

**Case Report**

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# Craniovertebral Junction Tuberculosis with Atlantoaxial Dislocation : A Case Report and Review of the Literature

Craniovertebral junction (CVJ) tuberculosis is a rare disease, potentially causing severe instability and neurological deficits. The authors present a case of CVJ tuberculosis with atlantoaxial dislocation and retropharyngeal abscess in a 28-year-old man with neck pain and quadriparesis. Radiological evaluations showed a widespread extradural lesion around the clivus, C1, and C2. Two stage operations with transoral decompression and posterior occipitocervical fusion were performed. The pathological findings confirmed the diagnosis of tuberculosis. Treatment options in CVJ tuberculosis are controversial without well-defined guidelines. But radical operation (anterior decompression and posterior fusion and fixation) is necessary in patient with neurological deficit due to cord compression, extensive bone destruction, and instability or dislocation. The diagnosis and treatment options are discussed.

**KEY WORDS :** Tuberculosis · Craniovertebral junction · Atlantoaxial dislocation · Retropharyngeal abscess.

## INTRODUCTION

Craniovertebral junction (CVJ) tuberculosis is an uncommon disease, accounting for only 0.3% to 1% of tuberculous spondylitis<sup>3)</sup>. Clinical manifestations range from mild or nonspecific symptoms to severe instability and neurological complications. Diagnosis may be difficult and management can be challenging, requiring different treatment modalities depending on the clinical symptoms or radiological features. Treatment options are also controversial without well-defined guidelines, including a conservative approach (anti-tuberculosis medication and external fixation), relatively simple surgery (posterior fusion and fixation), or radical surgery (debridement, decompression and stabilization)<sup>1-4,6-13,15-18,20)</sup>.

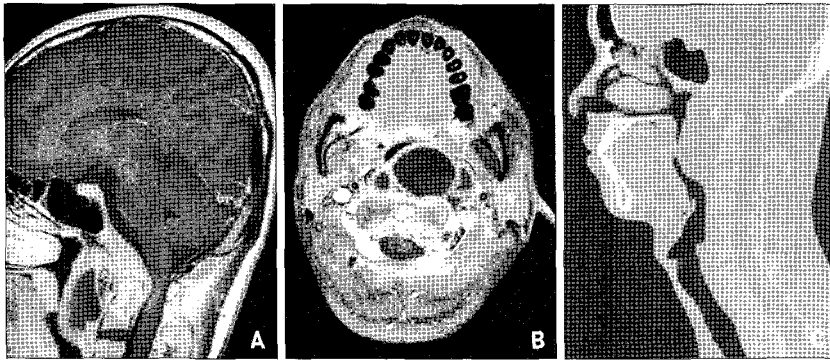
The authors present a case of CVJ tuberculosis with atlantoaxial dislocation (AAD) and retropharyngeal abscess. The diagnosis and treatment options are discussed.

## CASE REPORT

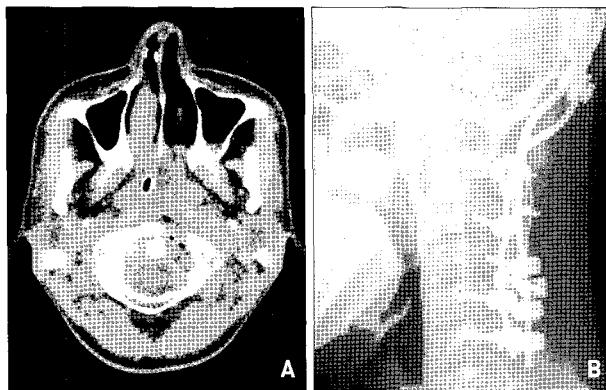
A 28-year-old male was admitted to the hospital with a five-month history of worsening posterior neck pain and quadriparesis. His pain was aggravated by moving or walking but was relieved by bed rest. He was healthy with no medical or surgical history, and no systemic illness was noted. He reported a transient tongue deviation to the left side. He had experienced generalized malaise and weight loss for 4 weeks before admission to the hospital. Neurological examination revealed mild weakness and increased deep tendon reflexes of both arms and legs, with positive ankle clonus and Babinski sign. Laboratory results including serologic tests, tumor marker and rheumatoid factors were not significant except for elevated white blood cell (WBC) count and erythrocyte sedimentation rate (ESR). Chest radiography showed no evidence of pulmonary lesion.

