

Occipital condyle syndrome in a case of rotatory atlantoaxial subluxation (type II) with craniovertebral junction tuberculosis: Should we operate on “active tuberculosis?”

ABSTRACT

Tuberculosis of the craniovertebral junction is rare as well as intriguing. We present a unique amalgamation of three rare entities: craniovertebral tuberculosis, occipital condyle syndrome, and nontraumatic type II rotatory atlantoaxial dislocation in one patient. We reviewed the limited literature available and the underlying pathophysiology to highlight the pattern of the disease presentation, progression, and response to management options. A 13-year-old girl presented with neck pain, torticollis, and right hypoglossal weakness following a fall from stairs 10 months back. Radiological investigation suggested right occipital condylar destruction with right-sided neck tilt and rotatory atlantoaxial dislocation. The contrast magnetic resonance imaging was suggestive of craniovertebral tuberculosis with primary foci in the right lung (apical cavitory lesion). In view of an intact neurological condition, she was started on antitubercular treatment and she continues to do well during the follow-up. It remains debatable if an anticipation of this problem calls for a surgical addressal at the acute stage of the disease as a delayed correction is likely to be more complex. While a halo device is preferable in these cases, it remains cumbersome and less preferred in comparison to the Philadelphia collar.

Keywords: Collar, conservative, craniovertebral junction, halo, surgery, tuberculosis

INTRODUCTION

Tuberculosis is a disease that can affect virtually any organ of the body and can have protean manifestations. Craniovertebral junction (CVJ) involvement in tuberculosis is rare. The postinfective bony instability and neuraxial compression in this region make CVJ tuberculosis a special subgroup. In background of tuberculosis, the amalgamation of occipital condyle syndrome (OCS) and nontraumatic type II rotatory atlantoaxial dislocation poses serious concerns regarding whether to intervene surgically or not. We reviewed the limited literature available and the underlying pathophysiology to highlight the pattern of disease presentation, progression, and response to various treatment strategies. In our patient, we opted conservative medical management on antituberculosis, based on Dr. Goel's principles, but the dilemma still persists on the timing and need of fixation in follow-up.^[1]

CASE REPORT

A 13-year-old girl presented with complaints of neck pain and a typical “cock robin” attitude of the head with complete

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restriction of the neck movements following a fall from stairs 10 months back. Tongue deviation to the right side suggested an ipsilateral hypoglossal nerve palsy. She had bilateral tender cervical lymphadenopathy involving the levels II, III, and IV. The patient typically held her head in the hands to avoid any movements and there was a significant improvement in pain on application of a cervical collar. Her blood erythrocyte sedimentation rate (ESR) and lymphocyte count were raised.

On computed tomographic scan of the craniovertebral junction, a right lateral subluxation of the atlanto-occipital and atlantoaxial joints with erosion of the superior cortex of the right lateral mass of atlas, occipital condyle, and adjacent occiput were seen [Figure 1a-c]. On contrast-enhanced magnetic resonance imaging (MRI) of the head and neck, there was a peripherally enhancing collection seen along the base of the skull on the right side and multiple enlarged necrotic as well as nonnecrotic posterior cervical lymph nodes were seen along the right internal jugular vein [Figure 2a-c]. A single cavitory lesion involving the apical lobe of the right lung was also seen with mediastinal lymphadenopathy. A C1-C2 rotatory subluxation was detected with an increased atlantodental interval (fielding and Hawkins classification Type 2) with an increase in the distance between the C1 posterior arch and C2 lamina. O-C1 subluxation with basilar invagination was present [Figure 3a-d]. The vertebral angiography was suggestive of a left dominant vertebral artery, while the course of right vertebral artery was normal.

The patient was thus diagnosed as a case of OCS due to fracture of the right occipital condyle and right lateral mass of C1, with type 2 rotatory C1-C2 dislocation, secondary to a craniovertebral junction tuberculosis. In background of high prevalence of tuberculosis in India and raised ESR, empirical antituberculosis treatment (ATT) was initiated. She was managed with neck mobilization using a Philadelphia collar. At 3-month follow-up, the patient has significant improvement in pain; however, there was persistent deformity. At the time of writing this article, we are still considering judicious indication and timing of intervention.

