Extradural spinal involvement by gout

Case report

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A case of extradural gouty tophus in the lumbar region in a teen-age girl is presented as an addition to the differential diagnosis of erosive lesions of the spinal canal.

KEY WORDS - gout - gouty tophus - extradural spinal mass - spinal neoplasm

URATE crystal deposition in connective tissue and joints is a well known clinicopathological entity. Crystals can be found in cartilage, tendon sheaths, synovial fluid, and subcutaneous tissues. Gouty involvement of the spine is, however, infrequently recorded despite Tkach's clinical finding of cervical or lumbar pain in nearly 75% of patients with gouty peripheral arthritis. There is poor correlation of radiological and clinical findings in gout. We are presenting this case as a rare addition to the differential diagnosis of chronic erosive intraspinal masses.

Case Report

This 17-year-old girl was admitted with a 3-week history of intermittent pain and swelling of the left ankle. Painful swelling and erythema of the left index finger was a recent additional complaint. There was no history of trauma, fever, chills, rashes, or rheumatic fever. She had no urinary tract symptomatology. A back brace had been prescribed for back pain 3 years before this admission. Her father had suffered the onset of arthritis at the age of 25 years, and had died at 30 years of age of an undocumented cause. The patient's paternal grandmother and grandfather allegedly had crippling arthritis. Since the age of 8 years, the patient had been treated with Tridione (trimethadione) for an ill-defined partial, complex, non-focal seizure disorder. An electroencephalogram during this hospitalization revealed no abnormalities, and anticonvulsant therapy was discontinued without further episodes of seizure.

Examination. Physical examination disclosed no stigmata of neurofibromatosis. Synovial thickening of the left second metacarpal-phalangeal joint and left talo-tibial joint was present without erythema or limitation of motion. No abnormalities of the spine were noted. Peripheral muscle tone and strength were symmetric and normal. Sensory function was normal, as were the deep tendon reflexes.

Hemoglobin values ranged from 10.4 to 12.5 gm/100 ml, with hypochromic, microcytic red cell indices. Blood urea nitrogen was 34 mg/100 ml, serum creatinine 2.4 mg/100 ml, and uric acid 19 mg/100 ml. The patient's creatinine clearance was 32 ml/min, improving to 48 ml/min at the time of discharge from the hospital. Lupus erythematosus preparation, antinuclear antibody, and rheumatoid factor were non-reactive. Serum lead levels were normal, and a calcium EDTA (ethylene-diaminetetraacetate) infusion test ruled out chronic plumbism. The serum uric acid level was reduced to 9 mg/100 ml after administration of allopurinol. Renal tubular functions were depressed, including inulin, uric acid, and para-amino hippurate clearances. Twenty-four hour delta-aminolevulinic acid and porphyrin excretions were within normal limits.

A percutaneous renal biopsy revealed well preserved cortex, with significant medullary fibrosis. Electron microscopy of this specimen showed marked degenerative changes in the proximal convoluted tubules with absence or deformity of the brush border in most loops. Distal convoluted tubule cells demonstrated membrane-bound lipid vacuoles containing the degenerative pigment lipochrome. There
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FIG. 1. Left: Anteroposterior tomogram revealing a sharp symmetrical destruction of the pedicles of L-4 and L-5 (arrowheads). Right: Lateral tomogram demonstrating scalloping of the posterior vertebral bodies of L-4 and L-5 (arrows) and widening of the spinal canal in the anteroposterior diameter (arrowheads).

was a striking absence of lysosomes in the tubular endothelium. Characteristic changes of gouty nephropathy were not found.

Radiological examination of the skull, feet, ankles, and long bones was normal. A retrograde pyelography demonstrated small kidneys with normal pelvocalyceal systems and ureters. Voiding cystourethrogram showed a dilated urinary bladder without vesicoureteral reflux. Tomograms of the patient's lumbosacral spine revealed extensive changes which appeared chronic. The laminae, pars interarticularis, inferior articulating facets of L-4 and superior articulating facets of L-5 were destroyed by a symmetrical convex expansion. The pedicles of L-4 were sharply eroded along their inferior aspects (Fig. 1 left). Scalloping of the posterior margins of the L-4 and L-5 vertebral bodies was present; the spinal canal was expanded in the lower lumbar and upper sacral regions (Fig. 1 right). Lumbar myelography demonstrated an extradural defect impinging upon the lateral aspect of the theca at the L-4 level. The dura was not ectatic and the conus medullaris was normal in size and position. Occult spina bifida at the L-5 and S-1 levels was noted on previous x-ray films.

Operation. The left L-4 lamina was removed to permit adequate exploration of the lesion. Amorphous white material occupied the epidural space of the anterior and lateral spinal canal. This material was partially encapsulated by fibrous tissue and grossly infiltrated the bone and muscle in several areas. The mass extended from L-3 through L-5 on the left side, and was easily removed with a curette.

Samples of the amorphous material and surrounding tissue were submitted for histochemical studies and hematoxylin and eosin sections. Microscopic examination of the specimen showed granulomatous tissue with numerous multinucleated giant cells and histiocytes surrounding regions of acellular material. The latter was composed of hexagonal crystal aggregates (Fig. 2). Special stains confirmed that the crystalline material contained uric acid. The finding of a uric acid granuloma is consistent with the usual description of a gouty tophus.

