Article *NIMHANS Journal*

# Tuberculous Atlantoaxial Disease Including Dislocations

**Volume: 06 Issue: 2S December 1988 Page: 89-98**

Anil P Karapurkar, - *Bombay*

Though tuberculosis is till rampant in India, tuberculous disease of the atlantoaxial region is uncommon. Tuli [1] states that the incidence is less than 1% of spinal tuberculosis. Not much has been written about it. Despite a diligent search of the literature few articles in the English language could be found, those of Pandya [2] in 1971 and Tuli [1] in 1974 being the last.

This report is based on 31 cases, 25 of these are from the department of Neurosurgery, 5 from the department of Orthopedics and one patient has been treated by Dr. Gajendra Sinh at the J J Hospital. The first patient was seen in 1958 and the last 2 are still in the wards. Though this is a short series some distinctive clinical, radiologic and therapeutic principles can be appreciated.

## Material and Methods

Of the 31 cases, the majority, 20 were female. Thirteen cases were between 20 and 30 years old, 7 were between 10 and 20 years of age, 5 were from 3 to 10, 2 were between 30 and 40 and 2 more between 40 and 50. One patient was 56 and the oldest was 76 years old. Each and every one of these patients had neck pain and neck stiffness as the presenting complaint. The duration of pain before the patient presented for treatment varied from 18 months to 10 days with an average of 6 months. Three of them woke up with neck pain and wry neck. Four patients gave a history of fall preceding the neck pain. Two patients were epileptic and the complaints were preceded by a grand mal convulsion. One patient who had neck pain for some weeks woke up with quadriplegia. One patient who had neck pain for some duration noted that suddenly her neck had become short. Another patient heard a distinct snap which was followed by progressive weakness. One patient was cleaning the floor of his water tank when he had neck pain and a wry neck.

There were five type of clinical presentation

The commonest was that of progressive neck pain and restriction of neck movements. The pain in low occipital and behind the ears. It may radiate upto the vertex. The neck becomes progressively fixed. The head is supported on both hands or on the chin and any movement is strongly resented. Torticollis was present in 12. Seven patients of this group had no neurological deficit. One of these had a stuttering neurological deficit, 11 had progressive neurological deficit and 1 had acute quadriplegia.

The next commonest presentation was that of low grade evening fever, progressive neck pain and stiffness. The x'rays were passed off as normal. Some of these were taken with the head turned, some were improperly centred and some were misinterpreted and one frank AAD was missed. In one patient the lateral tomograms were also normal. Paydirt was struck when AP tomograms were done before

referring her to the psychiatry department it showed extensive disease of the left lateral mass and a lateral AAD. All these patients subsequently presented frank atlantoaxial dislocation and progressive or acute neurological deficit. Eight patients fall into this category.

The third type of presentation is that of fever, neck pain restriction of neck movements and difficulty in swallowing. There is a bulge in the oropharynx. This is from the cold abscess which is drained in the mouth. The X'rays show no evidence of bone disease so the patients are allowed out of bed. They come back weeks or months later with progressive neurological deficit. X'rays now show frank AAD. There were two patients in this category.

In the fourth category there is a hoarseness of voice, dysphagia, tongue weakness in addition to neck pain and stiffness. These patients have multiple lower cranial nerve palsies. The limbs are usually spared. There is disease of the clivus and atlas. Careful tomograms are necessary to demonstrate the abnormality. There were three patients in this group.

In the fifth type of clinical presentation, the patients presented with fever, neck pain and a progressive quadriparesis starting from the lower limb spreading to the other lower limb then the upper limbs (e.g. LLL $\rightarrow$ RLL $\rightarrow$ RUL $\rightarrow$ LUL) with bladder and bowel involvement. This tumour like presentation was noted in 4 patients who had disease of the lamina.