DISCUSSION

This case was a unique experience where craniovertebral tuberculosis presented with OCS with rotatory subluxation of atlantoaxial joint that was precipitated by a history trauma. Unilateral suboccipital pain with ipsilateral 12th nerve palsy is termed as OCS.^[1-6] It was first described by Greenberg in 1981.^[6] It is characterized by occipital pain radiating toward the mastoid, ear, and vertex. There is painful restriction of neck movement leading to a cock robin deformity. Supporting

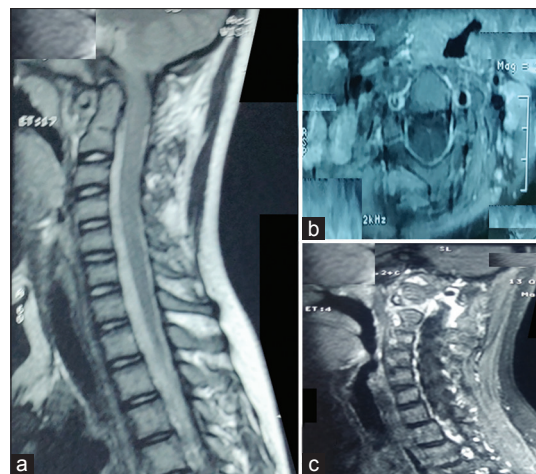


Figure 1: (a) T2-weighted magnetic resonance imaging of cervical spine, sagittal image shows narrowing at craniovertebral junction with anterior compression of cord at cervicomedullary junction but no intensity changes are seen. (b) T1-contrast, axial image showing multiple enlarged homogeneously enhancing lymph-nodes. (c) T1-contrast, sagittal paramedian image showing enhancement at craniovertebral junction

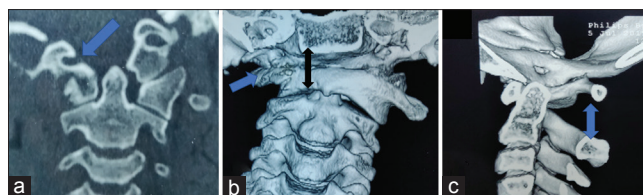


Figure 2: (a) Plain computed tomography of craniovertebral junction showing the hypoglossal canal in relationship to the right condyle. (b) Three-dimensional reconstruction of craniovertebral junction was suggestive of erosion of right occipital condyle and lateral mass of C1. (c) Three-dimensional reconstruction of craniovertebral junction sagittal image showing increased distance between the C1 posterior arch and C2 lamina, suggestive of instability

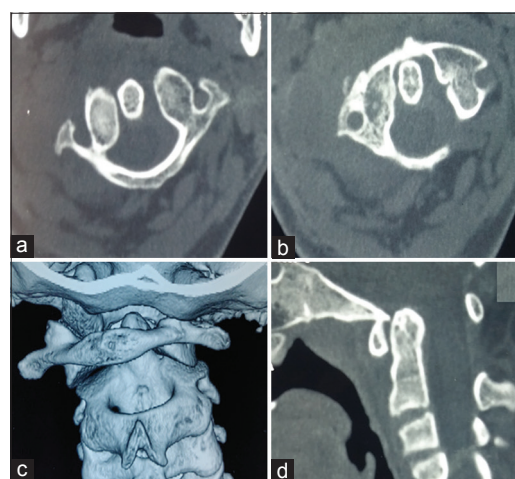


Figure 3: (a and b) Axial computed tomography cervical spine images showing rotated C1 in relation to odontoid process. (c) Three-dimensional reconstruction of craniovertebral junction, posterior view, showing subluxation of C1 over C2. (d) Plain computed tomography of craniovertebral junction showing basilar invagination and increased (atlantodental interval = 3.8 mm)

the head in hands, a unique sign of “apprehension” seen in tuberculosis was also present in this case. OCS due to CVJ-Potts spine is rare. The most common etiology for OCS is malignancy, with metastasis mostly from prostate and lungs.^[7-9] Other rare causes include inflammatory pseudotumor and Wegener’s granulomatosis. Table 1 summarizes previously reported cases of OCS associated with CVJ tuberculosis.