Postoperative Course. The patient had an uncomplicated postoperative course and was discharged 1 week after the procedure. She continues to be free of pain and without neurological deficit nearly 2 years after the operation.

Discussion

Intraspinal deposition of urates is a rare occurrence despite widespread involvement of other anatomic sites by this process. Spinal cord or spinal root compression by tophi is even more uncommon. Table 1 lists six cases of confirmed spinal gout reported in the literature. All patients were adult males with longstanding peripheral gout. Spinal involvement was evenly distributed between the cervical, thoracic, and lumbar areas. Only two patients presented with com-
TABLE 1

Summary of seven cases of spinal urate deposition

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age, Sex</th>
<th>Symptoms</th>
<th>Neurological Examination</th>
<th>Involved Level</th>
<th>Uric Acid (mg/100 ml)</th>
<th>Blood Urea Nitrogen (mg/100 ml)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kersley, et al., 1950</td>
<td>24, M</td>
<td>cervical pain; peripheral arthritis; splenomegaly</td>
<td>normal</td>
<td>C1-2</td>
<td>2-16</td>
<td>40-53</td>
<td>died, pneumonia</td>
</tr>
<tr>
<td>Koskoff, et al., 1953</td>
<td>44, M</td>
<td>peripheral arthritis; urinary retention; paralysis, bilateral</td>
<td>paraplegia; hypertensive reflexes; sensory level T-11</td>
<td>T9-11</td>
<td>10.7</td>
<td>33</td>
<td>improved</td>
</tr>
<tr>
<td>Lichtenstein, et al., 1956</td>
<td>56, M</td>
<td>mental confusion; weakness; peripheral arthritis; femur fracture</td>
<td>normal</td>
<td>thoracolumbar</td>
<td>8.6-14.1</td>
<td>uremia</td>
<td>died, uremia</td>
</tr>
<tr>
<td>Hall &amp; Selin, 1960</td>
<td>51, M</td>
<td>dizziness; peripheral arthritis</td>
<td>normal</td>
<td>L4-S1</td>
<td>not available</td>
<td>69-172</td>
<td>died, uremia</td>
</tr>
<tr>
<td>Vinstein &amp; Cockerill, 1972</td>
<td>46, M</td>
<td>cervical pain; peripheral arthritis</td>
<td>normal</td>
<td>C3-4</td>
<td>13.2</td>
<td>not available</td>
<td>improved on medication</td>
</tr>
<tr>
<td>Litvak &amp; Briney, 1973</td>
<td>73, M</td>
<td>back pain, leg weakness; peripheral arthritis</td>
<td>paraparesis; absent reflexes absent cremasteries</td>
<td>L3-5</td>
<td>5-7.5</td>
<td>not available</td>
<td>improved, decompressive laminectomy</td>
</tr>
<tr>
<td>Wald, et al., 1979</td>
<td>17, F</td>
<td>peripheral arthritis</td>
<td>normal</td>
<td>L3-5</td>
<td>19</td>
<td>34</td>
<td>unchanged, laminectomy</td>
</tr>
</tbody>
</table>

FIG. 2. Photomicrograph of granulomatous tissue (g) composed of multinucleated giant cells (arrows) surrounding an amorphous core (am). Hexagonal crystalline forms (c) are demonstrated. H & E, X 400.
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pressive spinal symptoms, although pain was often noted at the affected level. Our patient is an adolescent female with extensive radiological changes but only mild symptomatology because of the low spinal lesion involving the cauda equina region. The extensive literature of gout documents a marked male preponderance in all age groups except the first decade. Treadwell found 55% females in the first decade of life, declining to 26% in the second decade.

A strong family history of gout is common in juvenile cases.

The frequent association of gout with altered renal function has led many authors to assume a causal metabolic relationship beyond that of mere deposition of urates in the tubules. Berger and Yu, however, found that hyperuricemia alone had no deleterious effect on renal function in 112 patients with gout who were followed for up to 12 years. Deterioration of renal function was incidentally ascribed to aging, renal vascular disease, non-urate renal calculi, pyelonephritis, or unrelated nephropathy. Four of the six patients with confirmed gout were uremic, and renal failure was the primary cause of death in two of these patients. Our patient manifests an ill-defined interstitial nephritis with subcellular tubular alterations. No evidence of gouty nephropathy could be found.

The underlying feature in the development of gout is hyperuricemia. Although the pathogenesis is still uncertain, monosodium urate salts presumably precipitate in supersaturated tissues. The crystals evoke an acute inflammatory response with migration of polymorphonuclear leukocytes to the area. In the process of phagocytosis, lysosomal hydrolytic enzymes are released. A chronic inflammatory reaction ensues, resulting in granuloma formation and tophus. The absence of lysosomal structures in the renal biopsy specimen of our patient is intriguing.

An important consideration following operative diagnosis in our patient is control of the responsible metabolic process. Suppression of serum urate levels with allopurinol, a xanthine-oxidase inhibitor, may arrest the pathological deposition of urates and result in regression of the gouty tophus.

Although focal back pain has been present intermittently in this patient, there is currently no evidence of instability or collapse of the lumbar spine.

References


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