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CASE REPORT

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Hemiplegic/monoplegic presentation of cervical spine (C1-C2) tuberculosis

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Abstract Tuberculosis of the craniovertebral region is very rare. Neural deficit in this region is reported in between 24% and 64% of cases, and mainly takes the form of quadriparesis. Hemiplegic and monoplegic presentation among this set of patients is rarer. Out of 32 patients treated at our institution between May 1989 and February 2001, only one had hemiplegia, while two had monoplegia. These three cases are discussed. Case 1 involved a 45-year-old woman who presented with hemiplegia following a trivial fall. Plain radiographs and computed tomographic (CT) scans of the skull appeared normal, but CT scans of C1-C2 and the craniovertebral junction revealed destruction of the dens and atlantoaxial subluxation. The patients in cases 2 and 3 had monoplegia. Plain radiographs in both cases showed an increased prevertebral soft tissue shadow in front of C1-C2. CT in case 2 and magnetic resonance imaging (MRI) in case 3 revealed destruction of the arch of C1 and the dens, with subluxation. All three patients were successfully treated with rest, skull traction, anti-tubercular drugs and suitable braces. Case 3 required stabilization. All three patients achieved complete neural recovery. Patients 1, 2 and 3 had 22, 48 and 4 months' follow-up respectively. Patient 3 was subsequently transferred to a neurosurgery ward for stabilization of the occipito-C3 vertebrae. Hemi/monoplegic presentation is extremely rare; no author in the literature is able to give reason for the rarity or the pathomechanics of the condition. We believe that if medullary cervical junctional involvement extends slightly higher (in rare circumstances), with involvement of one of the branches of the vertebral or lower basilar artery, medial medullary syndrome will occur, sparing medial lemniscus and emerging hypoglossal nerve fibres. Thus the pyramids will be involved, causing contralateral hemiparesis, and if the pyramids are selectively involved, it will cause contralateral monoparesis.

Keywords Hemiplegia · Monoplegia · Tuberculosis of spine · Cervical spine · Atlanto axial junction

Introduction

Tuberculosis of the craniovertebral region is very rare and is reported in less than 1% of all cases of spinal tuberculosis [8]. It is nonetheless the most common infective lesion of the craniovertebral region. Most patients present with pain, limitation of neck movement, local tenderness, tilt of the neck due to spasm and a tendency to support the neck. They may have difficulty in breathing, talking or swallowing due to a large prevertebral abscess. Neurological deficit of varying degrees can be detected in 24–64% of cases at presentation [2, 3, 4, 5, 8]. Most of the patients with neurological deficit have either quadriparesis or

quadriplegia. In a series of 31 patients [4], 11 had quadriparesis and one each had hemiparesis and monoparesis. The rarity of hemiplegia or monoplegia as a presentation of tuberculosis of C1-C2 makes these cases worth reporting.

Case report

Thirty-two cases of C1-C2 tuberculosis were studied between May 1989 and Feb 2001. Of these, 22 had neural deficit; 19 patients had grade I–III quadriplegia, one had hemiplegia and two patients had monoplegia. We present the latter three cases.

Case 1

A 45-year-old woman was admitted to the medical ward with complaints of sudden-onset right-sided hemiplegia following a trivial fall. She did not give any history of neck pain. On clinical examination she had tenderness in the upper cervical region. Her neck movements were restricted. She had grade 2 power in her right shoulder, elbow and wrist, and grade 0 power in her right lower limb. She had upper motor neuron signs in the right lower limb. Findings of a neurological examination of her left upper and lower limb were within normal limits. There was no sensory loss, but she had urinary retention. Results of haematological investigations were within normal limits. Findings of computed tomography (CT) imaging of her skull were also within normal limits. On plain radiography, the cervical spine appeared essentially normal. The CT scan of the craniovertebral junction and C1-C2 revealed destruction of the dens with a sequestrum in the centre, which gave an appearance of an irregular, soft, feathery, coke-like sequestrum ("image en Grelot") [9], and significantly increased atlanto-dental space (8 mm), consistent with atlantoaxial subluxation (Fig. 1, Fig. 2). Skeletal traction of 0.5 kg was applied through crutch field tongs, and four-drug anti-tubercular treatment comprising INH, rifampicin, ethambutol and pyrazinamide was started. After 6 weeks of recumbency, graduated ambulation with a suitable brace (four-post collar) was started. The patient continued with uninterrupted multidrug antitubercular therapy for 18 months. She showed complete recovery of motor power in 6 months. At 22 months of follow-up, the patient did not require fusion, as her spine was stable.

