

Pott's disease

TB extending beyond the thorax is classified as extrapulmonary TB. Reported to occur in 8% of all cases of TB in 1964, the proportion has steadily risen to approximately 16% in 1991 and to 20% in 2001. It might have further risen to 25% or more by end 2010. The etiology underlying this increase in extrapulmonary disease remains unclear.

Common sites of extrapulmonary TB include the musculoskeletal system, genitourinary tract, and lymphatic system. Pott's disease accounts for approximately 50% of extrapulmonary TB that involves the bones and joints, but has a reported incidence of less than 1% to 5% of all TB cases.

Pott's disease is a presentation of extrapulmonary tuberculosis that affects the spine, a kind of tuberculous arthritis of the intervertebral joints. It is named after Percivall Pott (1714–1788), a London surgeon who trained at St Bartholomew's Hospital, London.

The lower thoracic and upper lumbar vertebrae are the areas of the spine most often affected. Scientifically, it is called tuberculous spondylitis and it is most commonly localized in the thoracic portion of the spine. Pott's disease results from hematogenous spread of tuberculosis from other sites, often pulmonary. The infection then spreads from two adjacent vertebrae into the adjoining intervertebral disc space. If only one vertebra is affected, the disc is normal, but if two is involved the disc, which is avascular, cannot receive nutrients and collapses. The disc tissue dies and is broken down by caseation, leading to vertebral narrowing and eventually to vertebral collapse and spinal damage. A dry soft tissue mass often forms and super infection is rare.

Etiology of Tuberculosis of Spine

The onset is gradual.

- Causative organism: *Mycobacterium tuberculosis*.
- Spread: Hematogenous. (by blood)
- Commonly associated with: Debilitating diseases, AIDS, Drug addiction, Alcoholism.

Signs and symptoms

- back pain which is localised
- Restricted spinal movements
- fever
- night sweating
- anorexia (loss of desire to eat)
- weight loss
- Spinal mass sometimes associated with numbness, paraesthesia, or muscle weakness of the legs.
- There may be kyphosis. (Spinal curvature)
- Muscle wasting.
- A paravertebral swelling may be seen.
- They tend to assume a protective upright, stiff position.
- If there is neural involvement there will be neurological signs.
- A psoas abscess (may present as a lump in the groin and resemble a hernia).

Differential diagnosis

- Pyogenic osteitis of the spine. (Pus)
- Spinal tumours.

Diagnosis

- blood tests - elevated erythrocyte sedimentation rate
- tuberculin skin test
- radiographs of the spine
- bone scan
- CT of the spine
- bone biopsy

- MRI

Diagnosis of Pott's disease can be challenging, since in the early phases the clinical presentation and radiographic imaging are often nonspecific. Malignancy as well as pyogenic or fungal vertebral infections can have similar presentations and imaging characteristics. The chances of misdiagnosis can be very high.

Diagnosis can therefore be delayed for months to years, increasing the potential for neurologic complications. History of a positive tuberculin test, active TB, HIV, immuno-suppression, or contact with an individual with TB should raise the index of suspicion for Pott's disease, but these 'flags' are often not present.

Clinically, the most common symptom is back pain. Pertuiset et al. evaluated 103 patients with Pott's disease and found 97% presented with back pain, which was consistent with previously reported data. Found to a lesser degree was fever in 31% and weight loss in 48%. Neurologic deficit was noted in 50% of patients, but rates ranging from 22% to 76% have been observed in other studies. Specific deficits resulting from compression of the spinal cord, conus medullaris, cauda equina, or nerve roots consist of bowel/bladder incontinence, sensory disturbance, and weakness including paraplegia.

Although nonspecific, laboratory tests of value include an elevated erythrocyte sedimentation rate (ESR), abnormal liver function tests, and a rise in the white blood cell count. Of these tests, the ESR appears most helpful, with an increase ($>20\text{mm/h}$) reported to occur in 88% to 100% of afflicted patients.