There was history of low grade fever in only 10 of these 31 patients. Three patients gave a history of tuberculosis in the family. One patient had tuberculosis of the dorsal spine 15 years for which she was in a plaster jacket for 5 months. One patient had suffered from pulmonary tuberculosis in the past for which he had taken inadequate treatment. One patient had neck swelling. Eleven patients had sensory symptoms like parasthesiae and tingling and numbness. Eleven patients had complained of motor weakness involving one or more limbs. In some patients both sensory and motor symptoms became worse on flexion. Seven patients had difficulty in swallowing.

On examination all patients had a stiff neck. twelve patients had torticollis. It is difficult to examine the oropharynx because of the neck stiffness. However 14 patients had a bulge in the oropharynx. Of the 5 patients with neck swelling, one had a soft discharging sinus in the neck, 2 had cervical lymphadenitis. The other 2 had a soft tissue neck swelling possibly cold abscess. Eleven patients had no neurological deficit. 4 had only spasticity and brisk jerks. Thirteen patients had motor weakness. Eleven had quadriparesis, one had a hemiparesis and another had monoparesis. Thirteen had sensory deficit, 11 had only lateral column and all had posterior column involvement. Three patients had paralysis of the lower cranial nerves. Two had involvement of the 9th, 10th, 11th and 12th cranial nerves and another had paralysis of the 12th. The last also had spastic quadriparesis. Four patients had respiratory distress. One of these had a cardiorespiratory arrest on the way to the x'ray department and died. One other patient had a similar arrest at another hospital where he could be revived and transferred to our care. Skull traction was inserted and the patient's breathing eased and he showed some recovery.

The ESR was high in all patients except 3. Mantoux test was done in 3. It was positive in a child and negative in the other 2. The chest X'ray showed pulmonary tuberculosis in only six patients. One patient had concomitant tuberculosis of the elbow and another who had hilar lymph node enlargement had tuberculosis of the skull as well. Cervical spine X'rays included straight lateral in flexion-extension and AP views. Usually it is impossible to demonstrate the odontoid in the open mouth views. Lateral tomograms were done in flexion and extension. These are necessary to demonstrate whether the AAD is mobile. AP tomograms were done if necessary. Soft exposure may be necessary to demonstrate the nasopharyngeal mass.

When the patient is transported for X'rays extreme care must be taken to prevent any serious mishap. Two of our patients had respiratory arrest on their way to the X'ray department and one died. If deemed necessary it may be safer to send the patient with a cervical collar. Again one must not be too enthusiastic to try and take perfect radiographs at the expense of the patient's life. Flexion and extension unless done in a controlled manner and carefully may be extremely dangerous. Myelogram may sometimes be necessary. It was done in 4 patients of the present series.

Plain radiographs were done in all patients. Lateral tomograms were done in 20 and AP tomograms in 5. As many as 18 patients had atlantoaxial dislocation. Fourteen had a nasopharyngeal mass. One of these in addition had a large soft tissue mass posteriorly. In 16 the atlas was diseased. In 4 the lateral mass was diseased and 10 posteriorly. In 16 the atlas was diseased. In 4 the lateral mass was diseased and 10 had disease of the anterior arch. In 22 patients odontoid was diseased. In some the contiguous body of the axis was destroyed. In 2 there was destruction only of the base of the odontoid. Four patients had disease of the lamina. In one of these the C2-C3 vertebrae were congenitally fused (Figure 1). One had disease of the lamina, atlas and odontoid. The former 4 did not have any AAD but the last one did. He was the one to have had a respiratory arrest. Three patients had destruction of the clivus and contiguous anterior arch. Eight patients were thought to have normal radiographs. X'rays repeated weeks or months later had shown either AAD or nasopharyngeal mass. In one patient as described earlier the plain X'rays and lateral tomograms were normal. AP tomograms clearly showed destruction of the left lateral mass of the atlas and lateral AAD.

## Management

As soon as disease of the AA region was supported the patient was given a collar. If tuberculosis was suspected antituberculous treatment was started. In the earlier years streptomycin 1g. daily was given upto 90 injections and again after a break of 9 months 45 injections every alternate day were given. Isonicotinic acid hydrazide and para aminosalicylic acid were given for 2 years. In recent times our regime is as follows: injection streptomycin 1g. daily upto 90 injections, pyrazinamide 20 mg/kg for 3 months, rifamycin 10 mg/kg for 9 months, INH for 2 years, and ethambutol 20 mg/kg is started after rifamycin is stopped and continued for 2 years. Supplements of B complex especially pyridoxine are added. The collar is worn for a minimum of 1 year. X'rays are taken every three months and if healing is satisfactory the collar is discarded.