Fig. 1 Reformatted computed tomography (CT) scan of the craniovertebral junction and C1-C2, showing destruction of the dens with a coke-like sequestrum in it, in a 45-year-old woman (case 1)

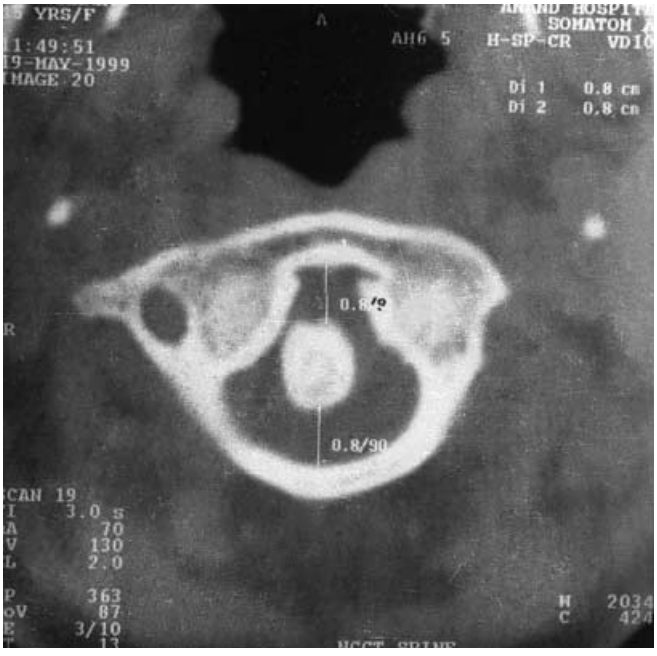

Fig. 2 Axial cut of C1-C2 showing significantly increased atlanto-dental space (8 mm) consistent with atlantoaxial subluxation, in a 45-year-old woman (case 1)

Case 2

A 23-year-old man presented with pain and restriction of neck movements of 2 months' duration. He developed gradual onset of weakness in the left upper limb. On examination, he had grade 2 power around the shoulder, elbow and wrist, with no sensory deficit. Plain radiographs showed increased prevertebral shadow (21 mm) in front of C1 (Fig. 3). CT scan revealed destruction of the right lateral mass of C1, with subluxation of C1 on C2 (Fig. 4). The patient was treated with head-halter skull traction, rest and multidrug anti-tubercular therapy. After 6 weeks of recumbency (with near complete motor recovery), graduated ambulation was started with a suitable brace. The patient continued on uninterrupted multidrug therapy for 18 months. After 3 months of treatment, lateral views of the cervical spine in flexion and extension showed the spine to be stable. At 24 months, the brace was discarded. After a total follow-up of 48 months, the patient has achieved complete neural recovery and a stable spine.

Case 3

A 20-year-old female patient was admitted with a 1-year history of swelling in the neck (right-sided), which had started discharging pus within 1 month of its appearance. The patient had gradually developed pain and restriction of neck movement. For the 2 months prior to admission, the patient had experienced left upper limb weakness and an increase in severity of pain following a trivial trauma. On clinical examination, she had a 1×0.5-cm puckered scar in the right supraclavicular region. There was tenderness in the upper cervical region, with gross restriction of all neck movements. Her left upper limb had grade 3 power at the shoulder, elbow and wrist, while the findings of neurological examination of

Fig. 3 Plain radiograph (lateral view) of the cervical spine of a 23-year-old man, showing increased prevertebral soft tissue shadow opposite the upper cervical spine (case 2)

