Tubercular atlantoaxial dislocation in children: an institutional experience

SAMIR KUMAR KALRA, M.CH., RAJ KUMAR, M.CH., AND ASHOK KUMAR MAHAPATRA, M.CH.
Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

Object. In this paper the authors analyzed the clinical and imaging-documented profile of pediatric patients with tubercular atlantoaxial dislocation (AAD).

Methods. Seventeen children 16 years of age or younger with tubercular AAD were included in the study. Patients with reducible AAD underwent direct posterior stabilization. All patients were treated with a four-drug antituberculosis therapy (ATT) regimen (10–20 mg/kg/day rifampicin, 10–20 mg/kg/day isoniazid, and 15 mg/kg/day ethambutol in a single daily dose; and pyrazinamide 20–35 mg/kg/day in two divided doses) for 3 months. The pyrazinamide was then discontinued after 3 months and the ethambutol after 1 year. The rifampicin and isoniazid were continued for 18 months.

Results. Most of the patients had irreducible AAD. There was a high incidence of long tract signs, and the restriction of neck movements, as well as neck pain, was also very common. There was a significant delay in seeking neurosurgical consultation. Most patients were assigned poor preoperative grades, but they experienced excellent improvement postoperatively.

Conclusions. The presence of tubercular AAD in children can have subtle manifestations leading to delayed diagnosis. The successful management of tubercular AAD can be achieved after determining the extent of the disease process and the underlying instability. The goal of surgery is tissue diagnosis and relief of neural compression and stabilization. Medical treatment with ATT is an integral part of the treatment protocol. (DOI: 10.3171/PED-07/08/111)

KEY WORDS • atlantoaxial dislocation • craniovertebral junction • pediatric neurosurgery

The CVJ is an extremely rare site to be infected with TB, accounting for 0.3 to 1% of the total incidence of spinal TB.5,6,8,14,26 The incidence of TB in the spine is also relatively low; fewer than 1% of cases occur in the spine.1,6,21 With the resurgence of TB in immunocompromised patients, especially in developed countries, the incidence of TB in general is increasing worldwide. In recent years there has been an increased incidence of spinal TB, and a few large series of TB in the CVJ have been reported.3,4,10,18,24,25

Determining the presence of TB in the CVJ is important because of the potentially disastrous complications to which it can lead. The extensive osteoligamentous destruction caused by TB in the CVJ leads to instability of this complex transition zone. The common pathological conditions that occur due to this extensive destruction include the following: AAD, secondary basilar impression, epidural abscess, and/or granulation tissue. The resultant instability is associated with specific management issues, and its successful treatment depends on adhering to specific investigational and therapeutic norms. The presence of AAD following TB (tubercular AAD) in this region, especially in pediatric patients, may cause serious complications in that its symptoms are nonspecific, resulting in a delay in diagnosis and hence late presentation in an advanced state. An established scheme for the management of such cases is also lacking.

The clinical symptoms of patients with tubercular AAD may range from nonspecific to severe cervicomедullary compression with resultant quadriplegia, bulbar dysfunction, and respiratory insufficiency. The delayed diagnosis can be detrimental in these cases because of the underlying important neural structures that can be damaged by this dislocation. The importance of obtaining a biopsy sample from such lesions is demonstrated by the fact that management plans are entirely different depending on whether the AAD is or is not associated with TB. The surgical management of AAD, whether of a tuberculous nature or not, provides relief of neural compression along with stabilization. Patients in whom a diagnosis of TB has been made must be treated with ATT. The biopsy of all such suspicious lesions is therefore recommended.
Clinical Material and Methods

Patient Population

Seventeen pediatric patients 16 years of age or younger (age range 5–16 years, mean age 10.94 ± 3.34 years; male/female ratio 12:5) with tubercular AAD were treated from 1997 to 2006 at Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India. All patients underwent a thorough assessment of respiratory status, which included pulmonary function tests whenever possible and bedside measurement of the single breath count, chest expansion, and breath holding time. The systemic signs and symptoms of TB were actively sought in all patients. The clinical symptoms and signs at presentation of the included patients are summarized in Table 1.

All patients underwent surgery and follow-up at our institute, and the diagnosis of TB was confirmed histopathologically. The patients were graded based on their neurological disabilities. The grading system is a combination of those used by Ranawat et al. and Di Lorenzo. (Table 2). All patients were assessed preoperatively as well as postoperatively with the same grading system.