***Destruction of fused C2-3 laminae. Note occipitalised posterior arch***

In 4 patients who had a large nasopharyngeal mass causing difficulty in swallowing, the pus was drained into the mouth. Two of these were put on complete bed rest for 3 months in a cervical collar which was worn at all times. Two others who were thought to have no bony lesion were mobilised. They reappeared several weeks later, one with spastic quadriparesis and one with ataxia and mild quadriparesis. The first patient was thought to have disease of the posterior elements of the upper cervical spine. Myelogram had shown posterior compression. Laminectomy of C2-3-4 was carried out. Large amounts of extradural granulation were excised. She was put on complete bed rest and improved. However as she had persistent dislocation anterior fusion between the C1 and C2 was done by Dr. G. Sinh according to his technique [3]. The other patient was subjected to a posterior bony on-lay fusion between occiput and C2 lamina after excision of the posterior arch of the atlas. In 2

patients who had disease of the clivus a transpharyngeal biopsy was done which confirmed tuberculous infection. Both these patients were put on complete bed rest in collar for 3 months.

Twelve patients who had no neurological deficit were put on complete bed rest with a cervical collar. Six of these had no AAD and 6 had a mobile AAD. The collar was hard and the neck was kept in extension. Eating, drinking and toilet were done in the recumbent position. Before mobilising the patient lateral tomograms were done in flexion and extension to confirm healing. All patients had healed well and had no AAD. Four patients were treated with prolonged skull traction extension. Two patients were treated with skull traction for 3 months. All had mobile AAD. Three of these had neurological deficit and the fourth severe AAD. Two patients are in traction in the wards. Weights may have to be adjusted to obtain adequate reduction and to correct overdistraction if it occurs. Initially Crutchfield tongs were used in our experience but prefer to use the Gardnere Wells callipers as they are able to turn in bed to prevent the formation of bed sores.

Eight patients who had AAD and neurological deficit had posterior fusion. All of these were put on skull traction. Adequate reduction was confirmed on portable X'rays and with the traction in place posterior fusion was carried out in the prone position. Intraoperative portable radiographs were taken to confirm reduction and then fusion was performed. In 4 patients posterior fusion with wire was done. In 3 the posterior fusion was in excellent position. In one patient it was between C1-2-3 lamina. Wires were passed under each lamina separately and these were interwined and tightened. In 2 patients the wire was passed under the posterior arch and wound round the spinous process of C2. The fourth patient who had been admitted in a quadriplegic state in an orthopedic unit had improved after skull traction. She was subsequently transferred under our care. A few days later the traction slipped out as the entry wounds were badly infected. The patient immediately became quadriplegic. Traction was reinserted at a suboptimal site which did not permit full reduction of the AAD. However she showed some neurological recovery. She was taken up for fusion the next day. The posterior arch was found too far in front of the C2 lamina causing severe cord compression. Hence a wire could not be passed under it. Occipito-C2 lamina fusion were performed with wire. However an hour after surgery she died after an acute cardiopulmonary arrest. Postoperative X'rays had shown wire fixation of the occiput and C2 lamina with the dislocated posterior arch of atlas still well in front. If confronted with such a patient today we would excise the posterior arch. At that time the experience had been that excision of the posterior arch would be fatal. Postoperatively the traction was removed and the patients mobilised in a stiff collar after X'rays flexion-extension had been taken to confirm that the spine was adequately fixed. In one patient wire and stored homologous bone had been used for posterior fusion. In 2 other patients occipito-C2 lamina fusion was done using an onlay graft from the iliac crest (Figure 2). All three were given a Minerva cast for 3 months. In one patient wire and acrylic were used for posterior fusion.

