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## Multifocal Skeletal Tuberculosis Presenting as Osteitis Skull and Atlantoaxial Dislocation

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Although skeletal tuberculosis is still frequent in many parts of the world, multifocal bone involvement in tuberculosis is rare. It is reported that 4.6% to 10% of skeletal tuberculosis is multifocal(1,2). Osteitis of skull with associated

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multiple cerebral tuberculoma with atlantoaxial dislocation of tubercular etiology manifesting in a child has not been reported in the English literature to the best of our knowledge. The present report describes a 14-year-old female who presented with the above findings with histopathological proven skeletal tuberculosis and reviews the literature on multifocal skeletal tuberculosis.

### Case Report

A 14-year-old female presented with a swelling in the occipital region associated with localized headache for the last 5 months. The swelling was initially pea size and progressively increased and finally burst discharging pus leaving a sinus. Two months prior to admission the swelling was excised by a local doctor but it recurred and increased in size. There was no history of preceding trauma. Ever since the appearance of swelling, patient had malaise, low grade fever with evening rise and progressive weight loss. One month after the swell-

ing was noticed, patient developed pain in the neck and movements of the neck were markedly restricted in all directions. There was no history of swellings in the neck, dysphagia, dysphonia and no associated cough or hemoptysis.

One month prior to admission she had a generalized tonic clonic seizure lasting for about 5 minutes. Ever since the seizure there was history of irritability, recurrent vomiting and occipital headache which was partially relieved with analgesics. There was no alteration of sensorium, history suggestive of any cranial nerve palsy, meningeal signs or focal neurological deficit. Even on repeated close questioning there was no history of contact with a case of tuberculosis.

Examination revealed a thinly built febrile child with a swelling in the occipital region in the midline, fluctuant, tender with an overlying sinus which was discharging pus. The skin over the swelling appeared attached but not inflamed and there was no cervical lymphadenopathy. On careful palpation of the swelling there was an underlying bony defect with well defined edges.

There was marked restriction of neck flexion, extension and rotation. Palpation of upper cervical spine revealed tenderness over the atlas and axis. On careful clinical examination no other discharging sinuses, lymphadenopathy or retropharyngeal abscess could be demonstrated. There was no BCG scar. Chest, cardiovascular system and abdominal examination were within normal limits. Central nervous system examination did not show any cranial nerve involvement or areas of

muscle wasting. Tone was slightly increased in all four limbs. There was generalized hyper reflexia. Babinski's sign was positive. Cerebellar signs could not be elicited.

Investigations showed a hemoglobin of 11 g/dl, TLC of 10,000/cu mm, DLC  $P_{80}L_{20}$  and ESR 50 mm in the first hour. Mantoux test was strongly positive. X-ray cervical spine (*Fig. 1*) showed atlantoaxial dislocation with no evidence of bony erosion and no increase of prevertebral shadow. Chest X-ray was within normal limits. Contrast enhanced computed tomographic scan (*Fig. 2*) showed multiple tuberculomas in left occipital lobe with extensive surrounding edema. Ventricular system and anterior fossa were within normal limits. There was erosion of the occipital bone in the midline with preserved inner table. The bony defect had well defined margins, with no periosteal reaction and there was overlying soft tissue swelling clearly suggesting osteitis of skull with scalp abscess. The patient was given antitubercular chemotherapy, anti-convulsants, glycerol to decrease brain edema and underwent surgery. At operation besides tubercular granulation tissue, there was atlanto axial dislocation due to destruction of the transverse and check ligaments. Posterior fixation of C1 and C2 with occipital bone using stainless steel wire, rib grafting debridement of tubercular sinus and curetting of granulation tissue was done. Post operative course was uneventful. X-ray cervical spine after surgery showed normal alignment with the stainless steel wire in place. Repeat CT scan after few months of ATT did not demonstrate any tuberculoma and bone healing was complete. Histopathology of excised tissue



Fig. 1. X-ray cervical spine showing atlanto axial dislocation.

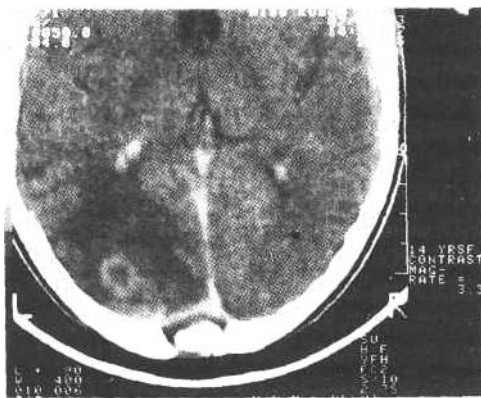


Fig. 2. CT scan showing multiple tuberculomas in left occipital lobe

was consistent with tubercular granulation tissue. The patient received anti tubercular therapy for 1 year and showed a complete neurological recovery. She was well with no neurological deficit when last seen 6 years after discharge from the hospital.

### Discussion

Despite improvement in socio economic status and improved hygiene, tuberculosis remains an important cause of morbidity and mortality in the developing world. Extrapulmonary tuberculosis and skeletal disease in particular may be insidious and often poses a diagnostic challenge. Extrapulmonary tuberculosis accounts for 15-25% of reported cases of tuberculosis in adults and children(3,4). Skeletal lesion account for approximately 4-10% of extrapulmonary tuberculosis(3,4). Medical literature on multifocal skeletal disease especially among Indian population is both brief and sparse. *Table I* summarizes the various series of skeletal tuberculosis including those with multifocal bone involvement.

