Outcome in neurologically impaired patients with craniovertebral junction tuberculosis: results of combined anteroposterior surgery

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Object. The authors studied the immediate and long-term outcome after transoral decompression, occipitocervical fusion, and antituberculous therapy (ATT) in patients who had neurological deficits due to craniovertebral junction (CVJ) tuberculosis.

Methods. In this retrospective study, the authors reviewed the management and outcome in nine consecutive patients in whom features of spinal cord compression were observed and CVJ tuberculosis was diagnosed between 1993 and 1999. They ranged in age from 9 to 55 years. Onset of symptoms was acute or subacute and rapidly progressive (median 4 months, range 1–12 months). Patients presented with neck pain (89%), progressive limb weakness (89%), sensory symptoms (22%), and urinary dysfunction (33%). The mean preoperative functional grade based on the Nurick Scale was 3.4 (range 1–5). The disease caused reducible atlantoaxial dislocation (AAD) in three patients (33%), irreducible AAD in two (22%), basilar impression (BI) in one (11%), AAD with BI in one (11%), and C-2 vertebral body (VB) destruction without dislocation in two (22%). Surgery was performed in all cases. Five patients (56%) underwent transoral odontoidectomy, two (22%) transoral decompression of retropharyngeal abscess and granulation tissue, and two (22%) transoral decompression of abscess and diseased parts of the C-2 VB. All patients then underwent occipitocervical (occiput–C3) fusion in which a contoured Steinmann pin and iliac bone grafts were used. Postoperatively, ATT was prescribed for 18 months. In the immediate postoperative period, function in these patients improved from a mean Nurick grade of 3.4 to 2.3 (p < 0.01). At long-term follow-up examination (median 18.8 months, range 7–46 months) function improved from a mean Nurick grade of 3.4 to 0.3 (range 0–2) (p < 0.001).

Conclusions. Patients with CVJ tuberculosis with features of cervical myelopathy are ideally managed with transoral decompressive procedures followed by occipitocervical fusion because this therapy provides immediate neurological improvement, stability, and allows early mobilization. The long-term prognosis in patients with this disease is excellent provided it is treated with appropriate surgical intervention(s) and with adequate duration of ATT.

KEY WORDS • cervical spine • craniovertebral junction • spinal cord • tuberculosis

Craniovertebral junction tuberculosis is a rare form of spinal tuberculosis accounting for less than 1% of all forms of spinal tuberculosis.18 Lifeso12 documented only 15 patients with CVJ tuberculosis in a total of 5393 patients (0.3%) with spinal tuberculosis. He reported that CVJ tuberculosis was observed in 12 (0.48%) of 250 adults with spinal tuberculosis. Craniovertebral junction tuberculosis primarily involves the atlas and the axis but can also involve the lower end of the clivus and the occipital condyles. Whereas tuberculous infection has been a frequent and prominent cause of cervical cord compression in developing countries, it is increasingly reported in individuals from developed regions of the world.1,6,11,16,22 Patients with CVJ tuberculosis may or may not present with neurological impairment. While conservative management may be appropriate in patients without evidence of spinal cord compression, those in whom neurological deficits have manifested might benefit from prompt surgical intervention. In a previous study,10 we found that a good outcome was possible in patients with CVJ tuberculosis who underwent different forms of surgery. We suggested, however, that patients with features of spinal cord compression should undergo anterior decompression followed by posterior fusion, to ensure immediate neurological improvement and stability. We have since adopted this protocol, and we present the results of this combined approach in nine patients.

Clinical Material and Methods

We recorded data obtained in patients treated in our unit between 1993 and 1999 who presented with cervical myelopathy and in whom CVJ tuberculosis was diagnosed. We analyzed the clinical and radiological features, microbrial findings, management practices, and the immediate and long-term outcome in these patients.
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Nine patients with cervical myelopathy and CVJ tuberculosis were managed in our unit between 1993 and 1999. Clinical data are summarized in Table 1. Age ranged from 9 to 55 years. There were eight male and one female patients. The duration of symptoms ranged from 1 to 12 months (median 4 months). Patients presented with neck pain (89%), progressive limb weakness (89%), sensory symptoms (22%), and bladder dysfunction (33%). Three patients suffered systemic symptoms of tuberculosis such as fever and weight loss. In all patients evidence of pyramidal tract involvement was documented; involvement of the sensory tracts was observed in five patients. In one patient with no complaints of limb weakness there was clinical evidence of pyramidal tract involvement. One 9-year-old boy initially presented with a large retropharyngeal abscess and no evidence of neurological impairment. The abscess was drained transorally, and ATT was initiated. He returned 3 months later with a spastic quadriplegia.

