Pigmented villonodular synovitis of the spine

Case report

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Pigmented villonodular synovitis commonly occurs in synovial joints of the appendicular skeleton, but rarely affects the synovial joints of the spine. It has both neoplastic and benign features, and the etiology is thought to be posttraumatic. The case of a young man presenting with paraparesis and a large thoracic lesion is reported.

KEY WORDS: pigmented villonodular synovitis; vertebral spine tumor

Pigmented villonodular synovitis is a well-characterized mass lesion of uncertain etiology associated with synovial membranes. It is found almost exclusively in the joints and tendon sheaths of the appendicular skeleton. This lesion has been associated with trauma and its clinical and histological features are consistent with reactive hyperplasia and neoplastic growth.

Pigmented villonodular synovitis is quite rare in the axial skeleton; only 10 cases have been reported previously in the English literature, none of which involved the thoracic spine. Reported cases involving the cervical and lumbar spine have been predominantly in females (80%), with presenting symptoms of either pain alone or pain with radicular neurological deficits. This paper reports an unusual case of severe acute paraparesis due to a thoracic intraspinal, extradural pigmented villonodular synovitis in a young man.

Case Report

This 23-year-old black man presented to the emergency department with a chief complaint of difficulty in ambulating. He had been involved in a motor-vehicle accident during the previous week, but had not appeared to suffer significant injuries. Two days prior to presentation, the patient had developed a diffuse low-back pain which intensified at the time of his emergency evaluation. He denied difficulty with bladder function, but did note patchy areas of sensory loss on both lower extremities.

Examination. The initial neurological examination demonstrated 5/5 segmental strength in the upper and lower extremities. Muscle bulk was bilaterally symmetrical but muscle tone in the lower extremities was increased. Deep-tendon reflexes were normal in the upper extremities and accentuated in the lower extremities. An upward toe-to-plantar stimulation was deli-

Fig. 1. A moderately T-weighted magnetic resonance image, sagittal view, of the thoracic spine showing the vertebral bodies of T4–10. A globular discrete mass of predominantly high signal intensity is apparent dorsal to the spinal cord at T7–8 (arrows).
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Fig. 2. Photomicrographs of the tumor specimen. Left: The hypercellular lesion is composed of mononuclear cells, giant cells, xanthoma cells, and moderate hemosiderin pigment. H & E, x 20. Right: Section demonstrating both a multinucleated giant cell and intracellular brown pigment. H & E, x 200.

...mitely noted on the right, with an equivocal response on the left. The patient’s gait was unsteady, with myelopathic features. The sensory examination showed a patchy loss of pinprick sensation in the lower extremities not involving the sacral dermatomes. No loss of proprioception or vibratory sensation was identified.

Plain thoracic spine films were unremarkable. A magnetic resonance (MR) image of the thoracic spine without contrast enhancement demonstrated a mass at the level of T7-8 with both extraspinal and intraspinal components (Fig. 1). The large intraspinal component had mildly eroded both ventrally and dorsally, but did not invade the vertebral bone, and the spinal cord was displaced to the left. No other lesions were identified in the thoracic region.

Operation. During the course of the evaluation the patient developed increased weakness of the lower extremities and urinary retention; therefore, an emergency operation was performed. The patient was placed in the prone position and a midline dorsal incision was used to expose the involved vertebral bodies. During exposure, a soft tumor mass invading the paraspinal musculature was identified. Obvious erosion with thinning of the T-7 and T-8 laminar arches was noted. A four-level bilateral laminectomy was performed with a drill,* following which the laminar arches spontaneously elevated due to a large grayish green multilobulated intraspinal mass. The mass was evacuated and a clean plane of dissection was identified along the dura and the eroded bone. The tumor also encased the head of the seventh rib, which was resected.

Postoperative Course. Postoperatively, the patient made an excellent recovery, with return of normal lower-extremity strength and sensation. His bladder and sexual function also returned to normal, although his gait continues to have mild myelopathic features. He has returned to his previous job.

