Craniovertebral junction tuberculosis with Subluxation

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Abstract
Tuberculosis of the Craniovertebral junction is a rare and potentially dangerous condition. Neurological complications are more serious and common than with that of other vertebrae. Hence the need for early recognition and appropriate treatment. Management of craniovertebral junction tuberculosis has been an ever debatable topic of discussion, varying from purely conservative to surgical interventional protocols with no proper consensus. Presenting here 10 cases of occipitocervical tuberculosis with destruction of the occipital condyle with subluxation with or without retropharyngeal abscess formation, managed surgically by posterior occipito-cervical stabilization and fusion with a review of literature.

Keywords: Tuberculosis, Cervical spine, Craniovertebral junction, Subluxation, Occipito cervical fusion

1. Introduction
Tuberculosis of the craniovertebral junction is an uncommon entity, comprising of 0.5% of tuberculosis in general, 6% of extra-pulmonary tuberculosis \(^1, 2\) and 0.3% to 1% of allspinal tuberculosis \(^2, 3, 4, 5\). Clinical presentation can vary from simple neck pain to severe neurological complications like quadriplegia. Early diagnosis in terms of radiological methods and treatment have shown excellent prognosis. Treatment options vary from conservative to surgical interventions, due to lack of adequate consenses it has ever been a debatable topic for discussion.

Fig 2: CT Shows bony erosions of the atlas, occipital condyles. With Atlantoaxial Subluxation.
Fig 3: left: sagittal T2-weighted MR image of the CVJ, revealing destruction of the odontoid base with atlantoaxial Subluxation with out cord compression. Right: sagittal T2-weighted MR image showing prevertebral hyperintensity suggestive of abscess.

Fig 4: intra operative fixation

Fig 5: C–Arm image after stabilization

Fig 5: pot op x-rays at 12 months and 24 months
Patients and management
All patients admitted with clinico-radiological diagnosis of Craniovertebral tuberculosis without neurological symptoms were considered, a total of 10 patients were treated (8 men, 2 women). Their age ranging from 28 to 60 years. Only 4 patients had received anti-tuberculosis therapy before presentation. All were treated previously with pain relieving medication, local heat and physiotherapy.

Patients presented with symptoms of neck pain, occipital headache, most had pain which was continuous in nature, non radiating, aggravated at night and on movements of the head. Retropharyngeal abscesses of varying size were present in six patients, symptomatic dysphagia occurred in only three. All had constitutional symptoms such as fever, loss of weight and loss of appetite, while two patients had vomiting as a prominent symptom. One patient kept his neck supported by both hands at all times. On examination an axial-loading force or pressure applied over the head elicited pain at the affected site in all. No patients presented with weakness of limbs or involvement of the lower cranial nerves, bladder and bowel functions.

All the patients had a full blood count, and the ESR ranged from 50 to 110 mm/hr. They all had radiographs of the cervical spine and of the chest to identify the primary pulmonary focus. Plain radiographs of the cervical spine in six patients showed prevertebral bulge of 6 to 8 mm at the C1-C2 region, with varying degrees of bone destruction. Evidence of atlantoaxial instability was present in all patients. All patients had a mobile, reducible atlantoaxial dislocation (AAD) which was confirmed on carefully performed flexion and extension views. MRI of the cervical spine was carried out in all patients and established the diagnosis of tuberculosis by revealing the abscesses and the extent of cord compression, and by identifying the extent of destruction of the bone. No patient was subjected to abscess drainage. All patients were managed with anti-tuberculous drugs and by rigid immobilisation using an occipitomental collar before and after surgery. The drug treatment was continued for at least one year. The collar was worn post operatively until there was clinical and radiological evidence of healing, the average duration being four to six months.

Anti-tuberculous treatment consisted of standard doses of four drugs; isoniazid (5 to 10 mg/kg/per day), rifampicin (10 to 20 mg/kg/per day), ethambutol (15 mg/kg/per day) and pyrizanamide (25 mg/kg/per day), with vitamin supplements. Pyrizanamide was discontinued after four months and the others were continued for one year. All patients were subjected to occipitocervical fusion and stabilisation in the anatomical position All patients were kept in bed for the first five days, with sandbags on each side of the head to prevent rotation. Elevation of the head end of the bed was started from the third day and on the sixth the patient began walking with a Philadelphia collar. These patients were discharged on 10th post operative day. The collar was used until there was clinical and radiological evidence of healing, usually at between 16 and 20 weeks. The minimum follow-up period in these patients was 12 months and the maximum was 24 months.

Results
All patients treated surgically had excellent relief of pain. Fusion was achieved at 16 to 20 weeks, as determined by consolidation of the bone graft on plain radiographs. No patient suffered reactivation of the disease after completion of chemotherapy. No patient had deterioration of their neurological status after surgery.

Discussion
Tuberculosis has been an important health problem in the developing countries since time memorial. Craniovertebral (CV) junction as defined by Wackenheim, is the area bounded superiorly by a line joining the internal occipital protuberance to the sphenoid occipital synchonosis and inferiorly the lower limit of the body of axis [1]. Due to lack of localizing symptoms or signs there is a delay in the diagnosisis is considered an extremely rareand potentially dangerous condition [1, 3]. With the resurgence of tuberculosis and theavailability of MRI, CT scan the incidence is now estimated to be about 32% [1]. It is more commonly seen in the immunocompromised (AIDS/Transplant /Malignancy). The disease has an insidious onset, that usually presents with neck pain, very commonly misdiagnosed in its early stages as cervical spondylitis, myofascitis etc [4].