Magnetic resonance (MR) imaging of the brain and cervical spine revealed a widespread extradural lesion around the clivus, C1, and C2, causing spinal cord compression at the cervicomedullary junction. Two cystic lesions, one in the retropharyngeal space and the other in the spinous process of C2 were noted; these cysts were connected to each other. Cervical computed tomography (CT) showed erosion at the left side of the clivus, anterior arch of C1,

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**Fig. 1.** A : T1-weighted Gadolinium-enhanced magnetic resonance image revealing an extradural lesion around the clivus, C1, and C2, causing spinal cord compression at the cervicomedullary junction. Cystic lesions are noted in the retropharyngeal space and C2 spinous process. B : T1-weighted Gadolinium-enhanced axial magnetic resonance image showing retropharyngeal abscess and involving C2 body. C : Sagittal computerized tomography image showing that the lesion is eroding clivus, C2 body and spinous process and increased atlantodental interval.



**Fig. 2.** A : Axial computerized tomography image showing cord decompression and reduced atlantoaxial dislocation after transoral decompression. B : Postoperative lateral view of the cervical spine showing the occipitocervical fusion and fixation (occiput to C5) in place.

dens, lamina of C2, spinous process of C2, and increased atlantodental interval (Fig. 1).

Transoral aspiration and drainage of the retropharyngeal cyst were performed, and the exudate was sent for bacteriologic evaluations. Staining and culture for acid fast bacilli (AFB) were negative. However, polymerase chain reaction (PCR) for *Mycobacterium tuberculosis* (*M. tuberculosis*) showed positive results. Following these procedures, the impression of CVJ tuberculosis was made. Subsequently, the combination therapy of isoniazid (INH), rifampicin (RFP), ethambutol (EMB) and pyrazinamide (PZA) was initiated empirically.

Two stage operations with anterior (transoral) decompression and posterior occipitocervical fusion with autologous iliac bone graft and instrument were done for the confirmation of tuberculosis and for the decompression of the cervicomedullary cord compression and the reduction and fixation of AAD. First, transoral decompression of the diseased bone and granulation tissue were done. On the left side, there was destruction of the lower clivus, anterior arch of C1, and dens.

Whitish yellow, thick, calcified tissue was noted and multiple tissue biopsies were obtained. The frozen section revealed chronic granulomatous inflammation with caseation necrosis, which was consistent with tuberculosis. After curettage of the granulation tissue and removal of the left side of lower clivus and anterior arch of C1, the left side of dens was drilled and epidural granulation tissue was removed to decompress the spinal cord. The ligaments around the dens were not definitely identified because the granulation tissue was adhere to and mingled with these structures. Postoperatively, the patient

was immobilized in a halo vest. The second surgery was performed 3 weeks later. The lamina and spinous process of C2 were severely destroyed by the disease and removed. Fixation with the occipital plate, the rods, and the screws (SUMMIT™, DePuy Spine Inc. Raynham, MA, USA) from the occiput to C5 was performed (Fig 2). Pedicle screw insertion to the right C2 pedicle, lateral mass screw insertion to the C4, C5, and left C3 lateral mass. Pedicle fixation to the left C2 pedicle could not be performed due to bony destruction, narrow pedicle diameter, and proximity to the vertebral artery on CT. Lateral mass screw insertion to the C1 and the right C3 could not be performed due to poor bone quality by the disease. After the insertion of screws and the occipital plate, rods and transverse bar were placed. After modification of the autologous iliac bone graft harvested from the left posterior iliac crest, C1 and C3 fusion by Gallie method was done. Cancellous autograft from posterior iliac crest was placed after decortication of C3, C4, and C5 lamina and facet. After the second operation, the patient had no posterior neck pain and improved quadriplegia. The tuberculosis was well-controlled with medication.

## DISCUSSION

Cervical Pott's disease is unusual. Furthermore, CVJ tuberculosis has been reported to affect 0.3% to 1% of all Pott's disease patients<sup>3</sup>.