Pathological Examination. Microscopic examination of the tumor demonstrated a hypercellular lesion composed of mononuclear cells, giant cells, xanthoma cells, and moderate hemosiderin pigment (Fig. 2). Five mitoses per high-power field could be identified in conjunction with a review of the radiographs; this was interpreted as a pigmented villonodular synovitis, a diagnosis that was confirmed by a nationally recognized authority in bone pathology at the Mayo Clinic. Since the lesion did not invade the vertebral body, giant-cell tumor of bone was excluded from the differential diagnosis.

Ploidy analysis of DNA by flow cytometric techniques was performed on paraffin-embedded tissue from the spinal lesion. A diploid population of cells was detected with a low-to-moderate proliferative index (that is, summation of cells in the synthetic (8.1%) and G2 + mitotic (6.2%) phases of the cell cycle).

Postoperative MR imaging of the thoracic spine was performed with and without contrast enhancement. This showed no residual disease (Fig. 3).

Discussion

Many terms have been applied to this lesion, including “fibrous histiocytoma of the synovium” and “nodular tenosynovitis.” The currently accepted term, however, is “pigmented villonodular synovitis,” as proposed by Jaffe, et al. The terminology has been expanded generally to account for two clinically and radiographically distinct entities: the sharply localized and diffuse forms.

Clinical Presentation

The presentation of pigmented villonodular synovitis in the appendicular skeleton involves the knee in 28% to 80% of cases. No clear gender predominance has been noted and it is typically diagnosed in the third decade of life. The most common presenting symptom is discomfort in the involved joint. The number of cases affecting the axial skeleton is small but the

* Drill with S1 bit and footplate manufactured by Midas Rex Institute, Fort Worth, Texas.
TABLE 1

Reported cases of pigmented villonodular synovitis of the spine

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Location</th>
<th>Sex, Age (yrs)</th>
<th>Pain</th>
<th>Major Presenting Neurological Deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kleinman, et al., 1980</td>
<td>C3-6</td>
<td>F, 65 yes</td>
<td>subacute myelopathy</td>
<td>none</td>
</tr>
<tr>
<td>Campbell &amp; Wells, 1982</td>
<td>L4-5</td>
<td>F, 54 yes</td>
<td>none</td>
<td>L4-5 radicular hypealgesia, weakness</td>
</tr>
<tr>
<td>Savitz, et al., 1982</td>
<td>L4-5</td>
<td>F, 53 yes</td>
<td>none</td>
<td>L-5 radicular hypealgesia</td>
</tr>
<tr>
<td>Pulitzer &amp; Reed, 1984</td>
<td>C1-4</td>
<td>F, 35 no</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>Palitzer &amp; Reed, 1984</td>
<td>L5-S1</td>
<td>M, 23 no</td>
<td>none</td>
<td>L-5 radicular hypealgesia</td>
</tr>
<tr>
<td>Weidner, et al., 1986</td>
<td>L5-6</td>
<td>F, 48 yes</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>Retrum, et al., 1987</td>
<td>L5-S1</td>
<td>F, 34 yes</td>
<td>none</td>
<td>L-5 radicular hypealgesia</td>
</tr>
<tr>
<td>Karnezis, et al., 1990</td>
<td>C-7</td>
<td>F, 37 yes</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>Clark, et al., 1993</td>
<td>T7-8</td>
<td>M, 23 yes</td>
<td>acute paraparesis</td>
<td>none</td>
</tr>
</tbody>
</table>

epidemiology consistently deviates from that described above (Table 1); these patients have been predominantly women in the fourth to sixth decades of life. The presenting chief complaint is almost always pain, but root and cord deficits were also present in half of the cases. Our case of pigmented villonodular synovitis of the spine is unusual in that the patient is a young man who presented with acute paraparesis and a minimal amount of pain.

Etiology

The etiology of pigmented villonodular synovitis is poorly understood. Trauma is often mentioned, but there are no data that statistically demonstrate a causal relationship between these lesions and injury. It has been noted that hemophilic and Charcot joints fail to develop pigmented villonodular synovitis. Attempts to induce this pathological entity in animal models with trauma alone have failed. Our patient denied even a remote episode of significant back trauma and it is very unlikely that the low-velocity motor-vehicle accident he sustained 1 week prior to presentation caused the lesion, although it may have “destabilized” the pathology or caused new hemorrhage into it. A neoplastic etiology of pigmented villonodular synovitis, although long suspected, remains controversial and has recently been carefully reviewed by Weidner, et al. Our case showed a diploid population of cells, which supports a benign diagnosis. There was also a low-to-moderate percentage of cells in the synthetic and G2 + mitotic phases of the cell cycle. It is likely that the low-to-moderate proliferative index indicates a low-to-moderate potential for recurrence. However, further retrospective studies are needed to verify this hypothesis.

