Unusual presentation of more common disease/injury

A rare case of basilar impression

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A teenage girl presented with progressively worsening neck and occipital pain since 8 months ago that was associated with restriction of neck movements, low to moderate grade fever, anorexia and weight loss followed by gradually progressive quadriparesis since 3 months ago. Neurological examination revealed spastic quadriparesis without cranial nerve palsy. MRI of the cervical spine revealed prevertebral and paravertebral abscess from clivus to C2/C3 level, which extended into the anterior epidural space at C1–C3 level, causing atlantoaxial dislocation. There was evidence of basilar invagination. The dislocated dens caused compression over the cervicomedullary junction. On this basis, a diagnosis of craniovertebral junction Pott's disease was made. The patient was given neck immobilisation with cervical collar, and antitubercular treatment was started. She showed significant improvement in her neurological deficit during follow-up.

BACKGROUND

Pott's disease (tuberculosis of the spine) is one of the important manifestations of extrapulmonary tuberculosis. The craniovertebral junction (CVJ) Pott's disease is rare, accounting for less than 1% of all cases. Atlantoaxial dislocations (AAD) are the most common abnormality seen in CVJ Pott's disease. CVJ tuberculosis rarely causes basilar impression. Basilar impression is an acquired type of basilar invagination. It is usually seen in diseases such as rickets, osteogenesis imperfecta, hyperparathyroidism, Hurler's syndrome, Paget's disease and acro-osteolysis (Hajdu-Cheney syndrome). It is believed that the bone and soft tissue involvement by the tuberculous process at the CVJ results in basilar impression. To the best of our knowledge, cases of tuberculosis as a cause of basilar impression have been rarely reported so far. This case highlights the importance of suspicion of tuberculous aetiology, especially in young patients with a chronic history of neck pain and with findings suggestive of high cervical compressive myelopathy.

CASE PRESENTATION

A 17-year-old, previously healthy, girl presented with progressively worsening neck and occipital pain since 8 months ago that was associated with restriction of neck movements. The pain was aggravated on movement of the neck and was relieved by rest. It was associated with low to moderate grade fever along with anorexia and weight loss. She then gradually developed progressive quadriparesis over the last 3 months. This was followed by dysphagia and vomiting for the last 15 days. There was no contact history with other patients with tuberculosis. General examination did not reveal any significant findings except for tenderness over the cervical spine and restricted neck movements in all directions. There was no lymphadenopathy and the neck height ratio was within normal limits. Neurological examination revealed quadriparesis with a bilateral extensor plantar response. There was no evidence of cranial nerve palsy and the sensory examination was normal.

INVESTIGATIONS

Laboratory examination was normal except for raised erythrocyte sedimentation rate. The chest x-ray did not reveal any abnormality. Cervical spine radiographs reveal AAD with basilar invagination (figure 1). MRI of the cervical spine revealed prevertebral and paravertebral abscess from clivus to C2/3 level, which extended to lateral recess and neural foramina at C1/2 level, causing compression over exiting nerve roots, and also extended into the

Figure 1 Radiogram lateral view of cervical spine, showing atlantoaxial dislocation.

Figure 2 MRI of the cervical spine, showing prevertebral abscess.

anterior epidural space at C1–C3 level, causing AAD (figure 2). There was basilar invagination with tip of dens significantly extending above Chamberlain's line (figure 3). The dislocated dens along with the epidural collection and thickened anterior longitudinal ligament caused compression over the cervicomedullary junction with signal intensity alterations within the cord. A vertebral biopsy was planned but could not be done as the relatives did not give consent for the invasive procedure.

DIFFERENTIAL DIAGNOSIS

On this basis, a diagnosis of secondary CVJ anomaly with atlantoaxial dislocation and retropharyngeal abscess probably caused by Pott's disease was kept.

TREATMENT

The patient was given neck immobilisation with cervical collar, and antitubercular treatment (ATT) was started.

Figure 3 MRI of the cervical spine, showing basilar invagination. The horizontal yellow line depicts Chamberlain's line.

ATT consisted of daily doses of Isoniazid (5 mg/kg), Rifampicin (10 mg/kg), Ethambutol (15 mg/kg) and Pyrazinamide (15–30 mg/kg). All the four drugs were given for the first 2 months after which Isoniazid and Rifampicin were advised to be continued for the next 7 months. A neurosurgeon's opinion was sought, who advised conservative management in view of mild compressive myelopathic changes. The patient was discharged and advised to be under regular follow-up.

OUTCOME AND FOLLOW-UP

During regular follow-up for the last 6 months, the patient showed a significant improvement in her neurological deficit. She is still on ATT and is under regular follow-up.

