Tuberculosis of the craniovertebral junction

Muhammad Asad Qureshi · Waseem Afzal ·
Ahmed Bilal Khalique · Ibrahim Farooq Pasha ·
Max Aebi

Received: 21 June 2012/Revised: 25 August 2012/Accepted: 29 August 2012/Published online: 5 October 2012
© Springer-Verlag Berlin Heidelberg 2012

Abstract

Purpose Tuberculosis (TB) of craniovertebral junction (CVJ) occurs in 1–5 % of cases of TB spondylitis. This can be a life-threatening condition due to mass effect of infective process or resultant instability. Surgical indications for TB of CVJ are not clear from literature.

Methods We have reviewed all the patients with TB of CVJ admitted at our center between 2005 and 2010.

Results There were 15 patients including 10 males and 5 females. Average age was 38 years and average duration of symptoms was 8 months. All patients were started on multidrug antituberculous chemotherapy and skull traction. Those patients who failed to respond in 4–6 weeks and had persistent instability or neurological deficit were offered surgery. Rest was treated conservatively by immobilisation or traction. All five patients who were surgically treated had occipitocervical fusion (OCF) with titanium screws and plate/rod construct combined with posterior decompression if needed. Only one patient needed anterior surgery in addition to OCF at a later stage. All patients improved neurologically whether they were treated surgically or conservatively. Only difference was that surgically treated patients had earlier pain relief, mobilisation, neurological improvement and lesser complications.

Conclusion We recommend that all patients with TB of CVJ with instability and neurological compromise, who

fail to respond to 4–6 weeks of antituberculous chemotherapy and skull traction should be offered occipitocervical fusion with or without posterior decompression. Anterior surgery will be needed only in those few cases who do not improve neurologically after OCF.

Keywords Tuberculosis · Caries spine · Potts disease · Craniovertebral junction · Occipitocervical fusion

Introduction

Tuberculosis (TB) of the craniovertebral junction (CVJ) is a life-threatening condition. This can lead to abscess or granuloma formation with or without instability at the CVJ. Both of these conditions can cause compression of the upper cervical spinal cord and brain stem, leading to tetraparesis and bulbar symptoms. It is not known whether it is an isolated disease or associated with manifestation of tuberculosis in other organs of the body. It is also not clear from the literature how many of these patients had pulmonary tuberculosis in the past and how long afterwards the craniovertebral junction gets involved and why.

The management of this condition is not very clear from literature as only retrospective reviews and case reports are published so far. Some papers suggest surgery in all patients while other produce similar outcome with conservative management [1–3, 5, 6, 8]. It is not clear whether surgery should be anterior, posterior or combined. No clear guidelines are available about use of implants in CVJ TB. We have reviewed all the cases admitted with CVJ TB at our centre and the literature with a view to present a management protocol for this disease.

M. A. Qureshi (✉) · W. Afzal · A. B. Khalique · I. F. Pasha
Spine Unit, Orthopedic Department,
Combined Military Hospital, Rawalpindi, Pakistan
e-mail: asad.qureshi@yahoo.com; maqpg@yahoo.com

M. Aebi
University of Bern, Bern, Switzerland

Patients and methods (Table 1)

This study was carried out at the Spine Unit, Orthopaedic Department, Combined Military Hospital, Rawalpindi, Pakistan. Data of all patients admitted with TB of the spine between 2005 and 2010 was retrieved from Spine Database. Out of all these patients, those having involvement of the craniovertebral junction (occiput-C2) were selected.

Craniovertebral junction tuberculosis was diagnosed on the basis of history, clinical examination, raised ESR, positive Mantoux test, positive findings on MRI or CT scan of the craniovertebral junction and transoral biopsy of the lesion. Surgery was offered to patients showing signs of instability at the craniovertebral junction with neurological compromise. Patients were called for follow up and a structured interview was performed to find out the following parameters:

1. Management done
2. Type of surgery and implants used in operated cases
3. Regimen and duration of ATT
4. Duration of follow up
5. Complications
6. Change in pain
7. Change in neurology
8. Stability at the craniovertebral junction
9. Patient satisfaction

A structured patient satisfaction form is impossible to give to the patient, since quite a number of these patients are uneducated and even speak different languages therefore the

communication and the maintenance of the communication is often very difficult. All patients' satisfaction outcome parameters have been evaluated in an individual interaction between one of the staff surgeons and the patients.

Results

Out of 310 patients admitted with spinal tuberculosis, 15 patients had involvement of the craniovertebral junction (4.8 %). There were ten males and five females, average age was 38 years. Average duration of symptoms before onset of treatment was 8 months. No patient had a history of pulmonary or other organ TB in the past. Two patients had multi-level spinal tuberculosis. All patients had raised ESR, positive Mantoux test and signs of infection at the craniovertebral junction on MRI/CT scan.