Preoperative Assessments

All patients underwent an initial assessment with lateral radiographs of the CVJ in neutral, flexion, and extension for the presence of AAD, prevertebral soft-tissue shadow, and bone destruction.

Sagittal and coronal contrast-enhanced MR images of the CVJ were obtained in all patients, and CT scans were obtained in seven to assess for extensive bone involvement due to the disease process. All patients underwent chest radiography to detect primary chest lesions. The biochemical investigations included the erythrocyte sedimentation rate in all patients and mycobacterial DNA amplification in 14. Serological evaluation for HIV-1 and -2 was performed in 16 patients who were suspected of being in an immunocompromised state.

Treatment Protocol

The treatment protocol was based on the presence or absence of reducibility of the AAD. Patients with irreducible AAD commenced cervical traction using Crutchfield tongs, and they were evaluated after 2 weeks with translateral radiographs of the CVJ preoperatively to assess for reducibility of their AAD. All patients with irreducible AAD underwent anterior decompression via the transoral route followed by posterior stabilization during the same operation (Figs. 1 and 4). For patients with the reducible variety, posterior stabilization was performed without anterior decompression (Figs. 2 and 3). All patients with reducible AAD had involvement of posterior elements from which a biopsy sample was taken. The biopsy was thus performed in all patients in the study. The management protocol is represented schematically in Fig. 5.

All patients were treated with a four-drug ATT regimen (10–20 mg/kg/day rifampicin, 10–20 mg/kg/day isoniazid, and 15 mg/kg/day ethambutol in a single daily dose; and pyrazinamide 20–35 mg/kg/day in two divided doses). The four-drug treatment was given for 3 months; at this time pyrazinamide was discontinued, and the ethambutol was discontinued after 1 year. The rifampicin and isoniazid were continued for 1.5 years. Pyridoxine (25 mg) was continued along with isoniazid. Two patients developed hepatic dysfunction during treatment, and they were started on a regimen of 15 mg/kg/day ethambutol; isoniazid, rifampicin, and pyrazinamide were discontinued. When results of the liver function tests returned to normal, isoniazid and rifampicin were restarted. Seven of the included patients commenced a regimen of ATT before surgery based on clinical and imaging findings, and 10 patients commenced drug treatment following surgery. All patients underwent neck stabilization with a hard cervical collar, which was worn for a period of at least 3 months or until an osseous posterior union had been attained. A detailed neurological examination and dynamic translateral radiographs (in flexion and extension) of the CVJ were performed after 3 months. Findings we particularly sought were new bone formation and a decrease in the extent of granulation tissue.

Surgical Technique and Diagnosis

For the anterior cervicomedullary compression, the transoral route (12 patients) was undertaken via the standard technique. Posterior stabilization was performed in all patients, and both the transoral route and posterior stabilization, whenever done, were performed in the same operation. In four patients, the modified Brooks technique was used for atlantoaxial fusion, in which the decorticated C-1 posterior arch and C-2 lamina were united using a central sublaminar braided wire without any intervening strut graft followed by placement of onlay bone grafts. For occipitocervical fusions, we used the Jain technique.
in 11 patients and occipitocervical fusion using contoured Steinmann pins in two.

The diagnosis of TB was based on acid-fast bacilli staining (seven patients) or culture (four patients) of *Mycobacterium tuberculosis*. Histological evidence of caseative necrosis and granuloma formation was obtained in 13 patients.

Results

Clinical and Imaging Findings

The most common clinical features were related to the involvement of corticospinal tracts with weakness, spasticity, and pyramidal signs present in most patients. There was a high incidence of neck pain (15 patients) and restriction of neck movement (13 patients). The mean duration of symptoms was $10.47 \pm 4.24$ months (range 5–21 months). Most patients presented with slowly progressive disease; only two exhibited acute deterioration and presented with rapidly progressing motor weakness and respiratory compromise. All patients were referred from peripheral hospitals, and the mean interval from their first visit to referral to our institution was $6.24 \pm 3.9$ months (range 1–16 months). Most (13 patients) were dependent partially or totally on others for their daily needs, and the distribution was nearly similar in the irreducible (75%) and reducible (80%) types of tubercular AAD (Table 3).