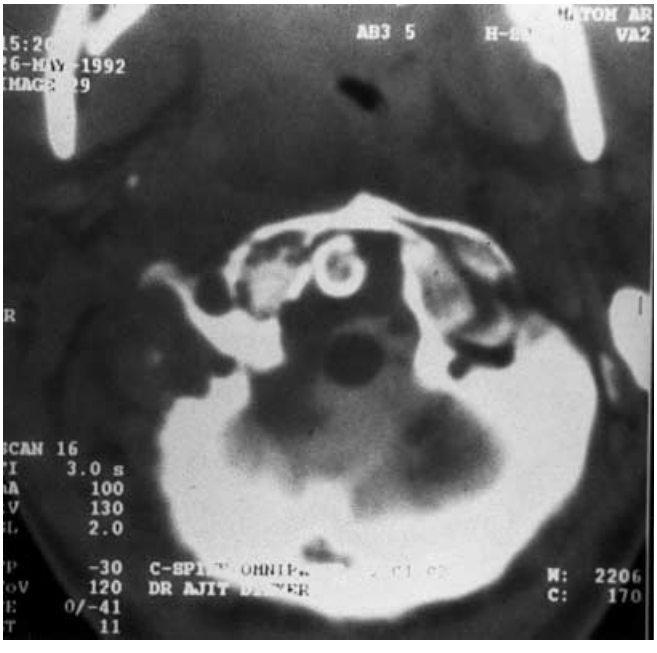

Fig. 4 CT scan (axial cut at C1-C2) of the same patient as in Fig. 3, showing destruction of right lateral mass with subluxation of C1 on C2

the other limbs were within normal limits. Haematological investigations revealed an ESR of 36 mm in the 1st hour, with other results being within normal limits. The cervical spine plain radiograph (lateral view) revealed increased prevertebral shadow, with destruction of the arch of atlas and odontoid process, resulting in dislocation of C1 over C2 (Fig. 5). Magnetic resonance imaging (MRI) revealed destruction of C1 and C2, with forward displacement of the destroyed odontoid and cord compression (Fig. 6). The patient was treated with head-halter traction and four-drug anti-tubercular therapy. After 6 weeks of recumbency, skull traction and multidrug chemotherapy, the patient showed complete neural recovery. Plain radiography revealed marked displacement of C1 on C2. She continued to lie in bed, on skull traction and anti-tubercular therapy. At 3 months, lateral plain radiographs in flexion and extension revealed an unstable spine (Fig. 7). She was transferred to the neurosurgery department, and occipito-C3 fusion using autologous grafts and wires is planned.

Fig. 5 Plain radiograph (lateral view) of the cervical spine of a 20-year-old woman showing increased prevertebral shadow, destruction of C1 and C2 and forward dislocation of C1 on C2 (case 3)

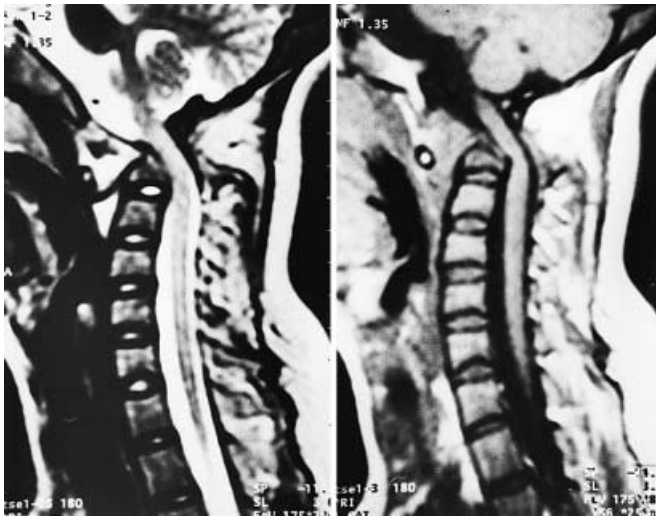

Fig. 6 Magnetic resonance image of a 20-year-old woman, showing destruction of the dens, atlantoaxial dislocation and kinking of the cervicomedullary junction (case 3)

