

Craniocervical Junction Tuberculosis: A Rare but Fatal Disease

Abdur Rahman^{1*}, Abhinav Srivastava², Vishal K Gojiya³, Kiran Verma⁴

1. Jawaharlal Nehru Medical College, Aligarh Muslim University, India.

2. Rohilkhand Medical College & Hospital, India

3. Assistant Professor MP Shah Medical College and Hospital Jamnagar Gujrat, India.

4. Assistant Professor Sarojini Naidu Medical College and Hospital Agra, India

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Corresponding Authors:

Dr. Abdur Rahman

Email:

abdurrahman6034@gmail.com

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Abstract

Background: Pott's spine describes tuberculous infection of the spine and the term "Pott's paraplegia" describes paraplegia resulting from the tuberculosis of the spine. Tuberculosis is a disease of poverty that affects mostly young adults in their most productive years. The progression of spinal tuberculosis is slow and insidious. Formation of a cold abscess around a vertebra is another characteristic feature of spinal tuberculosis. Retropharyngeal abscess can produce considerable pressure effects such as dysphagia, respiratory distress, or hoarseness of voice. Therefore, early diagnosis and treatment in life-saving in these patients.

Case Presentation: A 16 year old female patient presented with neck pain, difficulty in swallowing, slight restriction of neck movements and history of fever and night sweats. A detailed history was taken and complete examination was done. A swelling in posterior pharyngeal wall was seen and drained and the pus collected was sent for culture and sensitivity and for confirmation of the Tuberculosis. Radiological investigations were done which revealed retropharyngeal abscess and the extent of involvement of the craniovertebral junction. After confirmation of tuberculosis, antituberculosis treatment was started and patient was followed up and showed remarkable improvement.

Conclusion: Craniocervical junction tuberculosis is a rare disease with dangerous outcome if not diagnosed and treated early. antituberculosis therapy is the mainstay of conservative treatment for early diagnosed cases and has good prognosis.

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Introduction

Spinal tuberculosis is one of the oldest diseases known to mankind also known as Pott's spine, while the term Pott's paraplegia is the term used when paraplegia occurs due to spinal tuberculosis(1,2). The term Pott's disease classically includes disc space and nearby vertebral bodies with progressive kyphosis. It occurs slowly and the duration varies from months to years, ranging from 4 to 11 months' average.

Tuberculosis remains one of the most common chronic diseases in the developing countries, but only 10% of the patients of extrapulmonary tuberculosis present with skeletal involvement,

with spine being the most common (~50%) area affected, followed by hip and knee. Craniovertebral Junction (CVJ) tuberculosis is the rarest form of spinal tuberculosis presentation, with an incidence of 0.3% to 1%(3).

Classically, tuberculosis presents with weight loss, malaise, night sweats, generalised body ache, fatigue and night fevers. Spine tuberculosis patients usually present with severe pain, movement restriction and some other neurological symptoms (4, 5). When cervical spine involves, patient may present with the symptoms of root or cord compression, like weakness, pain, numbness of both extremities

may progress to tetraplegia too.

Cold abscess mostly involves the spine which is common and variable in size and occur anywhere within the length of vertebral column. In retropharyngeal abscess, patient presents with pressure symptoms like dysphagia, respiratory distress or hoarseness of voice. This occurs when cervical spinal tuberculosis resulting in pus accumulation behind the prevertebral fascia.

On plain x-ray, tubercular cold abscess can be seen as soft tissue shadows nearby the affected spine. In x-ray for retropharyngeal abscess, increase in prevertebral space in cervical region is notable(6). MRI is the most useful diagnostic tool to diagnose a case of spinal tubercular especially in cervicodorsal junction and craniocervical junction. MRI shows the disc destruction, vertebral bodies involvement, disc collapse, cold abscess and other spinal deformities (7, 8).

On cytology, tuberculosis can be confirmed by presence of epithelioid cell granulomas with necrotic granules in background and lymphocytic infiltration. In half of the patient we may see Langerhans' giant cells and scattered multinucleated cells. When bacteriological reports are negative, diagnosis must be made on clinical manifestations for tuberculosis (9).