- ***Occiput C1-C2 bone fusion***

In one patient who had a severe AAD satisfactory reduction was obtained on traction. Anterior fusion was attempted by laying on a silver of bone. However as the anterior arch and body of C2 were diseased this piece of bone could not be fixed. Over the next few days the patient complained of severe upper limb root pains which were thought to be due to the graft pressing upon the brachial plexus. The graft was therefore removed and under the same anaesthetic posterior fusion was performed. Anterior fusion was performed by Dr. G. Sinh in another patient who had a posterior decompression earlier. In this patient there was solid union between the atlas and axis.

In the 4 patients who had disease of the laminae of C1-2-3-4 laminectomy was done. Extradural granulation was excised extensively. In 2 there was a bead of pus. In one the C2-3 laminae were congenitally fused and atlas occipitalised. In one patient who had destruction of the anterior arch and lamina and a large anterior and posterior abscess, a posterior decompression was done by excising the foramen magnum and C1 and C2 laminae. This patient did remarkably well. In the presence of infection leaving a foreign body such as wire or acrylic was thought inadvisable hence fusion was out of question.

## Pathology

In 9 patients histology is available. In seven the tissues consisted of muscle fibres mixed with areas of necrosis resembling caseation, epitheloid cells, lymphocytes, plasma cells and Langhan's type of giant cells. In one patient infiltration with acute inflammatory cells, mainly lymphocytes, plasma cells and macrophages with vascular proliferation were seen. In this case there were no epitheloid or giant cells. Of the two patients who died in hospital, autopsy was done in one patient. The autopsy findings have been reported in detail by Pandya [2]. AFB were neither seen nor cultures in any of the cases.

## Results

One died in another hospital 2 months after she was discharged after a 2 months stay in our hospital. The cause of death could not be ascertained. One patient died in the immediate post-operative period. Autopsy was not done. One patient died whilst on her way to the x'ray department. Autopsy was done on this patient

The 11 patients who had no neurological deficit on admission had none on discharge. The patient who had respiratory arrest, anterior and posterior disease and had a posterior decompression gradually improved. On discharge he had residual quadriparesis with posterior column loss. The 2 patients who had paralysis of multiple lower cranial nerves were fed through a Ryle's tube for 3 weeks. They were then gradually able to swallow. On discharge 3 months later they were able to swallow well, speech had improved and the voice was less hoarse. Of the other 10 patients who had quadriparesis, all had recovered and were continuing to recover on discharge from the hospital 3 months later. X'rays of all patients on discharge had shown satisfactory reduction and fusion except in four who had persistent AAD. As the clinical improvement continued no corrective surgery was offered.

## Follow-up

Clinical long term follow-up is available in 19. No follow-up is available in 7. Two patients are still in the wards. The follow-up in the 19 patients ranges from 6 months to 17 years. Fifteen of these had no neurological deficits. One had residual spasticity alone. Another has residual spasticity and posterior column loss but no motor weakness. The boy with the shortest follow-up (6 months) who has respiratory arrest and quadriparesis had residual left hemiparesis and posterior column loss, but is

freely ambulant. One patient who had multiple lower cranial nerve palsies had residual hoarseness of voice despite laryngeal implants in the vocal cards because of local paralysis. All patients have mild residual restriction of neck movements.

Radiological follow-up is available in 16. In 14 of these the disease healed well nicely and there is no AAD. In 4 the diseased bone has healed but there is residual AAD. One of these had shown satisfactory reduction on the post operative X 'rays but the x'rays done lateral showed a fixed posterior AAD. The wires used for fixation had snapped. As the patient had no neurological deficit no further treatment has been offered. One other patient who had severe AAD has residual spasticity, but she is freely ambulant and able to perform all her house hold duties. Posterior decompression has been offered but she has refused surgery. The patient who had lateral AAD also has persistent AAD. But as she has no neurological deficit and only mild restriction of neck movements no further treatment has been suggested.