**Clinical Features**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Duration of Symptoms (mos)</th>
<th>Signs</th>
<th>Nurick Grade</th>
<th>Systemic Symptoms</th>
<th>Imaging Study</th>
<th>Imaging Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>17, M</td>
<td>6</td>
<td>pyramidal, dorsal column, spinothalamic</td>
<td>4</td>
<td>fever</td>
<td>x-ray</td>
<td>reducible AAD</td>
</tr>
<tr>
<td>2</td>
<td>24, M</td>
<td>1</td>
<td>pyramidal, spinothalamic</td>
<td>5</td>
<td>none</td>
<td>x-ray</td>
<td>reducible AAD</td>
</tr>
<tr>
<td>3</td>
<td>55, M</td>
<td>5</td>
<td>pyramidal, spinothalamic</td>
<td>4</td>
<td>none</td>
<td>x-ray</td>
<td>irreducible AAD + BI</td>
</tr>
<tr>
<td>4</td>
<td>55, M</td>
<td>3</td>
<td>pyramidal, dorsal column, spinothalamic</td>
<td>5</td>
<td>weight loss</td>
<td>x-ray, MR</td>
<td>reducible AAD</td>
</tr>
<tr>
<td>5</td>
<td>25, M</td>
<td>6</td>
<td>pyramidal</td>
<td>1</td>
<td>none</td>
<td>x-ray, MR</td>
<td>BI</td>
</tr>
<tr>
<td>6</td>
<td>35, F</td>
<td>4</td>
<td>pyramidal, dorsal column, spinothalamic</td>
<td>5</td>
<td>fever, night sweats</td>
<td>x-ray, myelography</td>
<td>C-2 VB disease</td>
</tr>
<tr>
<td>7</td>
<td>16, M</td>
<td>1</td>
<td>pyramidal</td>
<td>4</td>
<td>none</td>
<td>x-ray, MR</td>
<td>C-2 VB disease</td>
</tr>
<tr>
<td>8</td>
<td>9, M</td>
<td>5</td>
<td>pyramidal</td>
<td>2</td>
<td>none</td>
<td>x-ray, CT</td>
<td>irreducible AAD</td>
</tr>
<tr>
<td>9</td>
<td>18, M</td>
<td>12</td>
<td>pyramidal</td>
<td>1</td>
<td>none</td>
<td>x-ray</td>
<td>irreducible AAD</td>
</tr>
</tbody>
</table>

**Imaging Studies**

All patients underwent dynamic plain radiography and tomography of the CVJ. In all except one patient (Case 2), the plain radiographs and tomograms revealed an enlargement of the prevertebral soft-tissue shadow in front of the C1–2 vertebrae (Fig. 1). A diagnosis of AAD was rendered when the atlantoaxial distance was more than 3 mm in adults and 4 mm in children on a lateral x-ray film of the CVJ. The presence of BI was diagnosed by drawing the McGregor line and clival canal line on plain radiographs of the CVJ. In one patient who presented with history of trauma, neck pain, and no neurological impairment, findings on an initial lateral radiograph of the cervical spine were interpreted as normal (Fig. 2 upper). Three months later he presented with progressive quadriparesis, at which time a lateral radiograph of the cervical spine revealed AAD and BI (Fig. 2 lower). In addition to plain radiography, myelography was performed in one patient, CT scanning in one, and MR imaging in three patients (Figs. 3 and 4). Based on plain radiography alone in four patients and plain radiography and other imaging studies in the other five patients, the following diagnoses were made: AAD in five patients (55%), BI in one (11%), AAD with BI in one (11%), and C-2 VB destruction without dislocation in two (22%). The AAD could not be reduced on neck extension in three of the six patients with AAD (two with AAD and one with AAD and BI). In one patient with pure BI and two with C-2 VB disease radiography demonstrated dynamic spinal stability. Chest radiography revealed evidence of tuberculosis in only one. None was in an immunocompromised state. Testing for human immunodeficiency virus infection was negative in all patients.