Similar to the clinical presentation and laboratory testing, imaging can be helpful but is typically not pathognomonic. Chest x-rays are often normal in patients with extrapulmonary TB, including those with Pott's disease. Although changes on spine x-rays may be delayed up to 8 weeks, there is, classically, a collapse of 2 adjacent vertebrae anteriorly with destruction of the intervening disc. Malignancy, in particular, can appear similarly but often will have preservation of the

intervening disc. CT imaging offers the advantage of earlier and more detailed visualization of bony involvement and detection of para-spinal soft tissue masses. Characteristics suggestive of TB on CT imaging include the combination of a multi-locular, calcified para-spinal abscess with an irregular enhancing rim in conjunction with vertebral body bony fragmentation.

Due to its multi-planar capability and high resolution of soft tissue and bone, MRI is now the imaging modality of choice. MRI can show additional sites of infection as well as detect early changes involving the endplate. In particular, T2 hyper-intensity, T1 hypo-intensity, and enhancement with contrast, while not completely specific, are often consistent with TB. The degree of spinal cord or nerve root compression as well as intra-osseous, intra-spinal, and para-spinal abscesses with characteristic patterns of rim enhancement can also be visualized. Based on suggestive imaging characteristics, CT-guided or open biopsy can then provide definitive diagnosis.

Diagnosing TBM. TBM occurs less frequently than Pott's disease, comprising just 15% of extrapulmonary TB cases and only 0.7% of all TB cases. In adults, TBM can develop in isolation or with another form of the disease, particularly miliary or pulmonary. Typically, TBM develops in 2 stages. There is an initial hematogenous dissemination with the primary infection, or alternatively, dissemination during the course of a chronic infection resulting in caseous foci of bacilli deposited within the brain. Subsequent rupture of a focus into the subarachnoid space results in meningitis. More rarely, the meninges can be infected from a site of tuberculous spondylitis, presumably from direct extension of the infection through the adjacent dura. Either route, hematogenous dissemination from chronic infection or direct extension, can contribute to the development of TBM in the patient.

The clinical manifestations of TBM are varied and result from multiple factors including the development of a thick basilar exudate, vasculitis and vascular occlusion, allergic reaction to tuberculo-protein,

cerebral edema, and formation of tuberculomas. Symptoms typically are gradual in onset with malaise, low-grade fever, intermittent headache, and change in personality. Subsequently, a more pronounced meningitic phase develops within 2 to 3 weeks involving persistent headache, meningismus, vomiting, confusion, cranial nerve palsies, or long-tract signs. Symptoms can progress to stupor or coma followed by seizures, hemiparesis, and hemiplegia. Death typically occurs within 5 to 8 weeks if left untreated.

Cranial nerve palsies from basilar exudates occur in 20% to 30% of patients and most commonly involve CN VI and less frequently CN III followed by CN IV and VII. Other cranial nerves are rarely affected, but involvement of CN II can cause optic atrophy and blindness. Similar to Pott's disease, the tuberculous etiology of the meningitis, when it occurs in isolation, can be diagnostically challenging. A high index of suspicion in the patient with a history of TB or evidence of extrapulmonary TB is essential. Laboratory testing is typically not helpful, although an elevated ESR and positive tuberculin test may be present. Lumbar puncture for cerebrospinal fluid analysis should be performed but can also be non-diagnostic.

Prognosis is highly dependent on the timing of treatment. Early initiation of treatment, prior to focal neurologic deficit or mental status changes, will lead to the best outcomes. Even short delays involving several days to a week can result in significantly increased risk for permanent neurologic deficit and mortality. Empiric therapy is therefore recommended based on clinical suspicion and suggestive laboratory data.

Late complications

- Vertebral collapse resulting in kyphosis (exaggerated backward curvature of the thoracic region of the spinal column)
- Spinal cord compression
- sinus formation
- paraplegia (so called Pott's paraplegia)

Prevention

Controlling the spread of tuberculosis infection can prevent tuberculous spondylitis and arthritis. Patients who have a positive PPD test (but not active tuberculosis) may decrease their risk by properly taking medicines to prevent tuberculosis. To effectively treat tuberculosis, it is crucial that patients take their medications exactly as prescribed.

Therapy

- non-operative - antituberculous drugs
- Analgesics
- immobilization of the spine region by rod (Hull)
- Surgery may be necessary, especially to drain spinal abscesses or to stabilize the spine
- Richards intramedullary hip screw - facilitating for bone healing
- Kuntcher Nail - intramedullary rod
- Austin Moore - intrameduallary rod (for Hemiarthroplasty)

Pott's disease is a presentation of extrapulmonary tuberculosis that affects the spine, a kind of tuberculous arthritis of the intervertebral joints. Scientifically, it is called tuberculous spondylitis. Pott's disease is the most common site of bone infection in TB; hips and knees are also often affected. The lower thoracic and upper lumbar vertebrae are the areas of the spine most often affected.