Kumar and Saxena reported 48 patients between the ages of 4 and 42 years with multifocal skeletal involvement and suggested that poor host immunity

may be a predisposing factor(9). The spine is involved in 50% of the cases, the hip in 15%, knee in-15%, wrist, ankle, elbow, shoulder and other bones in 20%(10). Lesions may occur throughout the spine but often seen in thoracic or lumbar regions with two or more vertebrae typically affected. Goldblatt and Cremin described 499 cases of osteo articular tuberculosis including affliction of upper cervical spine in 11 cases of which three involved the atlantoaxial joint(8). Tuberculosis of the cranio vertebral junction forms 1% of all cases of spinal tuberculosis and primarily involves the atlas and axis(11). Tubercular infection is an infrequent but notable cause of cervico medullary compression ' in developing countries(12).

Tuli described 25 patients with cranio vertebral junction tuberculosis

**TABLE I**—Summary of Reported Series of Skeletal Tuberculosis

Year	Authors	Case of bone tuberculosis	Multifocal skeletal involvement
1942	Meng and Wu(5)	40	27
1949-1957	Sinha(6)	No case out of 12000 cases of tuberculosis.	
1965-1967	Tuli(7)	-	86
1978	Goldblatt and Cremin(8)	499	41
1976-1985	Kumar and Saxena(9)	-	48

who were successfully treated with local rest, chemotherapy and surgery for drainage of cold abscesses(11). Details of various modalities of management were described by Lai *et al.*(12). A diagnosis of atlanto axial dislocation of tuberculous etiology is suspected when a patient has all ,of the following(12): (i) Severe neck pain and restriction of neck movements; (ii) Atlantoaxial dislocation with erosion of the bony elements; and (iii) Raised ESR.

These patients usually present with marked neck stiffness, a history suggestive of tuberculosis and finally progress to quadriparesis due to cord compression. The reported patient had marked tenderness over the atlas and severe pain prohibiting any rotatory neck movements. A retropharyngeal or cervical abscess was not demonstrated. As radiological studies showed no destruction or fracture of the odontoid process and the atlas ring too was unbroken, dislocation could have occurred as a consequence of ligamentous involvement.' The reported studies describing autopsy findings demonstrated destruction of the entire atlantoaxial ligament complex including the vital check and transverse ligaments as seen in this case(13). The ligaments are extensively infiltrated by the disease process and give way leading to dislocation and cord compression.

Tuberculosis of the bone in general usually beings in the cancellous portion of the bones involved. It is because there is little cancellous bone in the flat bones of the vault of the skull that these bones are seldom affected by tuberculosis(14). The condition occurs most frequently in early childhood and one half of the cas-

es occur before the tenth year. Strauss in his analysis of the 220 previously reported cases gave a very comprehensive description of this condition(14). Ten years later Meng and Wu reported some 40 cases seen between the years 1926 and 1940(5). Calvarial tuberculosis has also been reported from India(15-23). Nagi and Sodhi in their analysis of 14 cases of tuberculosis of flat bones found skull involvement in only one case(21). Out of 156 cases of chronic osteomyelitis of skull Mohanty found 22 cases of tubercular origin(20). Out of these, only one patient had associated cerebral tuberculoma. Occipital bone involvement was not found in any of the above studies and has been reported in only 2 patients(22). A primary tuberculosis of the skull is rare as the condition is almost always secondary to tuberculosis elsewhere like lungs, orbit, paranasal sinus, nasal mucosa, lymphatic system. It is extremely rare for the lungs to be free from demonstrable disease as seen in this patient.

Two types of the disease are generally described; the circumscribed (perforating) type and the diffuse progressive type (Koenig). The circumscribed perforating type is the most frequent form(14). This type is characterized by a round usually punched out looking defect through the entire thickness of the skull. The initial focus begins as a localized area of granuloma formation within the diploe which slowly develops in size and presents the typical rarefying osteitis seen in the other flat bones. When a perforation has penetrated the outer table, the periosteum is lifted up and ruptures and pus escapes into the muscles and finally a typically discharging fistula develops as seen in this

patient. Headache if present is usually localized to the site of bone involvement. In a few cases there are signs of cortical irritation with epileptic seizures and rarely signs of cerebral compression. Other symptoms include hemiparesis, focal epilepsy, papilledema, superior sagittal sinus thrombosis and development of cerebral abscess(20). Associated meningitis and cerebral tuberculoma is extremely rare. Of the 223 cases reviewed by Strauss, tubercular meningitis was recorded in ten and cerebral tuberculosis in five(14).

Roentgenograms usually clinch the diagnosis as they almost always show one or more circumscribed punched out defects in the bone(14). The bone about the defect appears entirely normal even upto the very margin of the perforation. This picture is typical of circumscribed tuberculosis of the flat bones of the skull. It may also present as expanding destructive lytic lesions with ill defined irregular margins having at first a sclerotic border and later a osteoporotic edge. A button sequestrum or bone sand may be seen within lesion.

Diagnosis is by a high index of clinical suspicion, a positive Mantoux test, radiological features, aspiration of swelling for bacteriological examination and biopsy for histopathological examination. Treatment is mainly conservative. Surgery is indicated in case of extensive destruction, presence of secondary infection and intracranial involvement. The prognosis is remarkably benign and complete healing usually occurs as seen in this patient.

Thus tuberculosis should be suspected in cases such in which an apparently

normal host presents with osteomyelitis in an usually site. Careful diagnosis and aggressive management of multifocal bone tuberculosis usually leads to a complete clinical as observed in the present case.

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