Most patients (12) had evidence of the irreducible type of AAD and only five had reducible AAD. None of the patients with the irreducible type achieved reduction on cervical traction. There was evidence of significant extradural granulation tissue compressing the upper cervical cord anteriorly in nine patients and an abscess extending posteriorly in six patients. In three patients there were secondary signal intensity intrinsic cord changes. Multilevel involvement was noted in three patients and another three had active lesions in their chests.

Outcome Complications

The mean follow-up period was $21.53 \pm 21.37$ months (range 5–87 months), and no patient was lost to follow-up.
The condition of most of the patients improved: four patients (80%) with the reducible type and 10 (83.3%) with the irreducible type experienced improvement from their preoperative grades. The pre- and postoperative grades remained the same in two patients (one from each group). We treated one patient with irreducible tubercular AAD by using the Jain technique (that is, a single-stage transoral posterior stabilization and sublaminar wiring procedure). Following this operation, the patient exhibited neurological deterioration due to compression caused by the sublaminar wiring. The patient underwent repeated surgery and stabilization using contoured Steinmann pin fixation. His neurological condition continues to improve, but his grade remains unchanged.

The assessment of imaging-documented fusion was based on the appearance of continuous trabecular bone bridging adjacent bone surfaces with an absence of motion. There was imaging evidence of cervical fusion in 14 patients. Two patients did not undergo follow-up long enough to assess for bone union, and another patient at 10 months of follow-up had no evidence of trabecular union. However, stability was achieved in the latter patient as evidenced by an absence of cervical spine motion on lateral flexion and extension radiographs. None of the cases had nonunion or fibrous union.

One patient experienced oral wound dehiscence following surgery via the transoral route. This was treated by regular oral care, broad spectrum antibiotic coverage, and a high-calorie diet.

**Discussion**

**Incidence and Instability**

The increased incidence of tubercular AAD has been due to the worldwide resurgence of TB. About 6% of extra-

![Fig. 2.](image1)

![Fig. 3.](image2)
Pediatric tubercular atlantoaxial dislocation

pulmonary TB occurs in the vertebral column, and only 1% is considered spinal TB. Tuberculosis in the CVJ originates in the retropharyngeal space and progressively involves and destroys the osteoligamentous complex in this region. Only rarely is the bone primarily involved. The ligamentous involvement manifests as neck pain and restriction in neck movement commonly seen in these patients. Tuberculous lesions in the occipitoatlantoaxial complex can cause serious instability and neurological deficits. The CVJ has a wide range of motion and cervical rotation. It is the most mobile segment of the spine, and in the event of its destruction, such movements are significantly compromised. Atlantoaxial dislocation is the most common disease process resulting from TB in this region. In tubercular AAD, extensive destabilization of the joint complex by the destroyed bone, ligaments, and articular surfaces of the joints occurs, and the abnormal mobility may cause recurrent cervico-medullary compression with potentially fatal complications.

Clinical Spectrum and Diagnostic Delay

The neurological deficits experienced by these patients are due to mechanical compression of the neural structures by tubercular abscess, granulation tissue, AAD, or basilar invagination. Vascular compromise by endarteritis has also been described as a cause. The involvement of pyramidal tracts, spinothalamic tracts, and posterior columns by the disease process at the cervico-medullary junction reflects itself in the clinical presentation. The initial involvement, however, is in the form of neck pain or restriction of neck movements commonly and is due to the involvement of the ligamentous complexes. These may be present for a long time before the overt signs of neural compression manifest.

The clinical scenario in children is complicated by the paucity and imprecision of their complaints. In addition the diagnosis is commonly missed or delayed significantly because most patients are first seen at peripheral centers. This is demonstrated in our series in that the mean interval between the patients’ first presentation to peripheral hospitals and referral to our center was more than 6 months. This time period is significant enough for TB to progress and lead to serious disabilities. The presentation of the majority of patients with poor grades further strengthens this claim. A high index of suspicion is required at the peripheral level regarding the occurrence of tubercular AAD in children and especially so in the patients living in endemic areas, and those with a personal or family history of TB, a history of contact, or TB at another site.

Tuberculosis of the CVJ should be kept in mind during differential diagnosis in patients with neck pain and longstanding restriction of neck movements. Immunocompromised patients and residents of endemic areas must be considered as being at a high risk for spinal TB and thus tubercular AAD. All patients with spinal TB with such complaints should be investigated for the presence of associated AAD.