**Operative and Nonoperative Management**

The goals of surgery were to provide a diagnosis, obtain tissue or pus sample for culture studies, decompress the cervical cord, and ensure immediate stabilization of the CVJ (Table 2). Preoperatively, skull traction was undertaken in three patients in poor clinical grade. A transoral soft-tissue decompression and odontoidectomy was performed in five patients. The AAD was irreducible in two of these patients, in one BI and AAD were present, in one BI was documented, and in one with reducible AAD the odontoid base was destroyed and the odontoid was freely mobile. In two other patients with a reducible AAD, only transoral decompression of the soft tissue was performed. In two patients with C-2 involvement, transoral soft-tissue decompression was combined with removal of the diseased parts of the C-2 VB. In none of the cases was any attempt made to denude the dural tube of granulation tissue. Following the anterior decompression, all patients underwent posterior occipitocervical (occiput–C3) fusion in which a contoured Steinmann pin (loop or rectangle) and iliac bone graft were placed, either during the same stage and thus same period of anesthesia (five patients) or at a separate sitting 1 week later (four patients). Even patients in whom stability was demonstrated at presentation (one with pure BI and two with C-2 VB disease) were considered to be potentially at risk for CVJ stability after the transoral procedure. Furthermore, there was a possibility that CVJ instability would develop in these patients over time. Therefore, these patients underwent posterior fusion after the transoral surgery. Patients in whom staged procedures were performed were maintained in skull traction after the transoral procedure. They were mobilized the day after the occipitocervical fusion, in a hard cervical...
or Philadelphia collar, and were advised to continue wearing the collar for a period of 6 months.

All patients underwent ATT for an 18-month period (isoniazid 5–10 mg/kg/day for 18 months; rifampin 10–15 mg/kg/day for 18 months; and ethambutol 15 mg/kg/day for the first 3 months). The pus, tissue, and bone samples obtained at surgery were submitted for microbiological and histopathological studies. Smear or culture of the pus or tissue was positive for acid-fast bacilli in three patients (33%). Histopathological examination of the bone and/or tissue showed tuberculous osteomyelitis or granulation tissue in seven patients and chronic osteomyelitis in two patients.

Statistical analysis was conducted using the paired t-test for correlating observations.

**Results**

**Surgery-Related Morbidity**

In one patient transient confusion and urinary incontinence developed 1 day after occipitocervical fusion. He was found to be dehydrated and anemic, and improved with intravenous fluid replacement and blood transfusion. In one patient infection of the iliac bone graft donor site developed and was healed by application of daily dressings. There was no spinal cord–related morbidity or mortality in this series.

**Early and Long-Term Results**

At discharge from the hospital (10–14 days) following the second surgery, functional improvement of almost one Nurick grade was demonstrated in all patients. The mean Nurick grade at discharge improved from 3.4 to 2.3 ± 1.1 (range 1–4) (p < 0.01). At long-term follow-up examination (7–46 months, mean 18.8 months) further improvement was reflected by the mean Nurick grade of 0.3 ± 0.4 (range 0–2) (p < 0.001). All patients underwent plain radiography of the CVJ at follow up. Good incorporation of the bone graft was documented in all patients (Fig. 5). There was some degree of kyphotic deformity at the C1–2 level in two patients.

**Discussion**

Tuberculosis of the vertebral column accounts for about 6% of extrapulmonary disease or approximately 5% of all disease caused by *Mycobacterium tuberculosis*. In the spine, tuberculosis commonly affects the thoracolumbar regions whereas cervical tuberculosis is uncommon. Tuberculosis of the atlantoaxial region is the least common form of spinal tuberculosis. Most published series comprise isolated cases or a few patients. The
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commonest form of CVJ tuberculosis, AAD, was first described by Hilton.14

Pathophysiological Mechanisms

Craniovertebral junction tuberculosis leads to AAD by two mechanisms (osteomyelitis and destruction of the ligaments, mainly the transverse and alar).13,14 Lifeso12 has reported three stages of CVJ tuberculosis based on findings on plain x-ray films. In Stage I there is no evidence of bone destruction or displacement, whereas in Stages II and III there is progressively increasing evidence of bone destruction and AAD. Lifeso recorded one patient in whom the disease had progressed from Stage I to III. We also treated one patient in whom the disease progressed from Stage I to III despite provision of ATT. Based on his observations, Lifeso suggested that CVJ tuberculosis results from infection that spreads from the retropharyngeal tissues to the osseous elements; primary infection of the atlas or axis was rarely the cause of disease.

