Tuberculosis of the craniocervical junction
S. Y. Bhojraj, N. Shetty, P. J. Shah
From the P. D. Hinduja National Hospital and Medical Research Centre, Mumbai, India

Tuberculosis of the craniocervical junction is rare even where the condition is endemic. It poses problems in both diagnosis and management. We describe 25 cases followed over a period of 12 years, and relate the presentation, diagnosis and management. Of our 25 patients 16 were managed conservatively and nine by surgery. In order to diagnose this condition a high index of suspicion and advanced imaging techniques are necessary. Early diagnosis and adequate treatment led to good results without fatal complications.

Received 27 March 2000; Accepted after revision 25 August 2000

Tuberculosis of the craniocervical junction is rare and accounts for only about 1% of all cases of spinal tuberculosis. 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.

Patients and Management

Between January 1988 and January 2000 we treated 25 patients (15 men, 10 women) with tuberculosis of the craniocervical junction. At presentation their ages ranged from 18 to 75 years. Only a few had received anti-tuberculous therapy before presentation. Some had been treated for cervical spondylosis, myofasciitis, and other diagnoses with pain-relieving medication, local heat and physiotherapy.

The common symptoms at presentation were occipital headache and neck pain. Most had constitutional symptoms such as fever, loss of weight and loss of appetite, while some had vomiting as a prominent symptom. Clinical examination showed restricted rotation of the neck, with or without spasmodic tilting of the head. Local tenderness and spasm of the posterior muscles of the neck were common findings. Although retropharyngeal abscesses of varying size were present in all patients, symptomatic dysphagia occurred in only three. During clinical examination an axial-loading force or pressure applied over the head elicited pain at the affected site in all. Nine patients presented with varying degrees of neurological deficit, seven with early signs of compression of the cord (Frankel grade D) and two with severe compression causing difficulty in walking and weakness in all four limbs (Frankel grade C).

All the patients had a full blood count, and the ESR ranged from 30 to 80 mm/hr. They all had radiographs of the cervical spine to evaluate the local pathoanatomy and of the chest to identify the primary pulmonary focus. Plain radiographs of the cervical spine in all showed prevertebral bulging of greater than 7 mm at the C1-C2 region, with varying degrees of bone destruction. Evidence of atlantoaxial instability was present in nine patients. Of these, three had a fixed, irreducible, atlantoaxial dislocation (AAD) and six had a mobile, reducible AAD which was confirmed on carefully performed flexion and extension views. MRI of the cervical spine was carried out in all patients and established the diagnosis of tuberculosis by revealing the abscesses and the extent of compression of the cord, and by identifying intrinsic changes within the cord and the extent of destruction of the bone.

In the three patients who had drainage of a transoral abscess and in those who were operated on, the diagnosis was confirmed by histopathological reports showing the typical features of granulomatous disease, i.e., granulomas, central caseation, necrosis and infiltration with lymphocytes, histiocytes and Langhans-type giant cells.

The 16 patients who presented early were managed conservatively with anti-tuberculous drugs and by rigid immobilisation using an occipitomental collar. The drug treatment was continued for at least one year. The collar was worn until there was clinical and radiological evidence of healing, the average duration being four to six months.
Anti-tuberculous treatment consisted of standard doses of four drugs; isoniazid (5 to 10 mg/kg/per day), rifampicin (10 to 20 mg/kg/per day), ethambutol (15 mg/kg/per day) and pyrizanamide (25 mg/kg/per day), with vitamin supplements. Pyrizanamide was routinely discontinued after four months and the others were continued for one year.

There were nine patients who were treated surgically as they presented with bone destruction and varying degrees of spinal instability, and existing or impending compression of the cord. In one, C1-C2 wiring was carried out using Gallie’s method, but eight required occipitocervical fusion and stabilisation using a closed-loop rectangle with sublaminar or interspinous wiring and bone grafting (Fig. 1). Five of these had presented with mobile AAD and were fused in the anatomical position, whereas the remaining three had irreducible AAD which needed preoperative skeletal traction (Figs 2 and 3). In two of these reduction was achieved but one required fusion in a dislocated position.

All the patients who were operated on were kept in bed for the first five days, with sandbags on each side of the head to prevent rotation. Elevation of the head end of the bed was started from the third day and on the sixth the patient began walking with a moulded occipitomental support. These patients spent about two weeks in hospital. The collar was used until there was clinical and radiological evidence of healing, usually at between 16 and 20 weeks.