Of all transoral biopsies only one patient grew *Mycobacterium tuberculosis* on culture. All the rest had only positive histological findings of chronic granulomatous inflammation suggesting tuberculosis disease.

All patients had anti-tuberculous chemotherapy with four drugs for 2 months (Rifampicin, Isoniazid, Myambutol and Pyrazinamide), followed by three drugs for 10 months (Rifampicin, Isoniazid and Myambutol) Standard Regimen (SR). Four patients did not show improvement in ESR and CRP on this regimen and in them two more drugs were added (Levofloxacin and Amikacin) for 4 months Modified Regimen (MR) and ultimately their ESR and CRP were also improved.

Table 1 Craniovertebral junction TB patients; demography and management

S. no	Age (years)	Gender	Duration of symptoms (months)	Level	Asia impairment scale	Management	Follow-up (years)
1	23	Male	3	O-C2	C	OCF	2
2	38	Male	12	O-C2, L5-S1	C	OCF	2
3	43	Male	22	O-C2	D	OCF	4
4	32	Female	5	O-C2	C	OCF	6
5	28	Female	6	O-C2	D	OCF + AD	4
6	15	Female	24	O-C2	D	PHIL	1
7	75	Male	3	O-C3	D	PHIL	1
8	48	Male	1	O-C2	C	PHIL	1
9	23	Male	6	O-C1, C5-6	E	PHIL	2
10	47	Female	12	O-C2	D	TONGS	2
11	45	Male	6	O-C2	D	PHIL	2
12	24	Male	6	O-C2	E	TONGS	4
13	14	Male	6	O-C2	E	TONGS	5
14	32	Male	2	O-C2	D	PHIL	6
15	85	Female	5	O-C2	C	PHIL	1

OCF occipito-cervical fixation, AD anterior debridement, PHIL Philadelphia collar, TONGS cervical tongs traction

Patients having instability, i.e., atlantoaxial dislocation or rotatory dislocation or even lateral translation which are not fixed and having neurological deficit at the cervicovertebral junction as well, were primarily placed on traction and offered surgery if they did not improve in 4–6 weeks.

For traction treatment, patients were placed in bed and Gardner Well's tongs were applied. Main contra-indication to use of traction treatment was local wound or infective focus at site of pin placement. Initially 3 kg weight was used. Reduction was checked by weekly lateral X-rays of cervical spine. If there was inadequate reduction, 1 kg weight was added every week for a maximum of 6 kg or less until reduction was achieved. Pin tracts were daily inspected and cleaned with antiseptic solution. Main problems on traction were minor pin tract infections and loosening which were treated by local antibiotics and re-tightening of pins. At end of 4–6 weeks, traction was removed and stability was checked by flexion/extension films of cervical spine. For persistent instability, either surgery or Halo vest immobilization was offered to the patients.

Five of these patients were finally treated surgically, partially because they were not willing to be immobilised with Halo and the stability could not be maintained after removing the traction. In a specific case, the pathology was combined with a non-improving neurological deficit and neck pain and stiffness. The other ten patients were treated conservatively with a Philadelphia Collar for up to several months. In all operated cases, OCF (occipitocervical fusion) was performed, using titanium plates and screws and posterior decompression was added (Figs. 1, 2). The plates were fixed in the subaxial spine with lateral mass

screws and preferentially in the midline of the occipital bone. If possible, the segment C1/C2 was included in this fixation with a specific transarticular screw C2 to C1 according to Magerl.

Only one patient needed second stage anterior decompressive surgery by high cervical retropharyngeal approach. No transoral surgery has been performed.

All patients recovered fully, on last follow-up, which ranged from 1 to 6 years with an average of 3 years.

All patients were satisfied with the management. Patients who were treated surgically, had on average, earlier pain relief and mobilisation as compared to conservatively treated patients. There were no major complications. Four patients developed pressure sores, specifically during the time of traction. One patient with second stage anterior surgery had minor discharge from the wound which settled without any further surgery. Three more patients with instability at the craniovertebral junction were offered surgery but they refused surgery and preferred traction, which was continued for more than 3 months and they also improved ultimately.