Clinical Features

Only between 30 and 70% of patients with CVJ tuberculosis have been reported to present with neurological deficits. Early-onset symptoms may be mild and nonspecific, and there may be associated features such as malaise, nocturnal increase in body temperature, or weight loss.9 Because there are no localizing symptoms or signs, a delay in the diagnosis of CVJ tuberculosis can occur.6,9,10,11 Most patients (> 90%) present with severe neck pain and restriction of neck movements. Torticollis, dysphagia, and dysphonia due to a large retropharyngeal abscess may also be present. Sudden death has been associated with tuberculosis of the atlas and axis, probably secondary to unrecognized instability.4 Craniovertebral junction tuberculosis has been reported in immunocompromised patients such as those with acquired immune deficiency syndrome.20 Although others4,12 have reported a high incidence of extraspinal tuberculosis in patients with CVJ tuberculosis, we found evidence of systemic tuberculosis in only one of our cases.

Imaging Studies

Most often a diagnosis of CVJ tuberculosis is evident on a lateral plain radiograph of the CVJ. The combination of erosion of the atlas or axis, AAD, and increased prevertebral soft tissue is almost pathognomonic of CVJ tuberculosis in an endemic region. The use of CT provides additional details of the soft-tissue abnormality, focal bone erosion, and sclerosis.11 Fragmented, osteolytic, and subperiosteal, localized sclerotic bone lesions are features of bone involvement in cases of spinal tuberculosis.5 The use of MR imaging is the ideal diagnostic procedure to ascertain the presence and extent of compromise of the spinal canal and cord compression, as well as to determine the size of the perispinal abscess if present.17 Basilar impression has not been frequently reported in combination with CVJ tuberculosis;2 however, BI was present in one of our patients, and BI associated with AAD was present in another.

Management and Outcome

In initial reports on CVJ tuberculosis the authors recommended conservative measures that included bedrest
for 6 to 9 months and long-term antibiotic therapy. Tuli recommended performing surgery only in patients with persistent or recurrent neurological deficits and in those with CVJ instability. Other authors have recommended a variety of anterior and posterior procedures in patients with CVJ tuberculosis. Management strategies for patients with CVJ tuberculosis and neurological impairment reported by various authors are summarized in Table 3.

Most authors have advocated drainage of the extradural abscess and removal of diseased bone and ligaments, but none has recommended odontoidectomy. We found that the AAD was irreducible in three of six patients with AAD (two with AAD and one with BI and AAD). Because the odontoid was the compressing element in two patients with AAD and two with BI, an odontoidectomy was required to achieve decompression of the cervico-medullary junction. In one patient with reducible AAD, the odontoid was resected because it was completely detached from the C-2 VB and was freely mobile. We believe that removal of the odontoid when it is the compressing element will lead to early neurological recovery. Although we did not routinely use skull traction preoperatively, we believe that even in patients in whom the AAD is partially reduced following traction, an odontoidectomy will provide immediate decompression. Only in those in whom the AAD is completely reduced following traction would a strategy of posterior fusion alone combined with anterior soft-tissue decompression be an valid option.

The transoral procedure and the posterior fixation can be performed in one sitting, as was accomplished in the initial five patients in our series. Such surgery, however, can be protracted and we believed that in patients who are in poor clinical grade the posterior fixation can be staged 1 week after the transoral surgery. Thus, later in our series we performed staged surgery. Edwards, et al., have argued that a longer delay of approximately 3 months between the two procedures would enable the team of physicians to obtain sensitivity results of the causative organism and modify the ATT accordingly. They recommended that the patient be immobilized in a halo vest until the second surgery. We believe that this delay is not warranted because cultures might not grow the M. tuberculosis in all cases; culture was positive in only one third of our patients. Because the disease is anteriorly located in most cases, the posterior fusion should be effective even in patients with drug-resistant tuberculosis. By performing the occipitocervical fusion within 1 week of the transoral surgery, the patient is also spared the cost and possible morbidity associated with halo vest immobilization.

Although there is a trend toward conservative and ambulatory therapy in patients with thoracolumbar tuberculosis, the same cannot be recommended for those with cervical or CVJ tuberculosis because of the risk of sudden and permanent spinal cord damage due to instability of the cervical spine. Prolonged bedrest, which is often a component of conservative therapy, can lead to complications such as decubitus sores, pulmonary infection, and deep vein thrombosis, especially if the patients are quadriplegic. Therefore, internal stabilization of the CVJ so that the patient can be mobilized soon after surgery seems a prudent strategy to avoid all complications of prolonged bedrest. Internal fixation of the occipitocervical region also obviates the need for a halo brace. A hard cervical or