Fig. 7 Radiograph (lateral view) of the cervical spine of the same patient, taken in flexion and extension at 3 months, showing instability of the spine

Discussion

The clinical presentation of hemiplegia or monoplegia in tuberculosis of the atlanto-axial region is rare. What is true of spinal tuberculosis in general is also true of the craniovertebral region. This condition is noted most commonly during the first three decades of life, and is equally distributed across both sexes [7]. The reported incidence of neurological deficit in such cases ranges from 24% to 64% [2, 3, 4, 5, 8], with most patients presenting with either quadriparesis or quadriplegia. Clinical presentation with hemiplegia or monoplegia is rarely seen in cases of upper cervical spine tuberculosis. Pandya reported one case of atlanto-axial tuberculosis presenting with "mild right limb weakness" in 1971, while in a series of 31 patients with atlanto-axial tuberculosis, reported by Karapurkar, only one had hemiplegia and another monoplegia [4, 6]. Jain et al. reported two cases of monoplegia in a series of 11 cases of C1-C2 tuberculosis [3].

Tuberculosis of the upper cervical spine seems to begin either in the retropharyngeal space, with secondary involvement of bone, or rarely in the bone itself. With progression there is increasing ligamentous involvement with minimal osteolytic erosions into the odontoid or into C1. This allows anterior subluxation and proximal translocation of the odontoid. Tuberculosis of C1-C2 has been classified into three stages by Lifeso [5]. This classification system has a bearing on the treatment and the type of surgery required.

1. Stage I: minimal bone destruction with intact ligaments and no evidence of anterior displacement of C1 on C2, with or without proximal translocation of the dens – Patient seldom requires surgery
2. Stage II: minimal bone destruction with ligamentous disruption and anterior displacement of C1 on C2 with or without proximal translocation of the odontoid – Patient may require fusion (Allie type with autologous bone graft)
3. Stage III: marked bone destruction with complete obliteration of the anterior arch of C1 – When fusion is required, it is done between the occiput and C2-C3

At the craniovertebral junction, the spinal cord is compressed by tubercular abscess, granulation tissue, tubercular debris, atlantoaxial subluxation, upward translocation the dens, tubercular invasion of the cord, or vascular ischaemia due to local tuberculous pathology [2]. Our case 1 falls into stage II, with atlantoaxial subluxation. Subluxation of the dens on one side represents a quantum of compression on one side. Cases 2 and 3 are in stage III, where destruction of lateral mass occurred, with forward subluxation of the dens and compression of the cord. An unequal pressure on the cord and medullary cervical junction, or just above it, on either side can rarely present as hemiplegia or monoplegia. Rarely, a medial medullary syndrome occurs, in which the pyramid becomes involved, sparing medial lemniscus and emerging hypoglossal nerve fibres, causing contralateral hemiparesis and sparing sensory involvement [10]. Further, if there is differential involvement, monoparesis/plegia can occur. This is rare, because it will occur only if a branch of vertebral or lower basilar artery is involved, or if there is ascending inflammatory oedema just above the medullary-cervical junction, pressing on an area of decussating pyramidal tracts and sparing medial lemniscus and emerging hypoglossal nerve. Thus, it will always involve the opposite side and spare sensations. This is one of the few atypical presentations of tuberculosis of spine reported in the literature. To the best of our knowledge, only one case of C1-C2 tuberculosis leading to hemiplegia has been reported in literature. Other atypical presentations include:

1. Involvement of the posterior elements of the spinal column with complete sparing of the anterior elements (vertebral bodies and discs)
2. Skip lesions separated far apart to involve the two extremities of the spine
3. Extradural spinal cord compression without radiographic evidence of bony involvement
4. Destructive lesions of the sacrum with palpable pelvic mass [6]

The incidence of such atypical presentations of spinal tuberculosis is reported to vary from 0.2% to 10% [1].

Tuberculosis of C1-C2 should be kept in differential diagnosis of hemiplegia/monoplegia in endemic areas of tuberculosis.

Recognition and prompt management of these atypical forms will help in reducing the continuing morbidity of this disease due to serious secondary effects on the cord, which may result from a delay in making the correct diagnosis.

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