Evidence of systemic symptom may be absent and early complaints may involve pain or stiffness in the neck. Diagnosis may be delayed until signs of advanced disease develop. Smear and culture for bacilli are reported to be positive in less than 50% of cases<sup>14</sup>. Histological study has shown granulomatous tissue with or without caseation necrosis in approximately 75% of cases. Sensitivity of the PCR test for *M. tuberculosis*

**Table 1.** Reported series of tuberculous AAD

Authors	Year	No. of Patients	Management*
Allali et al. <sup>1)</sup>	2000	2	Minerva jacket+PF (n=1), PF (n=1)
Arunkumar et al. <sup>2)</sup>	2002	6	Transoral drainage+PF (n=2), TOD+PF (n=4)
Behari et al. <sup>4)</sup>	2003	17	Minerva jacket (n=8), TOD+PF (n=2), PF (n=7)
Bhojraj et al. <sup>6)</sup>	2001	9	Occipitomenal collar (n=1), PF (n=8)
Edwards et al. <sup>7)</sup>	2000	1	Transoral drainage
Fang et al. <sup>8)</sup>	1983	4	Transoral debridement+AF
Gupta et al. <sup>9)</sup>	2006	16 <sup>†</sup>	Halo
Jain et al. <sup>10)</sup>	1999	4	Traction/four post collar (n=3), Traction+PF (n=1)
Lal et al. <sup>13)</sup>	1992	6 <sup>‡</sup>	Drainage of abscess+Minerva jacket (n=2), Foramen magnum decompression+PF (n=1), Resection of C1 posterior arch+PF (n=2), Drainage of abscess+PF (n=1)
Lifeso et al. <sup>15)</sup>	1987	8	Cervicothoracic orthosis (n=1), Halo (n=1), PF (n=6)
Shukla et al. <sup>17)</sup>	2005	14	TOD+PF (n=9), PF (n=5)
Sinha et al. <sup>18)</sup>	2003	18	Anterior transcervical retropharyngeal decompression+PF

\*All patients underwent antituberculous medication. †Treated after 1998. ‡All patients underwent skull traction for 1 week. §All patients underwent transoral biopsy. TOD : transoral decompression, PF : posterior fusion, AF : anterior fusion

was reported from 91.4% to 97.6%, but only between 40% and 69% for AFB smear-positive and-negative respiratory specimens. Specificity was high (99.5% to 100%)<sup>5,19)</sup>. In this case, *M. tuberculosis* was not isolated by conventional culture of pus and tissue specimen. The diagnosis was confirmed by pathologic findings. The role of PCR methods for extrapulmonary tuberculosis is still unknown, but it might be helpful to make a rapid diagnosis not conclusive just by smear and culture findings, as in this case.

Retropharyngeal abscess in adults can be caused by direct invasion of pathogen as a result of trauma, foreign body, or iatrogenic cause (e.g., endoscopy or intubation). The spread of tuberculosis infection usually takes a retrograde route, reaching the craniovertebral joints. Subsequently, the infection causes destruction of the bony and ligamentous structures, and eventually produces cervicomedullary neural compression and occipitocervical or atlantoaxial instability<sup>12)</sup>. The occipito-atlanto-axial complex has a wide range of motion, accounting for approximately 80 degrees of rotation and significant flexion<sup>21)</sup>. The destruction of this complex can cause abnormal translational and rotational movements, leading to severe morbidity and even death. In this case, the patient presented with severe progressive neck pain, indicating instability.

The treatment options for CVJ tuberculosis have been controversial, from an absolute conservative approach to radical extirpation<sup>1,2,4,6-13,15-18,20)</sup>. This inconsistency is mainly due to the absence of well-proposed guidelines; however, it is also true that the symptomatic or radiological features of this disease may be diverse enough to justify several definite treatment modalities. Furthermore, the choice of intervention has depended largely on the surgeon's preference. Many

authors favored anti-tuberculous medication with or without external immobilization in case of minimal bone destruction and minimal neurologic deficit. Gupta et al. concluded surgery was not necessary, even in patients with advanced stage of disease (retropharyngeal abscess with or without AAD, gross bony destruction or angulation) on the base of their experience more than 3 decades<sup>9)</sup>. But, many reports stress surgical treatment when there are neurological symptoms due to cord compression or significant degree of instability, AAD or bone destruction. Many authors recommend posterior fusion and fixation and/or anterior decompression especially when anterior cord compression or irreducible AAD

existed (Table 1)<sup>1,2,4,6-10,13,15,17,18)</sup>. In this case, the management plan followed a radical operation line (anterior decompression, posterior fusion and fixation) because the patient showed neurological deficit due to cord compression, significant bony destruction and AAD. The patient had no posterior neck pain and improved quadriparesis after surgery.

