rare occurrence of cervical subluxation with myelopathy as a consequence of severe tophus involvement of cervical spine in a patient on hemodialysis due to urate nephropathy. Although unusual, the diagnosis of gouty involvement of the spine should be entertained in patients with spinal symptoms and ruled out in patients with severe systemic gouty involvement who may undergo any manipulation of the spine, i.e., general endotracheal anesthetics. Posterior surgical fusion is an efficacious mode of treatment for the unstable cervical spine in patients with gout. However, careful routine follow-up examinations are advised, as the stability of adjacent cervical spinal segments after previous fusion may be compromised due to stress-loading, especially in patients with weakened supporting tissues due to gout.

REFERENCES


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