All patients were followed up regularly for a period of
one year. The minimum follow-up period in these patients was 18 months and the maximum 12 years.

**Results**

The patients who had surgical treatment had earlier resolution of symptoms compared with those treated conservatively. All patients treated surgically had excellent relief of pain with good neurological recovery. Fusion was achieved at 16 to 20 weeks, as determined by consolidation of the bone graft on plain radiographs. No patient suffered re-activation of the disease after completion of chemotherapy. All patients who had a neurological deficit improved and none deteriorated in their neurological status after surgery.

**Discussion**

Tuberculous spondylitis is common in many parts of the world, but tuberculosis of the craniocervical junction is rare, forming about 1% of all cases of spinal tuberculosis. 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. Any infection in this region is of paramount importance since delay in diagnosis or tardiness in treatment could be fatal. The spinal cord in this region in patients with tuberculosis is threatened by atlantoaxial subluxation, compression by an abscess, or by direct tuberculous invasion. The disease has an insidious onset, presenting usually with neck pain, and is often misdiagnosed in the early stages as cervical spondylitis, myositis, etc. Gross destruction and neurological deficit are evident only in the later stages.

Our experience over a 12-year period was of 25 cases of tuberculosis of the craniocervical junction. The diagnosis was based on a typical clinical presentation and characteristic findings, aided by advanced imaging techniques such as MRI. No pathological process has yet been demonstrated to produce a similar radiological and clinical picture as that of tuberculosis of the craniocervical junction. On MRI at an early stage, altered marrow signals on the T1- and T2-weighted images (hypointense on T1 and hyperintense on T2) were present, together with abnormal soft tissue with granulation tissue showing homogeneous enhancement, or an abscess showing peripheral enhancement (Fig. 4). In advanced stages of the disease there is destruction of the occipital condyles or of the lateral mass of the atlas or axis vertebra.

Extension of the disease into the spinal canal can result in abnormalities of the cord caused by mechanical compression, or by ischaemia. When there is gross destruction, subluxation or dislocation of C1-C2 also occurs (Fig. 5). It is easier to demonstrate these changes on MRI rather than CT because of its multiplanar capability and better soft-tissue contrast resolution.

The management of this condition depends on the extent of destruction. If diagnosed early, it can be effectively treated conservatively by anti-tuberculous drug therapy for one year and with a chin-to-occiput collar support (Figs 4 and 6). In advanced disease in which extensive destruction has occurred, the stability of the atlantoaxial joint is lost (Figs 2 and 5). In such cases, or in those with neurological
deficit or impending compromise of the cord, surgical stabilisation with fusion posteriorly is required, with or without transoral decompression.

It is important to note that care should be taken when draining large and tense retropharyngeal abscesses, since sudden evacuation or decompression of the abscess could lead to neurological deterioration as a result of sudden loss of the stability afforded by the abscess.

In nine cases in our series which had AAD with neurological deficit or impending compromise of the cord, posterior stabilisation with fusion was achieved. Patients with mobile AAD did not need preoperative traction but those with fixed AAD required reduction with preoperative traction. In one of these reduction was not achieved and fusion in the dislocated position was required. Whenever fusion was performed in the dislocated position the C1 arch was not instrumented because of the potential risk of neurological damage. In patients with persistent dislocation and residual neurological deficit, there is a need for anterior transoral decompression of the dens, although this was not necessary in our patients. The number of cervical segments fused below the axis was assessed by the quality of bone and the purchase of the wires at the various sites during surgery.

Conservative management for postinfective AAD,6 and a regime of bed rest, skull traction and anti-tuberculous treatment with mobilisation only after six to nine months,4 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.7

Neurological complications are more common and serious in tuberculosis of the craniocervical junction than when other vertebrae are involved. Progressive neurological deterioration has been documented in patients treated without decompression or stabilisation, and sudden death can occur.8-10

Posterior fusion allows rapid neurological recovery, prompt relief of pain and a high rate of fusion with minimal morbidity5 (Figs 1 and 7). In view of this, and the early healing and stability afforded by fusion, we recommend that patients with instability and neurological compromise should be stabilised by means of a posterior fusion.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References