Case Presentation

A 16-year-old female came to OPD in ENT Department with a 10-month history of pain in the posterior neck radiating to the suboccipital area and shoulders. The pain increased on standing and was relieved by lying supine. There was history of fever and night sweats, but no family history of tuberculosis (TB).

On examination, she looked sick and afebrile. Her pain could be reproduced by pressing on or rotating her head (Hilton test)(10). Patient was pale and had weight loss. Haemoglobin was 8.7gm% and differential count were normal; MCH (20.5), MCHC (29.6), MCV (69.3) and PCV (29.4) values were below normal range. ESR was 26 mm (normal range is 0-9mm in 1st hour). RBC count was 4.24 million (normal range is 4.5-6.5 million/cu-mm). CBNAAT was positive detected in pus drained from the retropharyngeal abscess and was sensitive to Rifampicin. Pus

aspirated from the retropharyngeal abscess was sent for culture and sensitivity. Acinetobacter was the bacteria isolated which is a common commensal of the moist areas of the human body. It was sensitive to ampicillin, cefuroxime, cefixime, gentamicin, tobramycin, levofloxacin, imipenem, aztreonam, cefepime and resistant to amoxicillin/clavulanic acid.

CECT neck (Figure 1) showed right atlanto-occipital joint that was irregular with associated destruction along with irregularity of the occipital condyle and right lateral mass of the atlas (C1 vertebra). A well-defined peripherally enhancing lesion measuring 5.5*2.2*4.5 cm with few foci of calcification seen in the retropharyngeal space predominantly on the right side extending to the perivertebral space in the retro-styloid region pushing the right styloid process anteriorly (Figure 2).

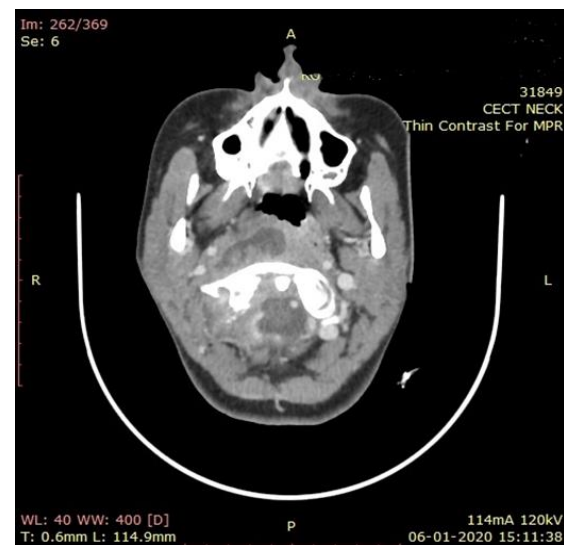


Figure. 1 CT Scan Axial views showing irregularity of right atlanto-occipital joint and destruction of occipital condyle.

Multiple subcentimetric enhancing level 1b, 2, and 3 cervical lymph nodes are seen bilaterally. MRI of this lesion confirmed the mentioned CT findings. Sections of the thorax showed patchy areas of consolidation with surrounding centrilobular nodules showing tree-in-bud appearance in the apical and posterior segments of the right upper lobe. A small patchy opacity is also seen in the apico-posterior segment of the left upper lobe.

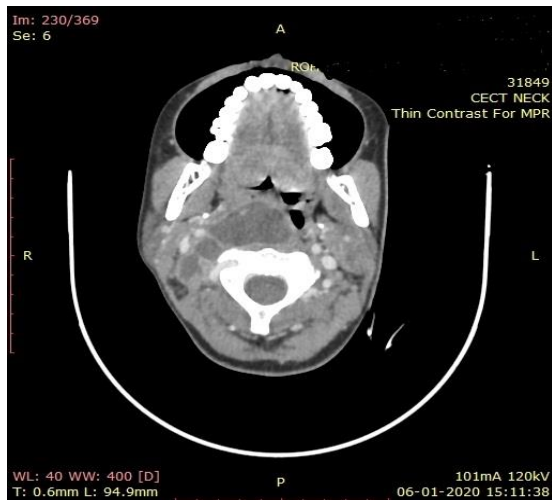


Figure. 2 CT Scan Axial view showing defined peripherally enhancing lesion in the retropharyngeal region.