## Discussion

Local pain, restrictions of neck movement and acute tenderness of the upper cervical spine form the cardinal clinical picture of atlantoaxial disease. Hilton's [4] description of a similar case cannot be improved upon. "There was a exquisite pain and some tenderness along the back of the head, extending to the vertex on both sides of the midline posteriorly (along the great occipital nerves). There was pain also at the back of the ears, more behind the right ear. (Lesser occipital nerve). There was no pain in front of the ears or within the auditory canals (fifth nerve free). The head was inclined to fall forwards and had to be constantly supported. On pressing the head directly downwards upon the spine, and attempting to rotate the head upon the neck, she could not bear it, and became nearly pulseless and fainted, the limbs tremulous and agitated". The last test should not be done as it is dangerous in the extreme. Dearden's 5 description too is apt. "His only complaint was excruciating pain ..... which rendered it difficult to raise himself from the recumbent to the sitting posture, the head having to be supported by both hands".

If there is a collection of pus or granulations it causes compression of the cervicomedullary junction. If the clivus is involved the compression of the medulla oblongata leads to paralysis of the lower cranial nerves. If it bulges into the nasopharynx it causes difficulty in swallowing. The waxing and waning picture may be due to acute compression of the anterior spinal artery on flexion, which gets relieved on extension. Three patients had woken up with a wry neck and one had woken up with quadriplegia. Hilton [4] explains this phenomenon well. "The pains were maximal at night waning in the morning, when she dropped off to a short sleep. The dropping off the sleep at any time induced jumping and startling of the limbs". "It is conceivable the during one of these "jumping and startlings" the neck may have acutely flexed causing acute cervical cord compression, and compression of the occipital nerves leading to wry neck. AAD causes pinching of the cervical cord and medulla between the posterior superior part of the body of axis and odontoid and the anterior surface of the posterior arch. The mechanisms of AAD have been adequately discussed by Pandya [2], [6]. When a patient is transported for X'rays care must be taken to prevent neck flexion. Transporting a patient in a cervical column which will keep the neck extended may be safer. Plain X'rays with the head unrotated are mandatory. If possible they should be done in flexion and extension. If plain X'rays are unsatisfactory and they

usually are, lateral tomograms in flexion extension are done. If these two are inconclusive AP tomograms may be necessary before deciding that the problem is functional. They would show disease of the lateral mass of the atlas, axis and occiput. Myelogram may be necessary if the radiological features warrant it. CT scan with metrizamide has been used to study the craniovertebral region. Osborne [7] has described studies in flexion and extension with measurements between the posterior surface of the odontoid and anterior surface of the posterior arch, the atlantodental interval, the distance between the posterior arch and the anterior foramen magnum on metrizamide enhanced CT scans. CT demonstrates the relations of the lateral mass very well. We have seen this area vividly in a case of myeloma involving the lateral mass. We have not studied any patient with atlanto-axial tuberculosis with CT scan. Hilton [4] showed the importance of rest in the treatment of traumatic or infective ailment. He stressed that the only possible treatment was absolute and long continued bed rest. "A small firm pillow under the neck, two large half filled bags of sand were placed one on either side of the neck to prevent any lateral movement of the head. She was not to be disturbed from the horizontal position for any purpose whatsoever, the bowels to be relieved by enemata, the urine to be drawn off if necessary". In describing another child who had recovered some what after a fortnight's bed rest, "the nurse specially appointed to attend the child, finding that her rest at night was now so calm and quite, that she was free from pain and fever that her appetite and her power of swallowing were so much improved, as well as her temper and thinking that she was altogether so much better, and willing no doubt to mark her own penetration, as well as to please the mother by telling her in the morning what had been done by her little charge ---- this meddling and officious woman, instead of giving the child her breakfast as usual, without disturbing her head or neck in the least degree, desired the child to sit up to eat the breakfast. The child did so: the head felt forwards and she was dead". How close to death such patients are can be appreciated from this description. In older patients prolonged bed rest may be fraught with danger of deep vein thrombosis and hypostatic pneumonia.