Discussion

This rare entity of tuberculous involvement of the craniovertebral junction occurs according to the literature in only 0.3–1 % of patients with tuberculosis/tuberculous spondylitis [9]. However, in our own series as well as in another series, 5 % of spinal tuberculosis was at the craniocervical junction [3]. On one side there are a lot of case descriptions and several case series with more or less cases, all of them

Fig. 1 **a** MRI showing TB abscess at CVJ. **b** Positioning during surgery. **c** Intra-operative picture showing two titanium recon plates used for occipitocervical fusion

Fig. 2 Post op AP (a) and lateral X-rays (b)

with some recommendations regarding classification [19], the establishment of diagnosis and the value of biopsy [13, 16, 18–21]; furthermore, their recommendations are also related to treatment modalities [15, 16] and to indications for treatment. Since craniocervical junction tuberculosis carries the risk of secondary instability and severe neuraxis compression [5], sometimes in the sense of severe bulbo-medullary complications [2] with painful torticollis, dysphagia and tetraparesis, early diagnosis is of utmost importance. In rare cases of patients with neurological deficit due to craniocervical junction tuberculosis, a hemiplegia or a monoplegia may occur. The cause may be an involvement of the pyramids with a contralateral hemiparesis or monoparesis. The reason for that may be an affection of the basilar or vertebral artery.

Most authors recommend CT-scan and/or MRI for the craniovertebral junction [4, 7] followed by the histology and bacteriology of the specimen gained usually through transoral biopsy of the retropharyngeal abscess. In our own material, however, we were unable to grow *Mycobacterium tuberculosis* on culture tests in all cases but one. But all had typical histomorphological findings of chronic granulomatous inflammation with giant cells present. We have no explanation for that, but it could be that these patients, coming sometimes from far rural areas, may have had already some tuberculostatic medication because of preceding lung tuberculosis which the patient has forgotten and which was not documented. Some authors recommend open posterior biopsy of the dens [10, 19] through the C2-pedicle. Several authors do not report any preoperative

histological results from the diseased craniovertebral junction, in contrast, diagnosis was made by CT/MRI examination and the accompanying clinical history only [2, 7, 18].

In spite of high probability of not being able to prove mycobacterium in the biopsy in our material, in contrast to other authors, we have treated those patients as tuberculous disease at the cervicocranial junction and all of them showed a cure in the follow-up period between 1 and 6 years. This proves that the treatment indication was adequate, although the bacteriological evaluation was not conclusive.

Literature is quite divided on management of patients with tuberculosis involvement of the craniovertebral junction [4, 7, 9–12, 14]. Some people recommend conservative treatment only [1, 2, 5, 11, 22], others recommend surgery in all patients [3, 6, 8, 23]. Some have shifted their management protocols from surgical to conservative and vice versa. Our study has also shown that all patients with tuberculosis at the craniovertebral junction improve on medical management. In agreement with most of the literature, our study has also shown that the only indication of surgery is persistent instability combined with neurological deficit which is not responding to medical management and skull traction after 4–6 weeks. Most of these patients improve neurologically just by reduction and stabilisation and if necessary by open posterior decompression. Rarely in our material anterior decompressive surgery is needed after posterior stabilisation, specifically if improvement fails. Standalone anterior decompressive surgery is not

recommended in our opinion as it adds to instability and it is difficult to stabilise the craniovertebral junction anteriorly.

Most recent studies are not conclusive whether non-surgical treatment is superior to surgical treatment or the other way around. Although our study was retrospective, the decision making regarding the treatment strategy was driven by the clinical presentation. A surgical treatment was only necessary in a minority group of patients with craniocervical junction tuberculosis, i.e. one out of three patients; their treatment consisted of immobilisation, be it by Philadelphia Collar and/or traction and adequate chemotherapy, although two-thirds of the patients had neurological deficits. Our conclusion is that even in neurological symptomatology surgery is not absolutely necessary if the doctor and the patient are willing to sit the time out until it comes to a resolution of the disease. It is rather the mechanical musculoskeletal instability which may lead to a non-resolving neurological deficit and may demand for a surgical procedure. The tuberculous disease may lead to a complete destruction of the ligamentous/bony structures which will then be unable to carry the “heavy head” and need the mechanical stabilisation.

