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Infection

Management of craniovertebral junction tuberculosis

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Abstract

Background: Tuberculosis of the craniovertebral junction (CVJ) is extremely rare. However, recent evidence suggests that the incidence of this condition may be increasing. The diagnosis is often difficult despite advances in imaging using magnetic resonance imaging. The transoral approach to the anterior CVJ provides excellent access to this region, has low mortality and morbidity, and enables biopsy of lesions and decompression of the neuraxis. Management of associated atlantoaxial instability, with regard to timing and method of stabilization, is controversial.

Methods: We report 24 cases of CVJ tuberculosis. Prominent manifestations of the disease included neck pain and stiffness, swelling of the retropharyngeal soft tissues, quadriparesis, osteolytic erosions, and atlantoaxial subluxation. Seven patients had acute presentation. All patients received antituberculous drug treatment for 18 months. Four patients were managed conservatively, 5 underwent only transoral biopsy, 9 patients underwent transoral decompression and posterior fusion, and 6 patients underwent only posterior fusion.

Results: Histopathologic analysis of biopsy material revealed abscess only in 5 cases, granulation tissue only in 6, abscess with granulation tissue in 4, granulation tissue with chronic osteomyelitis in 5, and chronic inflammation in 2. All patients improved, with mean improvement in Nurick grading of 1.71. Even patients with spinal cord signal intensity changes on magnetic resonance images showed improvement.

Conclusions: Although CVJ tuberculosis 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.

Keywords: Craniovertebral junction; Tuberculosis; Spinal tuberculosis

1. Introduction

Tuberculosis remains a major health problem in many developing countries. With the increase in incidence of acquired immune deficiency syndrome and intercontinental travel, central nervous system tuberculosis is becoming a major problem in developed countries as well. Tuberculosis of the craniovertebral junction (CVJ) is rare, comprising less than 0.3% to 1% cases of tuberculous spondylitis [3]. Early diagnosis and treatment are important in the prevention of long-term neurological sequelae. Transoral surgery has been advocated in both diagnosis and treatment of CVJ tuberculosis [3]. Antitubercular chemotherapy is an essential component of management. It is now well recognized that surgery plays an important role in decompression of neuraxis and diagnosis in these cases, although the method of surgical stabilization remains controversial [1-5,7].

2. Methods

During the period from 1987 to 2001, 24 patients (15 men and 9 women, with age ranging from 8 to 78 years; mean, 35.5 years) with CVJ tuberculosis who were managed at the National Institute of Mental and Neurosciences were followed up prospectively. All patients were assessed clinically and were investigated with plain

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x-rays of the CVJ, including lateral flexion/extension views to look for instability. In the initial part of the study, patients underwent postmyelography computed tomography (CT) scan; later, magnetic resonance imaging (MRI) was done to look for spinal cord compression. All patients received antituberculous drug therapy. Appropriate surgical intervention was done depending upon the extent of the disease and instability. All patients were followed up for a period ranging from 1 to 36 months (mean, 9.3 months).

3. Results

3.1. Clinical presentation

The most common clinical syndrome was of neck pain and progressive spastic quadriparesis. Mean duration of symptoms was 5.6 months. Only 7 patients had acute presentation in the form of rapid deterioration of motor strength, of which 6 also had sphincter disturbances. One patient presented with dysphagia due to retropharyngeal abscess. He was discharged after drainage of abscess and later presented with neck pain and progressive spastic quadriparesis. The detailed analysis of clinical features is given in Table 1.

3.2. Imaging findings

All patients underwent plain x-ray of the CVJ including flexion/extension studies on lateral view to look for instability. Increased prevertebral soft tissue shadow was seen in 2 (8.7%) cases (Fig. 1). Fourteen patients had atlantoaxial dislocation, of which 10 (41%) had reducible and 4 (16%) had irreducible type. Computed tomography scan of the CVJ was done in all patients. On the basis of CT scan results, the patients were categorized by Lifeso grading [11]. Two (8%) had grade I (increased prevertebral shadow) involvement, 16 (66%) had grade II (atlantoaxial dislocation and early bony changes) involvement, and 6 (25%) had grade III (atlantoaxial dislocation, gross destruction of bone, and pathological fractures) involvement (Fig. 2). Besides the above patterns of disease, 3 (12%) patients also had destruction of posterior atlantal arch. Magnetic resonance imaging done in 11 (45%) cases showed significant extradural granulation tissue compressing the upper cervical spinal cord anteriorly. There were 2 cases in which the extradural granulation tissue and abscess extended posteriorly (Fig. 3). Three (12%) cases also had secondary intrinsic cord signal intensity changes.

Table 1
Clinical features of CVJ tuberculosis

Symptoms and signs	Number (%)
Neck pain	24 (100)
Neck tilt	1 (4)
Dysphagia	1 (4)
Sensory deficits	13 (54)
Spastic quadriparesis	16 (67)
Sphincter disturbances	6 (25)

Fig. 1. Plain x-rays of CVJ showing increased prevertebral soft tissue shadow opposite C1-C3 suggestive of retropharyngeal abscess. There is also partial destruction of C1 anterior arch and odontoid process.

3.3. Histological diagnosis

Two patients underwent CT-guided biopsy and 2 underwent transoral open biopsy. In 16 cases, tissue was obtained during transoral decompression, and in 2 cases, tissue was obtained during posterior occipitocervical fusion. Two

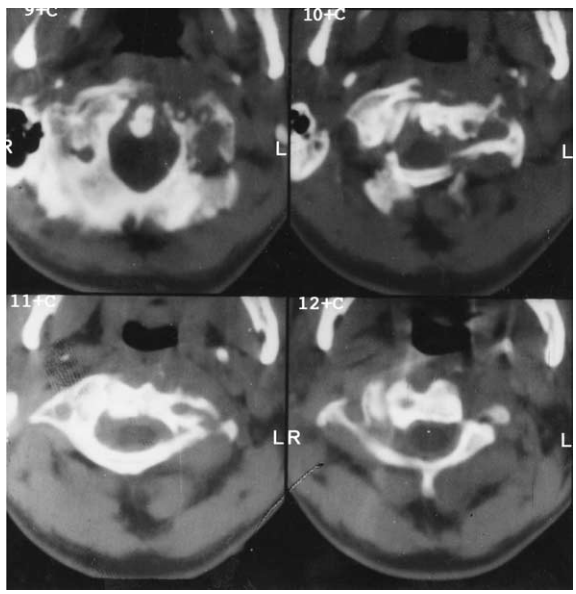

Fig. 2. Axial CT scan of CVJ showing extensive destruction of anterior arch of atlas, dens, and part of axis correlating with Lifeso stage III CVJ tuberculosis.