

Case Report

Craniocervical junction tuberculosis: Usual pathology at an unusual site

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Abstract

Background: Tuberculosis (TB) of the craniocervical junction is rare even where the condition is endemic. It poses problems in both diagnosis and management if not managed in time it may cause life-threatening complications.

Case Description: An 18-year-old male patient presented with pain in the nape of the neck since 12 months duration which was not improving with medication. After magnetic resonance imaging of cervical spine, he was diagnosed as craniocervical junction TB. We did a transoral decompression of abscess with biopsy along with posterior decompression of cord and occipitocervical fusion. Biopsy of pathological material came as TB. He was advised for anti-tubercular therapy for 18 months.

Conclusion: Although craniocervical junction TB is a rare disease, the outcome of treatment is good. Antituberculous drug therapy remains the mainstay of treatment after confirming the diagnosis. The surgical management options include transoral decompression with or without posterior fusion, depending upon the presence and persistence of atlantoaxial instability.

Key Words: Craniocervical junction, posterior fusion, transoral decompression, tuberculosis

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INTRODUCTION

Tuberculosis (TB) is a worldwide health problem, according to the World Health Organization it is the leading infectious cause of mortality worldwide killing 1.45 million people.^[13] Spinal TB, first described by Sir Percival Pott in the 18th century, accounts for 50% of these cases^[2] and results in immense morbidity and mortality. TB of the craniocervical junction is rare^[5] and accounts for only about 1% of all cases of spinal TB. It is difficult to diagnose and manage and is a therapeutic challenge. It primarily involves the atlas and axis and in some cases, the occipital

region. Because of potential fatal complications, any infection at this site must be diagnosed early and treated promptly. Death is usually due to atlantoaxial dislocation causing compression of the cord.^[9] Although the clinical spectrum of spinal TB is variable, back pain is the major symptom. Spinal TB can lead to bone destruction and vertebral collapse resulting in paravertebral abscess and deformity. If prompt management is not employed, severe neurological symptoms ensue, which can lead to a debilitating consequence. Moreover, spinal TB still remains the leading cause of nontraumatic paraplegia in developing nations.

CASE REPORT

An 18-year-old male patient presented with pain in the nape of the neck since 12 months duration which was not improving with medication. The patient had stiffness of the neck with decreased range of movement in all directions. Routine hematological investigations were within the normal limit, and he was screened for rheumatologic problems without revealing any pathology. On magnetic resonance imaging of cervical spine [Figure 1], there were irregular lytic bony destruction in anterior arch and left lateral mass of C-1, left occipital condyle, left half of C-2 vertebra with prevertebral and left para vertebral collection, mild erosion in the odontoid process, with cranial migration of odontoid with tip up to the level of foramen magnum, there was compression of cord at this level with cord edema, suggestive of Koch's. We did a transoral decompression of abscess with biopsy along with posterior decompression of cord and occipitocervical fusion [Figure 2]. Biopsy of the pathological material shows the background of extensive necrosis and inflammatory granuloma containing lymphocytes, plasma cells, epithelioid cells, and multinucleated Langhan's type of giant cells suggestive of TB. Postoperatively, he was advised for antitubercular therapy for 18 months. Now, the patient has relief of pain and leading a normal life.

DISCUSSION

The occipitocervical junction, a transitional zone between the skull and the spinal column, serves as the most mobile part of the axial skeleton. Bony abnormalities affecting this complex results in dysfunction of the neural structures by compression along the entire circumference, altering the arterial supply, venous drainage, and changing the cerebrospinal fluid dynamics.



Figure 1: On magnetic resonance imaging of cervical spine shows irregular lytic bony destruction C-1, C-2, and occipital condyle, with cranial migration of odontoid with tip up to the level of foramen magnum, compression of cord with cord edema, suggestive of Koch's

Lymphatic channels play a dominant role in the etiopathogenesis of any infective process at the craniovertebral junction (CVJ). The infection probably begins in the retropharyngeal space with secondary involvement of bone, and is rarely primarily in the bone itself. Progression of the disease causes increasing ligamentous involvement, and the later stages involve increased destruction of bone.^[3,6] Retrograde infection may spread to the CVJ resulting in instability or effusion as an inflammatory response.^[7,8,10]

The lateral masses are the initial to be affected, and destruction of these will eventually lead to collapse or weakening of the bony pillars. Depending on the severity of the disease process, varying degree of bony destruction of the atlas may ensue. This bony destruction may manifest as neck pain, torticollis, occipital neuralgia, dysphagia, dyspnea (retropharyngeal abscess) or even as neurologic dysfunction.^[4]

At this stage, the disease if unabated will cause ligament destruction aggravating the instability at the CVJ. Atlanto-axial dislocation (AAD) may result and if the prodens interval is more than 5 mm, it signifies the destruction of the transverse ligament of the atlas. Varying degree of rotatory malalignment is commonly associated and needs to be considered during treatment of the condition.

It is very unusual to find the disease advanced to the stage of complete liquefaction and dissolution of the atlas. When this occurs (as in our case), the instability at the CVJ is severe and demands urgent immobilization. It also forewarns complete osteoligamentous destruction and warrants emergency management. The associated problems of pressure such as dysphagia and asphyxia may assume alarming proportions. This may as in our case necessitate tracheostomy for effective airway ventilation.

As a sequelae, secondary basilar invagination may occur adding a different clinical dimension to the disease. An occipital bone may be involved as an extension of the



Figure 2: Posterior occipitocervical fusion

disease with softening allowing the spinal column to further telescope into the posterior fossa. The proximity of the lower cranial nerves makes them vulnerable to be affected.

Conservative management for postinfective AAD,^[12] and a regime of bed rest, skull traction and anti-tuberculous treatment with mobilization only after 6–9 months,^[11] have been advocated. Such management can lead to skin and pulmonary complications, especially in patients with neurological deficit. One-stage anterior surgical debridement and fusion of the atlantoaxial joint have also been recommended, but had a failure rate of 50% and is technically more difficult than a posterior fusion.^[13] Posterior fusion allows rapid neurological recovery, prompt relief of pain and a high rate of fusion with minimal morbidity.^[6] In view of this and the early healing and stability afforded by fusion, it is recommended that patients with instability and neurological compromise should be stabilized by means of a posterior fusion.^[1] Early diagnosis and treatment should be the goal with avoidance of fatal instability. The surgical option should be considered, in light of the patient's general condition, associated cranial nerve palsies, and neurologic status.

CONCLUSION

Although CVJ TB is a rare disease, the outcome of treatment is good. Antituberculous drug therapy remains the mainstay of treatment after confirming the diagnosis.

The surgical management options include transoral decompression with or without posterior fusion, depending upon the presence and persistence of atlantoaxial instability.

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