The sum of major studies about exclusive surgical treatment of craniocervical junction tuberculosis includes patients who fulfil the criteria of instability, others do not [6, 8, 9, 17, 22, 23]. Arunkumar et al. [6] proposed surgical treatment in spinal tuberculosis only in cases with neurological deficits due to craniovertebral junction disease. The neurological deficit may be due to reducible atlantoaxial dislocation (AAD), irreducible atlantoaxial dislocation, atlantoaxial lateral dislocation [13], basilar impression alone or with atlantoaxial dislocation and C2 vertebral body destruction without dislocation and with neurological symptomatology besides neck pain and torticollis. This neurological symptomatology usually includes: progressive limb weakness (in the majority of the cases), sensory symptoms (in about one-fifth of the patients) and urinary dysfunction (in about one-third of the patients). These authors recommend anterior transoral odontoid resection (in more than half of the cases), or transoral abscess decompression (in one-fifth of the patients) and transoral abscess decompression with partial odontoidectomy followed by a posterior occipitocervical fusion (usually from C0 to C3) [13]. This occipitocervical fusion usually is done with rods and sublaminar wiring. Our experience, however, taught us that anterior surgery is not necessary in appropriate posterior stabilisation from the occiput to C2 and/or C3 with plate/rod and screw constructs, whereas C1/C2 is posteriorly blocked with the transarticular screw fixation according to Magerl (Fig. 2). In only one case, an anterior retropharyngeal (not transoral) debridement was done.

Stable posterior construct does sufficiently immobilise the infection focus, so that, together with antituberculous chemotherapy, the infection may heal with secondary ossification. Behari, too [9] favours cervical treatment in mechanical instability and neurocompression, however, in less than 50 % of the cases with craniocervical junction tuberculosis. The surgical procedure was a single step anterior debridement and decompression and posterior fusion compared against a group of patients with simple posterior fusion and stabilisation alone [9]. The latter is proposed for patients who have a reducible atlantoaxial dislocation.

In our material we could not distinguish between reducible and irreducible atlantodental dislocation, since under long enough traction (days until 5–6 weeks [5]) of the diseased craniocervical junction, almost all atlantoaxial dislocations are reducible, as long as the ossification process of the infected focus has not yet been started. This concept has also been proven in cases which were all treated conservatively with gradual traction and secondary Halo vest as stabilisation. Instead of open posterior surgery, those authors use the Halo vest, once the reduction was achieved by traction. Surgical stabilisation is nothing else than the change from external Halo fixation into an internal splinting with instrumentation (Fig. 2).

Bapat et al. [8] demonstrated that transarticular screw fixation alone of C1/C2 is superior to standard occipitocervical fusion and again is sufficient as posterior surgery alone. The transarticular screw fixation may be combined, however, with an occipito-C1/C2-fixation (Fig. 2), if the C1 joint is also affected. In our series we have found that standalone transarticular screw fixation is not possible in most of the patients due to involvement of atlantoaxial joint. From the work of Behari et al. [9] as well as from our own experience can be concluded that patients with Grade I and II disability (only neck pain, independent, with only minor disability and pyramidal track involvement) can be treated with collar immobilisation and antituberculous therapy only.

Patients with Grade III disability (partially dependent on others for assistance, with all activities of daily living) and Grade IV (completely dependent on others for assistance, with activities of daily living with or without respiratory compromise), i.e. those patients who suffer from significant craniomedullary compression may definitively need surgery; either posterior fusion and stabilisation alone, specifically when the atlantoaxial dislocation is reducible under traction. In case of fixed dislocation and persistent hard (bony) ventral compression, a combination of anterior decompression/resection and posterior fusion may be indicated [24]. Low-cost reconstruction plates, including transarticular screws according to Magerl and possibly C3 lateral mass screws and screw fixation to the occiput, is

strongly recommended. The more expensive cervical rod systems may be an alternative.

Authors who favour conservative treatment of all craniovertebral junction tuberculosis [11] admit that in cases with doubtful diagnosis and initial severe or progressive neurodeficit, with or without respiratory distress, in presence of documented mechanical compression and documented dynamic instability following conservative treatment, surgery is a procedure of choice. Others come to the conclusion that surgery is not necessary in craniocervical junction tuberculosis [14], since finally all patients will clinically heal as well as radiologically under conservative treatment.

Conclusion

All patients with tuberculosis of upper cervical spine should be started on anti-tuberculous chemotherapy. Patients without signs of instability and neurological deficit can be treated with collar immobilization for 6 weeks to 3 months. Patients showing signs of instability and having neurological deficit should be placed on skull tongs traction. If they fail to improve neurologically on traction within 4–6 weeks, rigid occipito-cervical fusion should be performed with or without posterior decompression. If they still fail to improve, a rare anterior debridement/decompression should be added to posterior surgery.

Conflict of interest None of the authors has any potential conflict of interest.