Postinfectious rotatory atlantoaxial dislocation is rare and was first described by Charles Bell in 1830 and then defined as a syndrome in 1951 by Grisel.^[10,11] Multiple mechanisms of underlying pathophysiology have been postulated, the most well accepted being the “two hit hypotheses.”^[12] Following upper respiratory tract infection, the muscle spasm results from the inflammatory mediators carried to the muscles from pharyngovertebral plexus. When there is preexisting laxity of ligaments at C1 C2, unequal strain due to muscles spasm leads to subluxation. A combination of bony destruction and ligament laxity led to the clinicoradiological presentation in our patient.

The CVJ tuberculosis results after the spread from primary site via hematogenous, lymphatic, or contiguous route.^[13] It

is the rarest of all spinal tuberculosis (incidence 0.3%–1%).^[14] Involvement of occipital condyles is even rarer.^[15,16] In this case, there was fracture of the right occipital condyle extending to the ipsilateral hypoglossal canal leading to 12th nerve palsy in addition to the typical deformity. Cock robin posture was due to the ipsilateral muscle spasm and associated asymmetry of O-C1 joint with C1-C2 rotatory subluxation.

Three important issues were relevant in our patient and pose dilemma in decision-making when faced with such a case:

1. Should one opt for a conservative treatment strategy in the face of a bony instability at CVJ?
2. Is histopathological confirmation necessary before initiating ATT in radiologically suspected cases?
3. What is the best nonoperative way of neck immobilization?

Surgery was not contemplated at this stage due to evidence of active disease and severe muscle spasm that would not allow realignment. This approach was followed in most of the reported cases of CVJ tuberculosis with OCS or rotatory atlantoaxial dislocation.^[17] A course of ATT was given for 18 months and all patients had improved. Is surgery really

Table 1: Previously reported cases of occipital condyle syndrome associated with craniovertebral junction tuberculosis

Authors	Year	Age/sex	Presenting complaint	12 th nerve palsy	Neck deformity	Level involved	Management	Outcome
Dhaon <i>et al.</i> ^[2]	2003	20 years/male	Pain Swelling Difficulty in swallowing	Not mentioned	Cock robin attitude	Occiput, C1	ATT - 9 months Head halter traction - 1week Four post collar	Improved (pain and deformity) (1.5 years)
Mohindra <i>et al.</i> ^[22]	2006	21 years/female	Right otalgia with otorrhea and hearing loss Neck pain with torticollis	Not mentioned	Torticollis present with painful restriction of neck movement	Right petrous bone. Basiocciput, right occipital condyle, right lateral mass of C1 and C2	ATT - 18 months Hard cervical collar	Improved - pain and deformity (> 18 months)
Mohindra <i>et al.</i> ^[22]	2006	55 years/male	Slurring of speech Dysphagia	Present (with lower cranial nerve involvement)	Patient had quadriplegia (neck pain or deformity not mentioned)	Clivus Occiput Left lateral mass of C1 and left pars C2	ATT - 18 months	Improved - pain, myelopathy and nerve palsy (> 18 months)
Kapoor <i>et al.</i> ^[3]	2007	9 months/female	Quadriplegia	Not mentioned	Painful restriction of movement	Occiput, C1 and C2	Transoral decompression with ATT (1 year)	Improved (1 year)
Chaudhry ^[4]	2014	25 years/male	Left occipital pain	Present	Painful restriction of movement	Clivus C1, C2, C3, C4 Left paravertebral abscess	ATT - 18 months	Pain improved; tongue atrophy persists (24 months)
Krishnan <i>et al.</i> ^[5]	2018	30 years/female	Dysarthria, neck pain, difficulty in lifting head off bed	Present	Painful restriction of movement	Right occipital condyle and right C1 lateral mass	ATT Sternal-occipital-mandibular immobilizer brace	Pain and tongue weakness improved
Our case	2019	13 years/female	Neck pain Torticollis	Present	Cock robin attitude	Right occipital condyle and right C1 and C2 lateral mass	ATT Hard cervical collar	Pain improved (1 month)