Pott's disease, which is also known as Pott's caries, David's disease, and Pott's curvature, is a medical condition of the spine. Individuals suffering from Pott's disease typically experience back pain, night sweats, fever, weight loss, and anorexia. They may also develop a spinal mass, which results in tingling, numbness, or a general feeling of weakness in the leg muscles. Often, the pain associated with Pott's disease causes the sufferer to walk in an upright and stiff position.

Pott's disease is caused when the vertebrae become soft and collapse as the result of caries or osteitis. Typically, this is caused by

mycobacterium tuberculosis. As a result, a person with Pott's disease often develops kyphosis, which results in a hunchback. This is often referred to as Pott's curvature. In some cases, a person with Pott's disease may also develop paralysis, referred to as Pott's paraplegia, when the spinal nerves become affected by the curvature.

A person with Pott's disease may experience additional complications as a result of the curvature. For example, an infection can more easily spread from the para-vertebral tissue, which can cause abscesses to occur. Regardless of the complications that may occur, Pott's disease is typically slow spreading and can last for several months or years.

A person who has been diagnosed with Pott's disease may be treated through a variety of options. He or she may utilize analgesics or antituberculous drugs to get the infection under control. It may also be necessary to immobilize the area of the spine affected by the disease, or the person may need to undergo surgery in order to drain any abscesses that may have formed or to stabilize the spine.

Since Pott's disease is caused by a bacterial infection, prevention is possible through proper control. The best method for preventing the disease is to reduce or eliminate the spread of tuberculosis. In addition, testing for tuberculosis is an important preventative measure, as those who are positive for purified protein derivative (PPD) can take medication to prevent tuberculosis from forming. A tuberculin skin test is the most common method used to screen for tuberculosis, though blood tests, bone scans, bone biopsies, and radiographs may also be used to confirm the disease.

As a form of extrapulmonary tuberculosis that impacts the spine, Pott's disease has an effect that is sometimes described as being a sort of arthritis for the vertebrae that make up the spinal column. It is more properly known as tuberculosis spondylitis.

Pott's disease is often experienced as a local phenomenon that begins in the thoracic section of the spinal column. Early signs of the presence of Pott's disease generally begin with back pain that may

seem to be due to simple muscle strain. However, in short order, the symptoms will begin to multiply. Night sweats may become common, along with a running fever during the day. As the condition worsens, it is not unusual to experience a loss of appetite, resulting in an anorexic state and the resulting weight loss. There is also often periods in which there is a tingling or numb sensation in the legs, accompanied with a sense of not having much strength in the legs.

Fortunately, there are several ways to determine if Pott's disease is the root cause of the symptoms. Blood tests can help determine if there is an elevation in the rate of erythrocyte sedimentation. A bone scan will determine if there is some indication of problems, which may lead to the scheduling of a bone biopsy. Conducting a CT scan as well as a radiograph of the spine is also likely to provide valuable information about the presence and current status of Pott's disease.

Once the presence of Pott's disease is confirmed, there are several treatment options available. The first line of defense will involve the use of analgesics and various antituberculous drugs, which can help to arrest the progress of the disease, as well as begin to alleviate symptoms. In some cases, it may be necessary to insert a rod into the area of the spine, providing needed stability. This is often the case if some degree of spinal cord compression has been noted. Finally, more ambitious surgery may be required, especially in situations where there is a need to drain fluid from pockets or abscesses that have formed, or if conditions indicate impending collapse of the vertebrae in the spinal column.

Investigation for Tuberculosis of Spine

Blood

- TLC: Leukocytosis.
- ESR: raised during acute stage.

Tuberculin skin test

- Strongly positive.
- Negative test does not exclude diagnosis.

Aspirate from joint space and abscess

- Transparency: turbid.
- Colour: creamy.
- Consistency: cheesy.
- Fibrin clot: large.
- Mucin clot: poor.
- WBC: 25000/cc.mm.

Histology

- Shows granulomatous tubercle.