Posterior fusion are done by passing wire beneath the laminae. This manoeuvre may damage the cord. Alexander [8] has recently described the use of hooks which are laid on the lamina to obviate this danger. Acrylic, wire or homologous bone cannot be used in the presence of infection. Autologous bone has been used safely and has ossified. We have considered a collar which keeps the head extended adequate after fixation. Some people use the Minerva plaster cast or a halo brace for 3 months. If the dislocation is irreducible then the odontoid can be excised and occipitocervical fusion carried out in the same sitting. Crockard [9] and Ransford [10] have described their technique recently for fusion in rheumatoid disease and basilar invagination. However it would seem safer to first fuse and then excise the anterior arch and odontoid otherwise the spine would be unstable unless the patient is in traction. In patients having disease of the lamina laminectomy suffices. As the anterior ligaments of the atlas and axis are intact the spine is stable.

Antituberculous treatment must be given for 2 years at least. Anything less may lead to recrudescence of infection.

## Conclusions

1. One must maintain a high index of suspicion if one is not to miss these eminently curable conditions. The Chest X'ray, may be normal, the Mantoux negative. There may be no fever and no

exposure to tubercle. Largely there may be tubercle elsewhere e.g. skull or bones and joints.
2. X'rays must be done with head unrotated.
3. If in doubt always do lateral and AP tomograms.
4. ESR is a useful prognostic investigation.
5. Oral examination must be done to look for a bulge in its posterior wall.
6. For posterior disease the treatment is posterior decompression.
7. If there is no neurological deficit, with or without AAD, uninterrupted, rest in bed for 3 months in cervical collar.
8. If there is neurological deficit, traction and fusion are advisable.
9. Oral drainage if swallowing is compromised.
10. Cervical collar for 1 year.
11. Antituberculous treatment for 2 years.


## Acknowledgements

Thanks are due to Dr. R. S. Dhir, Professor and Head, Department of Orthopaedics, KEM Hospital for permitting me to include 5 of his cases and Dr. Gajendra Sinh for permission to include one of his. My thanks to Dr. S. Bhojraj for giving me the details of the 5 patients treated in Orthopedic department. My thanks to Dr. A. P. Desai for giving the histopathology reports and slides. I also express my gratitude to Mr. Ghorpade and his team for the slides and photographs and to Mr. Godhwani for secretarial assistance. Finally my thanks to Dr. S. K. Pandya, Professor of Neurosurgery, for his encouragement and to the Dean, KEM Hospital for allowing me to present this paper.



1.Tuli S M, Tuberculosis of the craniovertebral region Clinical Orthopaedics and Related Research: *General Orthopaedics* Page: 104: 209-212, 1974

2.Pandya S K, Tuberculous atlanto-axial dislocation (with remarks on the mechanism of dislocation) *Neurology India* Page: 19: 116-121, 1971

3.Sinh G, Congenital atlanto-axial dislocations : *Neurology (India)* Page: 24: 69-76, 1976

4.Hilton J, *Rest and Pain-A course of lectures on the influence of mechanical and psychological rest in the treatment of accidents and surgical disease and the diagnostic value of pain. George Bell and Sons. London 4th Ed*1887

5.Dearden J, Case of caries of the atlas and axis.: *Lancet* Page: 2: 568, 1982

6.Pandya S K, Atlanto-axial dislocation (A review) : *Neurology India* Page: 20: 13-48, 1972

7.Osborne D, Triaolo P, Dubois P, et al, Assessement of craniocervical junction and atlanto-axial relation using metrizamide enhanced CT in flexion and extension.: *AJNR* Page: 4: 843-845, 1983

8.Alexander E, Posterior fusions of the cervical spine *Clinical Neurosurgery* Page: 28: 273-296, 1981

9.Crockard H A, Transoral decompression and posterior fusion for rheumatoid atlanto-axial dislocation *Journal of Bone & Joint Surgery* Page: 68B: 350-356, 1986

10.Ransford A O, Crockard H A, Pozo J L & Thomas N P, Cranio cervical instability treated by

contoured loop fixation.:
*Journal of Bone & Joint Surgery* Page: 68B: 174-176, 1986