Chest X ray showed right side apical consolidation (Figure 3).



Figure .3 X ray Neck lateral view showing retropharyngeal abscess

Peroral aspiration of the retropharyngeal swelling was performed. Thick yellow pus was aspirated, necrotic tissue was removed, and multiple tissue biopsies were obtained. Aspirated pus was positive for Tuberculosis in CBNAAT. The biopsy tissue revealed epithelioid granulomas with Langerhans' giant cells.

The patient was given antitubercular treatment

under category 1 for directly observed treatment strategy for extrapulmonary tuberculosis patients. The treatment was started as per the recommendations of Revised National Tuberculosis Control Program (RNTCP) which included Rifampicin (350mg), Isoniazid (225 mg), Pyrazinamide (1200 mg) and Ethambutol (825 mg) daily for 56 days in the Intensive phase. The Continuation phase includes Rifampicin (450 mg), Isoniazid (225 mg) and Ethambutol (825 mg) for 112 days.

The follow ups reported improvement in fever, neck movements, difficulty in swallowing and had then started gaining weight.

Discussion

Spinal tuberculosis commonly involves thoracolumbar region, whereas craniocervical junction tuberculosis is the least common presentation and accounts approximately 1% of all cases of spinal tuberculosis (11-13).

Craniocervical junction tuberculosis mostly occurs as secondary infection of bone and may present as retropharyngeal abscess. With progression, it involves nearby ligaments, atlas and odontoid erosion may occur resulting in unstable articulation from occiput to atlas. Primary infection of atlas and axis bones is rare. Acute lesion of craniocervical junction may be fatal as it is a very important region for spinal canal. Movement restriction and severe pain are the most common symptoms of craniocervical tuberculosis. Osteomyelitis and destruction of transverse and alar ligaments resulting in atlantoaxial subluxation (14). So early fixation and appropriate spinal implementation is necessary.

Pandya revealed involvement of bone and ligaments by the disease process as possible causes for dislocation (15). Lifeso suggested that CVJ tuberculosis results from infection that spreads from the retropharyngeal tissues to the osseous elements; primary infection of the atlas or axis was rarely the cause of disease (16). Patient may present with abscess formation and atlantoaxial instability with symptoms of lower cranial nerve palsies or root compression. Respiratory distress and dysphagia may occur due

to pressure effect of retropharyngeal abscess and may extend up the mediastinum too. Head stabilisation with neck extension may require prolonged bed rest for better results. There are several methods and procedures suggested for craniocervical tuberculosis treatment. As the retropharyngeal abscess progresses it may cause dysphagia and airway obstruction due to mechanical obstruction and may extend up to the posterior triangle or the mediastinum. Following debridement, several methods of achieving stabilization have been proposed. Tuli proposed prolonged bed rest and skull traction with the neck held in extension(17).

The ATT alone provides an excellent neurological outcome in most of the patients with CVJ tuberculosis without atlantoaxial instability. This approach helps in immediate neurological improvement, immediate spinal stability and excellent long-term outcome.

Conclusion

Craniocervical junction tuberculosis is a rare disease and requires a detailed radiological and histological proof to arrive at the correct diagnosis. Antitubercular therapy along with prolonged immobilization is the mainstay of conservative treatment option and can cure the disease. The long term prognosis of this condition is good in patients with an early diagnosis and treatment.

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Conflicts of Interest

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Authors ORCID's

Vishal K Gojiya

<https://orcid.org/0000-0003-1518-508X>

Abbreviations

OPD- Out Patient Department

MRI: Magnet Resonance Imaging

MCH: Mean Corpuscular Hemoglobin

MCHC: Mean Corpuscular Hemoglobin Concentration

MCV: Mean Corpuscular Volume

PCV: Packed Cell Volume

ESR: Erythrocyte Sedimentation Rate

RBC: Red Blood Cell

CBNAAT: Cartridge based Nucleic Acid Amplification Test

CECT: Contrast Enhanced Computed Tomography

ATT: Antitubercular Treatment

CVJ: Craniovertebral Junction

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