Proliferative Potential

Although there is a great deal of literature concerning the behavior of pigmented villonodular synovitis, no previous studies have been published on the deoxyribonucleic acid (DNA) ploidy status and proliferative index as it relates to the potential for recurrence. Preliminary follow-up information indicates that, overall, 45% of patients develop local recurrence. For the most part, these lesions should be regarded as locally aggressive but nonmetastasizing lesions. Heretofore, no one has commented on DNA analysis and whether a high proliferative index indicates a greater propensity for recurrence. Our case showed a diploid population of cells, which supports a benign diagnosis. There was also a low-to-moderate percentage of cells in the synthetic and G2 + mitotic phases of the cell cycle. It is likely that the low-to-moderate proliferative index indicates a low-to-moderate potential for recurrence. However, further retrospective studies are needed to verify this hypothesis.

Neuroradiographic Studies

The radiographic features of pigmented villonodular synovitis have been characterized primarily in the extremities. Plain films demonstrate erosive bone changes and occasionally a definable soft-tissue mass. Other findings on plain films include increased synovial density due to hemosiderin deposition and multiple subchondral cysts. On bone scanning the bone lesions take up radionuclide, and on angiography larger lesions show numerous irregular vessels with a blush. Computerized tomography demonstrates the sclerotic margins of the lytic bone lesions, and often the soft-tissue mass will enhance at least at its periphery. Magnetic resonance imaging, especially with T2-weighted images, clearly defines the margins of the lesion. The mass usually has mixed signal intensity on T1 and T2 pulse sequences due to the presence of hemosiderin, cystic fluid, and hemorrhage.
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Magnetic resonance imaging is very sensitive but not specific; however, it represents the best single diagnostic test for detecting pigmented villonodular synovitis and separating nodular from diffuse lesions. Patients can be followed longitudinally with MR imaging for evidence of recurrence.

Treatment Considerations

The primary consideration that guides therapeutic intervention for this tumor is its tendency for local recurrence.\(^2\)\(^,\)\(^3\)\(^,\)\(^22\)\(^,\)\(^23\) Tumor-free survival of 65 years in the appendicular skeleton has been reported.\(^2\)

Surgical excision is curative if the entire lesion can be resected.\(^1\)\(^,\)\(^2\)\(^,\)\(^22\)\(^,\)\(^23\) This is the only form of curative therapy and every effort should be made to achieve a gross total excision at the time of the first operation. This is technically easier with discrete lesions and is very difficult with large diffuse lesions. Spinal lesions may require a staged approach to resection from the ventral and dorsal regions if disease is widely distributed. In the spinal cases reported, early recurrence was noted when total excision was not possible.\(^2\)\(^,\)\(^3\) In our case, the bone was eroded ventrally but was not frankly invaded, and the lesion peeled easily from the ventral aspect of the vertebral column, allowing for gross total excision.

The role of radiation therapy has not been clearly defined. Local irradiation has been described as a primary treatment since the 1940's.\(^7\)\(^,\)\(^12\) Although encouraging results were reported with this approach, the follow-up time was short. Potential problems including radiation-induced sarcomas, joint stiffness, and poor wound healing have been recognized. The use of radiation therapy as an adjunct either after subtotal resection or after local recurrence has been described, but no definitive data about efficacy are available.\(^1\)\(^,\)\(^2\) The use of radioisotope infusions has also been reported.\(^1\)

Repeat surgical excision of pigmented villonodular synovitis is a well-recognized form of therapy and may be very applicable to spinal lesions where there is an additional risk of radiation-induced neurological injury. However, early and multiple recurrences may necessitate local radiation treatment if repeat surgical resection is not possible.

References

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