ATT - Antituberculosis treatment

required for CVJ instability? Many publications have strongly recommended conservative treatment, particularly in patients without a neurological deficit. We also treated along the conservative lines; however, the follow-up status of this patient at 3 months indicates that the patient will probably be cured of the disease but left with a deformity. This makes us wonder if surgical debridement, realignment, and fixation, followed by continuation of ATT, was a better treatment choice. Goel and Shah described unilateral facet fixation for CVJ tuberculosis.^[18] Waiting with ATT may lead to bony fusion which would increase the operative difficulty if surgery becomes necessary later due to deformity or late myelopathy. It would most likely require an ipsilateral vertebral artery mobilization to gain access to the O-C1 and C1-C2 joints.^[14,15] In our patient, as the patient was responding well, a decision to continue the ATT was made. Absence of myelopathy and a progressive neurological deficit, with good response to ATT, favored a conservative management.^[19] Dr. Goel highlighted the fact that CVJ tuberculosis may be managed conservatively in Stage II, provided there is no neurological deficit and patient is responding to drugs, in terms of pain.^[1]

Is histopathological verification compulsory?

Qureshi *et al.* analyzed the patients of CVJ tuberculosis and found that majority of histopathology samples came out as negative.^[20] They summarized that clinical indication is enough to start management, and if surgery is needed, biopsy may be taken at that time. Appaduray and Lo . highlighted the nonoperative management for CVJ tuberculosis and wrote that tissue biopsy is helpful only where drug-resistant tuberculosis is suspected.^[21] In our experience, a contrast-enhanced MRI with ESR and lymphocytosis is sufficient to diagnose and start treatment. A clinical improvement, particularly a sharp decrease in the pain along with overall improvement in general health, indicates a good response to ATT, even in the face of unchanged radiology.

Halo Verses Hard collar immobilization

A number of assistive devices are prescribed to immobilize cervical spine in cases if instability.

The options include halo vest, cervical orthosis, Philadelphia collars, Minerva collars, Aspen, stiff-neck Miami collar, and NeLoc orthoses. Problems such as pin loosening, infection of pin sites, penetrating skull bone, and a poor cosmetic appearance of these devices make the halo orthosis a less preferred choice. Other rarer side effects include pressure sores from plastic vests, nerve injury, dual penetration, pain, change in swallowing function, stiffness of facial muscles, change in bolus flow, and occipital ulceration. Karimi *et al.*

reviewed literature on efficacy of cervical arthrodesis and found halo vest being the best choice, followed by Minerva collar.^[17] However, the above two are cumbersome and often patients are noncompliant. Hence, a halo vest should have been advised, but considering the family situation and unsure about regularity in follow-up, we decided to give the most compliant option of collar. Interestingly, Grady *et al.* supported the use a cervical collar for neck immobilization; they proposed that good results with a Philadelphia collar depended on three factors: degree of movement while in the collar, patient reliability, and the age of the patient.^[19]

CONCLUSION

It is rare for cervical Pott's spine to present as OCS with atlantoaxial rotatory subluxation. Radiological findings in a typical clinical setting are good enough to initiate ATT. While almost all of these patients will respond to a course of ATT, presence of a deformity creates a management dilemma as a healing with the persistence of deformity may be an unwelcome outcome. It remains debatable if an anticipation of this problem calls for a surgical addressal at the acute stage of the disease as a delayed correction is likely to be more complex. While a halo device is preferable in these cases, it remains cumbersome and less preferred in comparison to the Philadelphia collar.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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