## CONCLUSION

In this rare case of CVJ tuberculosis, the authors could get satisfactory outcome with two stage operations in a patient with neurological deficit due to cord compression, extensive bony destruction and AAD.

## References

- Allali F, Benomar A, El Yahyaoui M, Chkili T, Hajjaj-Hassouni N : Atlantoaxial tuberculosis : three cases. *Joint Bone Spine* 67 : 481-484, 2000
- Arunkumar MJ, Rajshekhar V : Outcome in neurologically impaired patients with craniovertebral junction tuberculosis : results of combined anteroposterior surgery. *J Neurosurg* 97(2 Suppl) : 166-171, 2002
- Attia M, Harnof S, Knoller N, Shacked I, Zibly Z, Bedrin L, et al : Cervical Pott's disease presenting as a retropharyngeal abscess. *Isr Med Assoc J* 6 : 438-439, 2004
- Behari S, Nayak SR, Bhargava V, Banerji D, Chhabra DK, Jain VK : Craniovertebral tuberculosis: protocol of surgical management. *Neurosurgery* 80 : 72-80, 2003
- Bennedsen J, Thomsen VO, Pfyffer GE, Funke G, Feldmann K, Beneke A, et al : Utility of PCR in diagnosing pulmonary tuberculosis. *J Clin Microbiol* 34 : 1407-1411, 1996
- Bhojraj SY, Shetty N, Shah PJ : Tuberculosis of the craniovertebral junction. *J Bone Joint Surg Br* 83 : 222-225, 2001
- Edwards RJ, David KM, Crockard HA : Management of tuberculomas of the craniovertebral junction. *Br J Neurosurg* 14 : 19-22, 2000
- Fang D, Leong JC, Fang HS : Tuberculosis of the upper cervical spine. *J Bone Joint Surg Br* 65 : 47-50, 1983
- Gupta SK, Mohindra S, Sharma BS, Gupta R, Chhabra R, Mukherjee KK, et al : Tuberculosis of the craniovertebral junction : is surgery

- necessary? *Neurosurgery* 58 : 1144-1150, 2006
10. Jain AK, Kumar S, Tuli SM : Tuberculosis of spine (C1 to D4). *Spinal Cord* 37 : 362-369, 1999
  11. Kanaan IU, Ellis M, Safi T, Al Kawi MZ, Coates R : Craniocervical junction tuberculosis : a rare but dangerous disease. *Surg Neurol* 51 : 21-25, 1999
  12. Kotil K, Dalbayrak S, Alan S : Craniovertebral junction Pott's disease. *Br J Neurosurg* 18 : 49-55, 2004
  13. Lal AP, Rajshekhhar V, Chandy MJ : Management strategies in tuberculous atlanto-axial dislocation. *Br J Neurosurg* 6 : 529-535, 1992
  14. Lee KS, Doh JW, Bae HG, Yun IG : Primary Infections Disorders of the Spine : Report of 40 Cases. *J Korean Neurosurg Soc* 25 : 1655-1660, 1996
  15. Lifeso RM, Weaver P, Harder EH : Tuberculous spondylitis in adults. *J Bone Joint Surg Am* 67 : 1405-1413, 1985
  16. Mohindra S, Gupta SK, Mohindra S, Gupta R : Unusual presentations of craniovertebral junction tuberculosis : a report of 2 cases and literature review. *Surg Neurol* 66 : 94-99, 2006
  17. Shukla D, Mongia S, Devi BI, Chandramouli BA, Das BS : Management of craniovertebral junction tuberculosis. *Surg Neurol* 63 : 101-106, 2005
  18. Sinha S, Singh AK, Gupta V, Singh D, Takayasu M, Yoshida J : Surgical management and outcome of tuberculous atlantoaxial dislocation : a 15-year experience. *Neurosurgery* 52 : 331-338, 2003
  19. Smith MB, Bergmann JS, Harris SL, Woods GL : Evaluation of the Roche AMPLICOR MTB assay for the detection of Mycobacterium tuberculosis in sputum specimens from prison inmates. *Diagn Microbiol Infect Dis* 27 : 113-116, 1997
  20. Tuli SM : Results of treatment of spinal tuberculosis by "middle-path" regime. *J Bone Joint Surg Br* 57 : 13-23, 1975
  21. White AA III, Panjabi MM : The clinical biomechanics of the occipitoatlantoaxial complex. *Orthop Clin North Am* 9 : 867-878, 1978