References

1. Akhaddar A, Gourinda H, Gazzaz M, Elmadhi T, Elalami Z, Miri A (1999) Craniocervical junction tuberculosis in children. *Rev Rhum Engl Ed* 66(12):739–742
2. Akhaddar A, Chakir N, El Hassani MY, El QA, Jiddane M, Boukhrissi N (2000) Sub-occipital Pott's disease. Diagnostic imaging in 2 cases. *J Neuroradiol* 27(2):144–149
3. Allali F, Benomar A, El YM, Chkili T, Hajjaj-Hassouni N (2000) Atlantoaxial tuberculosis: three cases. *Joint Bone Spine* 67(5):481–484
4. Allali N, El-Quessar A, Melhaoui A, El-Hassani MR, Chakir N, Jiddane M (2004) Sub occipital Pott's disease: report of 8 cases. *J Neuroradiol* 31(1):72–73
5. Arora S, Sabat D, Maini L, Sural S, Kumar V, Gautam VK et al (2011) The results of nonoperative treatment of craniovertebral junction tuberculosis: a review of twenty-six cases. *J Bone Joint Surg Am* 93(6):540–547
6. Arunkumar MJ, Rajshekhar V (2002) Outcome in neurologically impaired patients with craniovertebral junction tuberculosis: results of combined anteroposterior surgery. *J Neurosurg* 97(2 Suppl):166–171
7. Augier A, Zrig H, Roqueplan F, Brauner M, Dumas JL (2008) MR and CT features of craniocervical junction tuberculosis: a report of 5 cases. *J Radiol* 89(5 Pt 1):585–589
8. Bapat MR, Lahiri VJ, Harshavardhan NS, Metkar US, Chaudhary KC (2007) Role of transarticular screw fixation in tuberculous atlanto-axial instability. *Eur Spine* 16(2):187–197
9. Behari S, Nayak SR, Bhargava V, Banerji D, Chhabra DK, Jain VK (2003) Craniocervical tuberculosis: protocol of surgical management. *Neurosurgery* 52(1):72–80
10. Belanger E, Levi AD (2000) Tuberculosis of the axis in a patient with systemic sarcoidosis: technique of posterior open biopsy of the dens: case report. *Neurosurgery* 47(4):969–972
11. Chadha M, Agarwal A, Singh AP (2007) Craniovertebral tuberculosis: a retrospective review of 13 cases managed conservatively. *Spine (Phila Pa 1976)* 32(15):1629–1634
12. Dhammi IK, Singh S, Jain AK (2001) Hemiplegic/monoplegic presentation of cervical spine (C1–C2) tuberculosis. *Eur Spine J* 10(6):540–544
13. Dhaon BK, Jaiswal A, Nigam V, Jain V (2003) Atlantoaxial rotatory fixation secondary to tuberculosis of occiput: a case report. *Spine (Phila Pa 1976)* 28(11):E203–E205
14. Gupta SK, Mohindra S, Sharma BS, Gupta R, Chhabra R, Mukherjee KK et al (2006) Tuberculosis of the craniovertebral junction: is surgery necessary? *Neurosurgery* 58(6):1144–1150
15. Kalra SK, Kumar R, Mahapatra AK (2007) Tubercular atlanto-axial dislocation in children: an institutional experience. *J Neurosurg* 107(2 Suppl):111–118
16. Kapoor SK, Tiwari A, Chaudhry A (2007) An unusual case of craniovertebral junction tuberculosis in an infant. *Spine (Phila Pa 1976)* 32(23):E678–E681
17. Kotil K, Dalbayrak S, Alan S (2004) Craniovertebral junction Pott's disease. *Br J Neurosurg* 18(1):49–55
18. Krishnan A, Patkar D, Patankar T, Shah J, Prasad S, Bunting T et al (2001) Craniovertebral junction tuberculosis: a review of 29 cases. *J Comput Assist Tomogr* 25(2):171–176
19. Lifeso R (1987) Atlanto-axial tuberculosis in adults. *J Bone Joint Surg Br* 69(2):183–187
20. Mohindra S, Gupta SK, Mohindra S, Gupta R (2006) Unusual presentations of craniovertebral junction tuberculosis: a report of 2 cases and literature review. *Surg Neurol* 66(1):94–99
21. Raut AA, Narlawar RS, Nagar A, Ahmed N, Hira P (2003) An unusual case of CV junction tuberculosis presenting with quadriplegia. *Spine (Phila Pa 1976)* 28(15):E309
22. Shukla D, Mongia S, Devi BI, Chandramouli BA, Das BS (2005) Management of craniovertebral junction tuberculosis. *Surg Neurol* 63(2):101–106
23. Sinha S, Singh AK, Gupta V, Singh D, Takayasu M, Yoshida J (2003) Surgical management and outcome of tuberculous atlanto-axial dislocation: a 15-year experience. *Neurosurgery* 52(2):331–338
24. Teegala R, Kumar P, Kale SS, Sharma BS (2008) Craniovertebral junction tuberculosis: a new comprehensive therapeutic strategy. *Neurosurgery* 63(5):946–955