

## Tuberculosis of craniovertebral junction: Role of facets in pathogenesis and treatment

Tuberculosis of the craniovertebral junction usually occurs secondary to a primary focus, elsewhere in the body. Osteoligamentous destruction and deformities leading to a range of clinical presenting symptoms have been recorded in cases of tuberculosis involving the craniovertebral junction. The understanding of the site of involvement of tuberculosis in the craniovertebral junction and the pattern of its spread and nature of its pathogenetic effects on the osteoligamentous assembly is crucial for defining the strategy of the management. It has to be understood that tuberculosis involves the bone primarily and ligaments are affected only secondarily. Bones are involved by destruction while the ligaments are involved by displacement and disruption.<sup>[1,2]</sup>

### Natural Course of Disease

The general pattern of progression of craniovertebral affection secondary to tuberculosis can be divided into three stages.<sup>[1,2]</sup>

#### Stage 1

In this stage, there is unilateral involvement of the cancellous part of the facet of atlas. There is no destructive deformation in this stage. Less frequently, there may be isolated and unilateral involvement of cancellous part of the facets of axis or the odontoid process. Inflammatory granulomatous reaction and caseous necrosis may be seen. Granulation tissue is usually located around the involved facet. The other parts of the atlas (or axis) bone and the contralateral facet are not involved.


In this stage, the patient has pain in the neck and restriction of neck movements. Systemic symptoms such as loss of weight, loss of appetite, and fever are usually present.

#### Stage 2

In this stage, the disease progresses to involve the atlantoaxial joint by destructive necrosis and inflammation. The joint involvement is a result of extension of the inflammatory reaction. The destruction involves the atlantoaxial joint complex, and in addition, it extends to other parts of the atlas and/or axis bones. Tuberculous inflammation may extend widely. The contralateral joint is still unaffected in this stage. The incompetence of the joint and osseous destruction and the adjoining ligamentous disruption in such a situation has been known to result in atlantoaxial dislocation. The atlantoaxial dislocation is probably a result of ineffectiveness of the alar and transverse ligaments as their bone attachment site is destroyed. As the contralateral atlantoaxial joint is normal, the atlantoaxial dislocation is of “fixed” and rotatory variety, and grossly, mobile and reducible dislocation is never or seldom encountered. The facet of the atlas may be displaced laterally in relation to the facet of axis or may even be collapsed. Prevertebral or extradural spinal caseous necrosis or pus formation is usually encountered. On imaging, the joint space on the involved side is seen to be reduced or absent while on the contralateral side, it is normal.

In this stage, the patient has pain in the neck, neck muscle spasm, and severe restriction of neck movements. Torticollis is characteristic and most prominent symptom. Torticollis appears to be a natural defense process, wherein the neck turns to the contralateral side in an attempt to reduce all weight bearing by the affected lateral mass and the joint and protect the cord from compression by the infective granulation.

In this stage, the patient may or may not have neurological symptoms or deficits.

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### Stage 3

The disease involves the contralateral atlantoaxial joint and other bones and joints in the region. Evidence of instability of the craniovertebral junction is usually seen.

In this stage, the patient usually has a neurological deficit.

### Indications of Surgery

Neurological deficits are notably delayed and less pronounced despite the aggressive destruction and deformation by the disease. The disease is initially unilateral and the contralateral atlantoaxial joint is spared till late in the disease process. Due to the presence of relatively stable craniovertebral region despite the unilateral facet destruction and the effectiveness of the modern antituberculous drugs, surgery can be delayed or avoided. The extent and nature of involvement of the joints are determining factors for need for any kind of surgery for stabilization of the region. Surgery for evacuation of pus or granulation tissue is seldom required. Despite the extensiveness of the bone destruction and inflammation, the patient is usually neurologically quite stable, and the systemic symptoms are not pronounced till late in the disease process. In the absence of progressive neurological symptoms and the presence of intact contralateral atlantoaxial joint, surgery can be avoided. The patient needs to be placed on antituberculous drugs. In addition, the patient will need a firm four-poster cervical collar and all activities related to neck movements and weight bearing should be avoided for at least 3 months or till there is evidence of bone healing and regression of symptoms.

Surgery is indicated less frequently and may be required in Stages 2 and 3 of the disease. The most positive indication of surgery includes presence and progression of neurological deficits. Radiological deformity in the presence of intact neurological condition may only be a marginal indication of surgery. More crucial issue is to identify and relate the problem to tuberculosis and differentiate the lesion from several tumor-like pathological situations.

If the disease is localized to the atlantoaxial region, surgery can involve fixation of the contralateral atlantoaxial joint by the lateral mass plate and screw fixation. Our experience suggests that even if the fixation is done by this method unilaterally, it is strong and provides a stable craniovertebral junction.<sup>[3,4]</sup> Incorporation of the occipital bone in the fixation is seldom necessary. However, in the presence of gross and bilateral destructive disease, occipitocervical fixation can be a useful alternative.

### Lateral Dislocation of Facet of Atlas over the Facet of Axis

In the presence of destruction of the facets of atlas and axis on one side, it appears that the alar and transverse ligaments can become unilaterally incompetent.<sup>[2]</sup> Like in cases with bifid atlas, the shift of balance on the contralateral side and the obliquity of the inclination of the facet of atlas in the atlantoaxial joint can result in its lateral dislocation over the facet of axis.<sup>[5]</sup> Distraction of the facets of the atlas and axis, reduction of lateral dislocation, and unilateral fixation of lateral masses can result in effective stabilization of the region.

### Long-term Outcome after Conservative Treatment

The effectiveness of drugs usually begins in about 3 weeks of treatment. Pain reduces and the systemic symptoms begin to abate. Over the period, the patient recovers in the neurological symptoms and deficits to varying degree. Fibrous reunion of the region and the joint usually occurs over the period, and normality of the joint movements is at least partially restored. Patients nonresponsive to drug treatment will usually continue to have progressive destructive necrosis, deformity, and instability.

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