Imaging Findings

On plain radiographs of the CVJ, the bone destruction leads to the loss of conventional radiological markers for demonstrating AAD and leads to difficulty in diagnosis. The changes on plain radiographs may lag behind by 2 to 6 months as the radiographic evidence of bone erosion appears only after 50% of the vertebra has been destroyed. These investigations are still helpful in identifying the TB disease process as well as AAD. Although no imaging findings are pathognomonic of tubercular AAD, the use of CT and MR imaging readily demonstrates the bone fragmentation at the endplates and associated obliteration of fat planes around the vertebral bodies. These findings coupled with clinical suspicion are highly specific findings. The presence of a multiloculated and calcified abscess with a thick, enhancing irregular rim is highly suggestive of TB. Contrast-enhanced images reveal the spread of abscess and granulation tissue along the vertebral bodies under the longitudinal ligament and in the epidural plane. The differential diagnosis includes rheumatoid arthritis, brucellosis, sarcoidosis, fungal infection, lymphoma, or chordoma, and the presence of caseating necrosis usually helps in the differentiation. Confirmation, however, is still required by histological analysis of the tissue specimen.

The direct relationship of spinal column narrowing to the extent of neurological deficits has been proposed. Al-Mulhim et al. reported that narrowing of greater than 75% gives rise to severe neurological deficits, and narrowing of the cervical spine by less than 50% produces only mild to moderate deficits. This suggestion may be fallacious given that the underlying instability may not be due to canal narrowing alone, and in these cases there may be severe neurological deficits despite a wide canal diameter. Moreover, the disease...
process may be widespread, and yet there may be no significant canal compromise. These patients are more prone to experience sudden neurological deterioration following trivial events. During the stages of healing of the tuberculous process, the canal diameter along with stability should also be assessed in each patient.

The incidence of irreducible AAD was much more than that of reducible AAD (12 patients compared with five patients) in our series. This is contrary to the fact that TB is a destructive process and, expectedly, the incidence of reducible AAD should be greater. There is enough evidence in the literature that the reducible type of AAD in children later progresses to the irreducible type if it is not recognized; therefore, it is imperative to recognize reducible AAD early. In the series by Behari et al., it was seen that of the nine patients with minor deficits (Grades I and II), five (55.6%) had reducible AAD, whereas all eight patients with severe deficits (Grades III and IV) had irreducible AAD. This demonstrates that most patients with good grades have reducible AAD and those with irreducible AAD have poorer grades.

**Treatment Options and Protocol**

The management of TB in the CVJ in general and tubercular AAD in particular has been a subject of debate over the years. There have been proponents of medical treatment alone with absolute conservatism who question the use of any form of surgical intervention whatsoever, and there are proponents of radical surgical extirpation as well. This is mainly because of the paucity of available literature pertaining to TB of the CVJ, especially tubercular AAD; most of the information has come as case reports or as part of larger studies concerning tuberculous spinal disease with many unusual presentations. The management protocol is difficult to standardize and the treatment has to be tailored according to the condition of the patient and extent of dis-
Pediatric tubercular atlantoaxial dislocation

In the study by Gupta et al., in which they analyzed 51 patients with tuberculous AAD over a course of 26 years, conservative treatment was undertaken in 16 patients in the past 6 years. This has been a recent trend in their series; initially they also performed surgical decompression in patients with poor grades. Of these 16 patients, only three were children; a transoral needle aspiration was performed in all three. These authors used radiological staging for assigning grades to the patients, and this may not correlate well with the clinical profile of the patients. In their series, a 10-year-old child was in radiological Stage 1 and assigned Nurick Grade 5. Although they recommend the use of halo fixation in all patients, this device may not be well tolerated by children. Moreover they undertook surgical intervention in the form of aspiration or biopsy in all their patients.

The presentation in children is usually delayed in our setup due mainly to the improper and late diagnosis at the peripheral centers from where these patients are referred for neurosurgical care. The subtle clinical manifestations of the disease process make diagnosis more difficult in children, and this is exemplified in our series in that 13 (76.5%) of 17 children; a transoral needle aspiration was performed in all three. These authors used radiological staging for assigning grades to the patients, and this may not correlate well with the clinical profile of the patients. In their series, a 10-year-old child was in radiological Stage 1 and assigned Nurick Grade 5. Although they recommend the use of halo fixation in all patients, this device may not be well tolerated by children. Moreover they undertook surgical intervention in the form of aspiration or biopsy in all their patients.