**TABLE 2**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Traction</th>
<th>Operation*</th>
<th>Staged Ops</th>
<th>Outcome (Nur-ick Grade)</th>
<th>Follow-Up Duration (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>yes</td>
<td>transoral drainage, Oc–C3 fusion</td>
<td>no</td>
<td>0</td>
<td>24</td>
</tr>
<tr>
<td>2</td>
<td>yes</td>
<td>transoral drainage, Oc–C3 fusion</td>
<td>no</td>
<td>0</td>
<td>20</td>
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<td>3</td>
<td>no</td>
<td>transoral odontoidectomy, Oc–C3 fusion</td>
<td>no</td>
<td>2</td>
<td>46</td>
</tr>
<tr>
<td>4</td>
<td>yes</td>
<td>transoral odontoidectomy, Oc–C3 fusion</td>
<td>no</td>
<td>0</td>
<td>9</td>
</tr>
<tr>
<td>5</td>
<td>no</td>
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<td>no</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>6</td>
<td>no</td>
<td>C-2 decompression, Oc–C3 fusion</td>
<td>yes</td>
<td>0</td>
<td>7</td>
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<tr>
<td>7</td>
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<td>0</td>
<td>13</td>
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<tr>
<td>8</td>
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<td>yes</td>
<td>0</td>
<td>13</td>
</tr>
<tr>
<td>9</td>
<td>no</td>
<td>transoral odontoidectomy, Oc–C3 fusion</td>
<td>yes</td>
<td>0</td>
<td>30</td>
</tr>
</tbody>
</table>

* Occipitocervical fusion performed using contoured metal loop or rectangle and iliac bone graft.

Fig. 5. Case 4. Follow-up lateral radiograph of the cervical spine obtained 9 months after transoral odontoidectomy and occipitocervical fusion. Note the good incorporation of the bone graft.
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### TABLE 3

**Summary of reported series of patients with CVJ tuberculosis and neurological impairment**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Management*</th>
<th>Outcome</th>
<th>Follow-Up Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tuli, et al., 1974</td>
<td>8</td>
<td>traction &amp; drainage of abscess &amp; bedrest for (\leq 6) mos</td>
<td>good in 7; fair in 1</td>
<td>1–7 yrs</td>
</tr>
<tr>
<td>Fang, et al., 1983</td>
<td>4</td>
<td>transoral drainage &amp; anterior/posterior fusion + bedrest for 3 mos</td>
<td>good in 3; worsened &amp;</td>
<td>mean 9.25 yrs</td>
</tr>
<tr>
<td>Lifeso, 1987</td>
<td>5</td>
<td>transoral decompression + halo traction + posterior fusion</td>
<td>good in 4; preop death in 1</td>
<td>mean 36 mos</td>
</tr>
<tr>
<td>Lal, et al., 1992</td>
<td>6</td>
<td>transoral drainage &amp; Minerva jacket (3 cases); posterior</td>
<td>good in 5; death in 1</td>
<td>mean 24.8 mos</td>
</tr>
<tr>
<td>transoral drainage &amp; posterior decompression &amp; metal &amp; bone fusion (1 case)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>present series</td>
<td>2</td>
<td>transoral drainage (1 case); transoral drainage &amp; traction (1 case)</td>
<td>good in 1; death in 1</td>
<td>12 &amp; 18 mos, respectively</td>
</tr>
<tr>
<td>Kaanan, et al., 1999</td>
<td>9</td>
<td>transoral odontoidectomy/decompression &amp; occipitocervical fusion</td>
<td>good in 9</td>
<td>mean 18.8 mos</td>
</tr>
</tbody>
</table>

* All patients underwent antituberculous chemotherapy.

Philadelphia collar will provide adequate orthotic support and is generally recommended for a period of 3 to 6 months, which is the time required for the iliac bone graft to fuse the occiput to the upper cervical spine.

Most authors have reported an excellent neurological outcome in almost all patients with CVJ tuberculosis (Table 3). The good outcome is maintained over several years after the treatment. Death, however, has been reported in a few patients who presented late in their illness, underlining the importance of early management of this potentially lethal condition.

### Conclusions

Acute or subacute onset of neck pain with restricted neck movements and features of progressive spinal cord compression should arouse suspicion of a diagnosis of CVJ tuberculosis, which can be confirmed by imaging studies and an elevated erythrocyte sedimentation rate. Patients with CVJ tuberculosis in whom myelopathy is present are ideally managed with transoral decompressive procedures followed by posterior internal stabilization because this provides immediate neurological improvement, immediate spinal stability, and excellent long-term outcome.

### References


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