Infections of the Craniovertebral Junction

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ABSTRACT

Infections of the craniovertebral junction (CVJ) are rare, as compared to other sites of spinal column, in both tuberculous and non-tuberculous pathologies. Non-tuberculous abscess is unique and the incidence of CVJ tuberculosis ranges from 0.3 to 3% (7) of spinal TB infections. The scarcity of the infection is postulated to be secondary to the very good vascularity of the area and decreased epidural space in the region. The incidence is increasing secondary to the number of immunocompromised hosts, as seen with AIDS, transplant, drug abuse, CKD and DM, and improvements in the diagnostic tests that are available. However tuberculous osteomyelitis is more frequently seen in endemic areas.

KEY WORDS: Abscess, cranio-vertebral junction, CVJ, CVJ TB, infection, upper cervical spine

INTRODUCTION

Infections of the craniovertebral junction are rare, as compared to other sites of the spinal column, in both tuberculous and non-tuberculous pathologies. Non-tuberculous abscess is unique and the incidence of CVJ tuberculosis ranges from 0.3 to 3% (7) of spinal TB infections. The scarcity of the infection is postulated to be secondary to the very good vascularity of the area and decreased epidural space in the region. The incidence is increasing secondary to the number of immunocompromised hosts, as seen with AIDS, transplant, drug abuse, CKD and DM, and improvements in the diagnostic tests that are available. However tuberculous osteomyelitis is more frequently seen in endemic areas.

PRESENTATION

Patients with CVJ infection may present with a wide variety of mostly non specific symptoms ranging from simple neck pain to severe instability and quadriplegia, but the most common complaint is neck pain(1) and h/o fever and movement restriction; neurological signs usually appear later. Patients often have history of fever, recent or chronic ENT infection (Figure 1) or operation, cervical spine procedures and surgery, immunocompromised state, and trauma (2, 15). The authors have seen several cases after tonsillectomy. The symptoms include neck pain, stiffness, fever, neurological weakness, jaundice, nerve palsies, myelopathy, hoarseness of voice, dysphagia, headache, neck and back pain, and in case of tuberculous abscess night sweats and weight loss. Sometimes the diagnosis is delayed and confused in patients with history of trivial trauma so a high index of suspicion is required in endemic areas, with a positive history of risk factors, and in patients with neck pain. The presentation will depend on the progress of the disease, with progressive destruction of the facets and odontoid leading to instability and pain, and canal compromise and compression by abscess leading to the neurological symptoms (13).

WORK UP

Routine blood workup often shows raised white blood cells (WBC), C-reactive protein (CRP) in non TB etiology and raised erythrocyte sedimentation rate (ESR) and CRP in most infectious cases, although the range varies but 45-50 should be considered suspicious (6). These tests are non specific but help in the overall diagnosis. A raised ESR with neck pain should raise the suspicion of craniovertebral junction (CVJ) TB and appropriate investigations should be advised earlier.

Polymerase chain reaction (PCR) also has wide range of positivity, with average of 70 -75 % (16), and the reliability of image-guided biopsies also varies, so the need of these tests should be weighed in terms of cost benefits, local facilities and expertise, and effect on management plan

X-ray is a simple investigation which may show bony erosion and destruction, increased epidural space, and movement and mal alignment of atlas and axis, basilar invagination, while flexion extension views highlight inherent insatiability.

MRI with contrast is the investigation of choice among imaging modalities (Figure 2) and has predictive value and sensitivity of up to 95% and specificity over 90% (3). Clinical symptoms become evident when there is up to 60% canal compromise (17). Krishnan et al (11) have graded the spinal cord abnormalities on MRI in TB into three grades as follows

Grade 1- no displacement of theca and no evidence of compression

Grade 2- displacement of theca but no evidence of compression

Grade 3- compression of the cord with or without degenerative changes such as syrinx or myelomalacia.

CT scan is helpful in delineating the bony anatomy, and helps in surgical planning, Lifeso et al. had divided CVJ TB into 3 grades (9). CT myelography is also an option where MRI is contraindicated.

Tissue Diagnosis: In non-tuberculous infections, causative organisms can be identified in 70 % of the cases on CT-guided aspirations, while the yield is very low in tuberculous cases, and the presence of caseation should be considered as hallmark of TB (4).