The presentation in children is usually delayed in our setup due mainly to the improper and late diagnosis at the peripheral centers from where these patients are referred for neurosurgical care. The subtle clinical manifestations of the disease process make diagnosis more difficult in children, and this is exemplified in our series in that 13 (76.5%) of 17 patients were functionally dependent. The presentation of children in poor grades dissuaded us from meting out conservative treatment, and the good results following surgery (in 14 of 17 patients) further strengthened our claim. The scenario may be different in adults where the scope of conservative treatment may be more valid and has been tried by us as well. Adults tend to present when their disease is in the earlier stages. The series by Gupta et al., largely consists of adult patients and hence cannot be compared with our experience. Thus we believe that children should be treated more aggressively than adults.

Menezes and colleagues have observed that skeletal traction, in itself, is a type of operative reduction and must be considered as such. They have also commented that prolonged immobilization can be a problem and that surgical decompression and stabilization still play a significant role in the management of problems in the region of the CVJ, including TB.

Histological confirmation is very important because the radiological findings even at best are nonspecific. The primary aim of treatment is relief of cervicomedullary compression and stabilization of the CVJ. The role of medical treatment alone is well documented and accepted, and this should be tried in selected cases. In our series, none of the patients underwent medical treatment alone because none presented at an early stage or in a good grade. Even the one patient with irreducible AAD whose condition was assessed as Grade I on presentation eventually underwent surgery because the diagnosis was unclear. In addition the patient did not show signs of clinical or imaging-documented improvement despite an adequate trial of ATT. The neurological conditions of patients with tubercular AAD are more prone to deteriorate because of the relative mobility that results in recurrent cervicomedullary compression, even when the tuberculous disease at the CVJ has responded to ATT. The placement of cervical traction often results in reduction of the AAD after which direct posterior stabilization can be performed. In our series none of the 12 patients with irreducible tubercular AAD achieved reduction by cervical traction. Patients with TB of the CVJ without AAD have a greater chance of responding to medical treatment alone than those with AAD.

Successful management entails achievement of neural decompression and stabilization. The various techniques of posterior stabilization have been described and used over the years. Despite the fact that more contemporary fusion techniques can be used for atlantoaxial fusions, we chose this one because of our wide experience with it and are pleased with its consistently good results.

The key to continuing improvement and stabilization of the disease process is institution of ATT and ensuring drug compliance. Patients and their relatives must be made aware of the importance of drug treatment as well as its potential side effects. All patients in the present series were given ATT for 18 months, and there were no complications due to the medications.

### Tissue Diagnosis and Drug Treatment

Tissue diagnosis is absolutely mandatory. Edwards et al. proposed that without bacteriological or histological confirmation, one should not assume that lesions in the CVJ are caused by TB, and only in confirmed cases of systemic TB is a clinical trial of ATT is justified. This is primarily due to the absence of pathognomonic radiological features of tubercular AAD. In the present series, a tissue diagnosis was obtained in all cases.

The ATT regimen must be given to all patients with clinical or imaging-documented suspicion of TB to prevent dissemination of the disease. The difficulties associated with ATT are that the culture of the offending bacteria becomes difficult after commencing drug treatment. The aggressive debridement of offending granulation tissue, abscess, and necrotic bone along with ATT is the mainstay of the treatment.

### Outcome of Treatment

The chances of improvement following successful and judicious management are heartening. There is a likelihood of significant improvement when therapeutic options are used along with ATT. The disease process is essentially benign with an excellent response to treatment. The disease
course has to be controlled with ATT or without surgical decompression and stabilization. The approach is at best individualized and depends primarily on the extent of involvement of the disease process. Recognition at an early stage helps in avoiding surgical intervention; however, this may not be true for all cases as is demonstrated in our series.

Conclusions

The presence of TB at the CVJ in children can have varied pathological manifestations, the most common being the presence of AAD. The manifestations of tubercular AAD may be subtle in children and thereby cause a delay in diagnosis. The successful management of tubercular AAD can be achieved after the establishment of the extent of disease process and the underlying instability. The presentation in children is usually at a late stage and hence they are less suitable for treatment by conservative therapy alone. The goal of surgery is the relief of cervicomedullary compression and stabilization coupled with tissue diagnosis. Medical treatment with ATT is an integral part of the management protocol. Following the treatment guidelines with an individualized approach leads to good results.

References


Address reprint requests to: Raj Kumar, M.Ch., Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow 226014, India. email: rajkumar@sgrgi.ac.in.