Other common features are dysphagia, dysphonia, torticollis, airway compromise, tongue deviation, cervico-medullary compression, cranial nerve deficits, atlanto-axial instability, upward translocation of the dens, and abscess formation causing nerve root compression [1, 3]. Neurological deficit is present in 24% to 64% of patients [1] with spastic quadiplegia/quadriplegia 60% [6] to 100 %, [7] neurological deficits are more common when ligaments and bone are involved [2]. It commonly affects youngindividuals, ranging from 14 to 65 years. Early diagnosis with radiologically investigation is recommended. Xray changes appear late 2 to 6 months as 50% destruction is need to be detected [2]. If left unattended to, there is a high probability of irreversible involvement of neurovascular structures and grave prognosis. Management of CV junction tuberculosis has been an ever debatable topic of discussion with no proper consensus, treatment varies from purely conservative to operative interventional protocols with their own pros and cons.

Treatment options being [3]
1. Antitubercular therapy (ATT) only
2. Cervical Traction with ATT
3. Trans oral decompression and posterior fusion with ATT
4. Posterior fusion with out decompression with ATT

The main features that decide on the treatment choice are the neurological status, extent of bony destruction, cord compression, presence of Subluxation /dislocation and clinical response to ATT.

Conservative management with prolonged bed rest and skull traction as the treatment of CV junction tuberculosis with atlanto-axial subluxation has been supported by many authors even in advanced stage [8]. Unstable spine with atlanto-axial subluxation or those with occipital neuralgia can be initially managed with a cervical traction and then later changed to a rigid cervical collar for 6 months to 9 months [3, 9, 10]. Conservative management comes with the risk of sudden and permanent spinal cord damage due to instability [4, 10, 11] and other complications such as decubitus sores, pulmonary infection, and deep vein thrombosis.

Transoral drainage of abscess alone for decompression is recommended in patients with mild bony changes and no atlantoaxial dislocation [2], patients with instability and neurological compromisesshould be stabilised by means of a posterior fusion [11, 12], whether to be done in the same sitting with fusion or as a staged procedure is controversial. Edward et al., [13] suggested 3 months between the two procedures with halo vest after the drainage as it would allow for
sensitivity and adequate ATT regimens, never the less they can be performed in the same sitting \cite{10} or with a few weeks interval as cultures may not show growth of M. Tuberculosis \cite{10}, both studies showing adequate results. Decompression by removal of the odontoid is rarely done and is recommended when it is the compressing element \cite{10}. An alternative decompression is the anterior transcervical retropharyngeal approach \cite{2}.

Based on the extent of disease, disability and neurological deficit Lifeso, et al., staged the disease and has outlined the treatment accordingly which is widely accepted to date \cite{14}.

Table 1: Management protocols proposed by Lifeso et al. For different stages of cranio-vertebral junction tuberculosis.

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<tr>
<th>Stages</th>
<th>Description</th>
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<tr>
<td>I</td>
<td>The ligaments are intact, there is minimal bone destruction and there is no evidence of anterior displacement of C1 on C2. It would require cervico-thoracic orthosis until the stability has been achieved.</td>
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<tr>
<td>II</td>
<td>There is ligamentous disruption with anterior displacement of C1 on C2 but bone destruction is still minimal; there could be proximal translocation of the odontoid. A Halo traction following reduction with gentle traction is recommended. After achieving normal alignment posterior fusion is done with continuation of cervico-thoracic orthosis for three months.</td>
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<td>III</td>
<td>There is marked bone destruction with complete obliteration of the anterior arch of C1, and eventually complete loss of the odontoid process; these cases exhibit marked instability of the occipito-cervical junction. A Halo traction is initially used to achieve alignment which is followed by posterior fusion. Halo vest immobilization is then used until stability is achieved.</td>
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Irrespective of surgical intervention or not, ATT is the key stone for the management of CVJ TB. All cases should receive ATT with initial four drug regimen (rifampicin 10 to 20 mg/kg/day; isoniazid 5–10 mg/kg/day and ethambutol 15 mg/kg/day in a single daily dose and pyrazamide 20–35 mg/kg/day in two divided doses). The duration of the 4 drug regimen has been variedly reported as 3 months to 18 months \cite{1, 3, 4}.

Conclusion

Treatment of CVJ tuberculosis cannot be standardized, it has to be tailored to each patient broadly presence of abscess formation, atlanto-axial subluxation with cervico-medullary compression, severe or progressive neurological deficit should undergo surgical intervention at the earliest. Surgery provides for immediate neurological recovery, stability, allows for early mobilization and also reduces the cost of staged surgery. The long term prognosis is excellent provided it is detected early and treated appropriately with surgical interventions and with adequate duration of ATT.

References

10. Moses Joseph Arun Kumar, Vedantam Rajekhar. Outcome in neurologically impaired patients with craniovertebral junction tuberculosis: results of combined