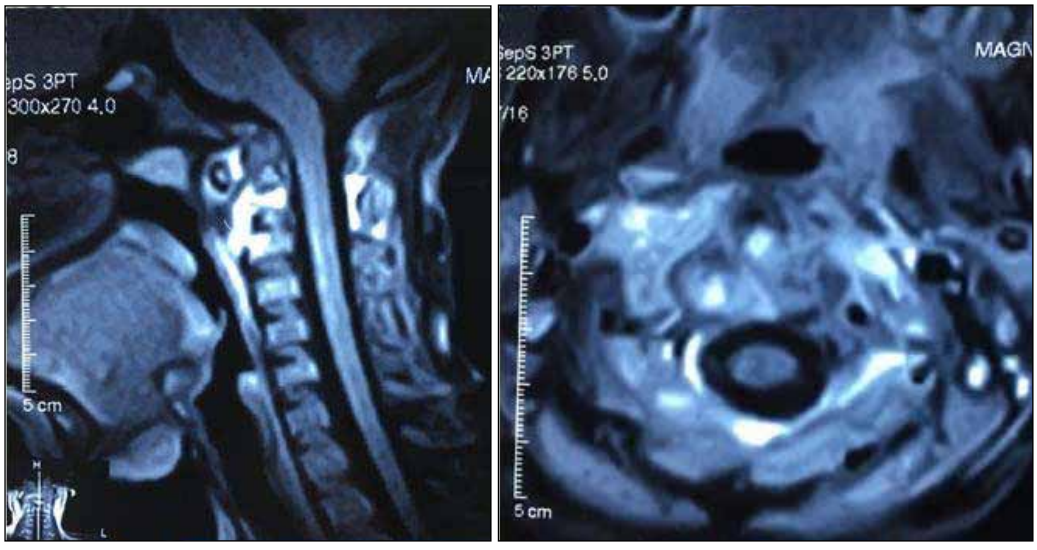

Figure 1: *T1 sag +T1 contrast image of 13 yr old with torticollis and neck pain had sore throat 2 weeks prior Increase WCC and ESR Settled with 6 weeks IV therapy.*

Figure 2: *T1, T2, and T1 axial Contrast of and elderly with TB abscess. Pus was aspirated anteriorly and patient was given 12 months of ATT with success.*

MANAGEMENT

The management of CVJ abscess includes both, operative and non operative treatments. The decision mainly depends on the neurologic symptoms, their progression and the instability present. The infection should be identified in both TB, and non TB infections. The blood cultures are negative in 40 % of the cases. The patients who have no instability and have no progressive neurologic symptoms can be managed with immobilization, and specific antibiotics. In cases of TB where cultures are negative empirical treatment can be started in endemic areas. The response to quadruple line therapy is seen in 3 to 6 weeks time with imaging and lab studies. Some risk factors that are a contraindication for conservative treatment are age more than 60, DM, and infection with MRSA (10). These factors are associated with over 90% chance of failure of conservative treatment (10).

The clinical improvement and radiological improvement should be followed and patients who are not improving, or have a neurological deficit, cord compression or instability with gross bone destruction should be considered for surgery. Options for surgery include, simple aspiration, trans oral decompression, and posterior decompression with or without fixation. A halo vest may be required during conservative treatment, or the early post op period.

Management of Tb infection is slightly different in that it requires long term chemo therapy ranging from 12-24 months (12,13,17). Commonly a regime of 4 drugs for 18-24 moths is used. If this is a good response, these can be reduced to 2 or 3 drugs in the maintenance phase (after 2 months). In unresponsive and resistant cases, second line drugs like streptomycin or levofloxacin may be added, and an increasing use of these drugs may be required with increasing frequency of resistant TB.

Failure of medical therapy is considered in cases of worsening inflammatory markers, worsening of neurological state, increasing pain or instability on follow up imaging. Instability can initially be managed with traction and halo vest, and reducibility of the unstable components can be assessed before formal fixations can be planned. Some studies have shown better motor outcomes of early surgical intervention in patients with neurologic deficit (1,7,10,11, 14,17).

Several grading systems were proposed depending on clinical parameters for e.g. De Lorenzo, grading by Behari et al,, and radiological grading and others using both criteria (1,5,8). All the grading systems still cannot fully guide towards conservative vs. surgical management.

CONCLUSION

Infection of CVJ is a rare condition, and one should have a high index of suspicion when the patient presents with neck pain, movement restriction, with or with fever and raised inflammatory markers, and in areas of endemic TB. Urgent radiological investigations should be performed and instability appropriately treated, with traction halo vest and fixation.

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