DISCUSSION

Pott's disease (tuberculosis of the spine) is one of the important manifestations of extrapulmonary tuberculosis and constitutes about 6% of all cases of extrapulmonary tuberculosis. Most commonly, Pott's disease involves the thoracic spine. Cervical spine involvement is less common, constituting 10% of all cases of Pott's disease. The CVJ Pott's disease is still rarer and accounts for less than 1% of all cases.^{1 2}

AAD are the most common abnormality seen in the CVJ Pott's disease.³ CVJ tuberculosis can lead to AAD by two mechanisms: (1) osteomyelitis and (2) destruction of the ligaments, mainly the transverse and alar.¹ It is suggested that CVJ tuberculosis results from the spread of infection from the retropharyngeal tissues to the bony elements; primary infection of the atlas or axis is rare. Eventually, the infection causes destruction of the bony and ligamentous structures, leading to cervicomedullary neural compression and occipitocervical and/or atlantoaxial instability.⁴ The occipito-atlantoaxial complex accounts for about 80° rotation and significant flexion. Destruction of this complex results in abnormal translational and rotational movements.⁵ Atlantoaxial instability and abscess can be present together. Abscess can cause lower cranial nerve palsies or spinal nerve root compression, leading to occipital headaches and paraesthesia. Mechanical obstruction from a retropharyngeal abscess can cause dysphagia and airway compromise.¹ The patient may show constitutional symptoms such as fever, anorexia and weight loss as in our case. However, these occur only in approximately 40% of patients with spinal tuberculosis. It is therefore important not to exclude spinal tuberculosis when constitutional symptoms are not present.

There is basilar impression along with the AAD in this case. Basilar impression is an acquired type of basilar invagination. It is seen in diseases such as rickets, osteogenesis imperfect, hyperparathyroidism, Hurler's syndrome, Paget's disease and acro-osteolysis (Hajdu-Cheney syndrome) (table 1). It is believed that the bone and soft tissue involvement by the tuberculous process at the CVJ results in the basilar impression. To the best of our knowledge, cases of tuberculosis as the cause of basilar impression have rarely been reported so far.^{3 6}

CVJ tuberculosis predominantly affected the anterior structures and, hence, the transoral approach is accepted as the method of choice for biopsy of lesions as well as

Table 1 Classification of craniovertebral junction abnormalities

Congenital anomalies/malformations	Developmental/acquired abnormalities
A. Malformations of the occipital bone 1. Manifestations of occipital vertebra a. Clivus segmentation b. Remnants around foramen magnum c. Dens segmentation anomalies 2. Condylar hypoplasia 3. Basilar invagination B. Malformations of the atlas 1. Atlas assimilation 2. Aplasia of arches of atlas 3. Atlantoaxial fusion C. Malformations of the axis 1. Dens dysplasia a. Os odontoidum b. Ossiculum terminale persistens 2. Irregular atlantoaxial segmentation 3. Segmentation failure of C2–C3	A. Abnormalities of foramen magnum 1. Secondary basilar invagination a. Paget's disease b. Osteomalacia c. Rickets d. Rheumatoid arthritis e. Osteogenesis imperfecta f. Hyperparathyroidism g. Acro-osteolysis h. Hurler's syndrome i. Tuberculosis 2. Foraminal stenosis a. Achondroplasia B. Atlantoaxial instability 1. Morquio syndrome 2. Down syndrome 3. Inflammatory a. Rheumatoid arthritis 4. Infections a. Grisel syndrome 5. Traumatic 6. Tumours a. Neurofibromatosis b. Syringomyelia

decompression of the neuroaxis.¹ Positivity of smear and culture for bacilli has been reported to be present in less than 50% of cases. Approximately 75% of cases have shown granulomatous tissue with or without caseation necrosis on histological study. Sensitivity of the PCR test for *Mycobacterium tuberculosis* ranges from 91.4% to 97.6%. Specificity is high (99.5–100%).⁵

The treatment options for the CVJ tuberculosis have been controversial, from an absolutely conservative approach to radical extirpation.^{4–7} This inconsistency is mainly due to the absence of well-proposed guidelines. Also the symptomatic or radiological features are diverse enough to justify several definite treatment modalities. Many authors favoured antituberculous medication with or without external immobilisation in the case of minimal bone destruction and minimal neurological

deficit. Surgical treatment may be required when there are neurological symptoms due to cord compression or a significant degree of instability, AAD or bone destruction. Many authors recommend posterior fusion and fixation and/or anterior decompression, especially when anterior cord compression or irreducible AAD existed.⁶ Owing to the presence of minimal neurological deficits in our case, the patient was subjected only to conservative treatment and showed improvement with it.

Learning points

- ▶ This is one of the rare cases with tuberculosis as the cause of basilar impression.
- ▶ This case highlights the importance of suspicion of tuberculous aetiology, especially in young patients with a chronic history of neck pain and with findings suggestive of high cervical myelopathy.
- ▶ Conservative line of management with antitubercular treatment still has a role in mild neurological deficits in cases of craniovertebral junction Pott's disease.

Competing interests None.

Patient consent Obtained.

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