X-Ray spine

Early:-

- Narrowed joint space.
- Diffuse vertebral osteoporosis adjacent to joint.
- Erosion of bone.
- Fusiform paraspinal shadow of abscess in soft tissue.

Late:-

- Destruction of bone.
- Wedge-shaped deformity (collapse of vertebrae anteriorly).
- Bony ankylosis.

Complications

- Vertebral collapse resulting in kyphosis.
- Spinal cord compression.
- Sinus formation.
- Paraplegia (so called Pott's paraplegia).

An highly unusual case of Pott's disease, undiagnosed until late in the destructive process, that subsequently resulted in the development of tuberculous meningitis (TBM) causing multiple cranial nerve abnormalities including blindness as recently reported.

Pott's disease treatment strategies

General management

- Bed rest.
- Immobilisation of affected joint by splintage.
- Nutritious, high protein diet.
- Drainage of abscess.
- Surgical decompression.
- Physiotherapy.

Before the advent of effective anti-tuberculosis chemotherapy, Pott's disease was treated with immobilization using prolonged bed rest or a body cast. At the time, Pott's disease carried a mortality rate of 20%, and relapse was common (30%). The duration of treatment, surgical indications, and inpatient care have since evolved. Despite questionable efficacy, prolonged recumbence and the use of frames, plaster beds, plaster jackets, and braces are still used. Cast or brace immobilization was a traditional form of treatment but has generally been discarded. Patients with Pott's disease should be treated with external bracing.

Studies performed by the British Medical Research Council indicate that tuberculous spondylitis of the thoraco-lumbar spine should be treated with combination chemotherapy for 6-9 months. According to the most recent recommendations issued in 2003 by the US Centers for Disease Control and Prevention, the Infectious Diseases Society of America, and the American Thoracic Society, a 4-drug regimen should be used empirically to treat Pott's disease. Isoniazid and rifampin should be administered during the whole course of therapy. Additional drugs are administered during the first 2 months of therapy. These are generally chosen among the first-line drugs,

which include pyrazinamide, ethambutol, and streptomycin. The use of second-line drugs is indicated in cases of drug resistance.

The cornerstone of treatment for Pott's disease and TBM involves combination chemotherapy. However, recent increases in drug-resistant organisms have led to the addition of a fourth or even a fifth anti-TB drug consisting of ethambutol and/or streptomycin.

While controversial, several clinical studies have also shown corticosteroids (12 mg/day in adults, 8 mg/day in children weighing less than 25 kg) to improve the morbidity and mortality associated with TBM.

Given the effectiveness of chemotherapy in even those patients with mild neurologic deficit, surgery is now predominantly reserved for significant or progressive neurologic deficit and deformity correction.

Regarding the duration of therapy, the British Medical Research Council studies did not include patients with multiple vertebral involvement, cervical lesions, or major neurologic involvement. Because of these limitations, many experts still recommend chemotherapy for 9-12 months.

Opinions differ regarding whether the treatment of choice should be conservative chemotherapy or a combination of chemotherapy and surgery. The treatment decision should be individualized for each patient.

While the role of surgery in Pott's disease is controversial, some have advocated surgery for every infected site. Routine surgery does not seem to be indicated. Surgical drainage of the abscess as well as debridement and stabilization can be indicated, even though spine-related neurologic deficits were relatively mild.

Most common indications for surgical procedures are discussed below:

Surgical Care

Indications for surgical treatment of Pott's disease generally include the following:

- Neurologic deficit (acute neurologic deterioration, paraparesis, paraplegia)
- Spinal deformity with instability or pain
- No response to medical therapy (continuing progression of kyphosis or instability)
- Large paraspinal abscess
- Non-diagnostic percutaneous needle biopsy sample

Resources and experience are key factors in the decision to use a surgical approach. The lesion site, extent of vertebral destruction, and presence of cord compression or spinal deformity determine the specific operative approach (kyphosis, paraplegia, tuberculous abscess). Vertebral damage is considered significant if more than 50% of the vertebral body is collapsed or destroyed or a spinal deformity of more than 5° exists. The most conventional approaches include anterior radical focal debridement and posterior stabilization with instrumentation. In Pott's disease that involves the cervical spine, the following factors justify early surgical intervention:

- High frequency and severity of neurologic deficits
- Severe abscess compression that may induce dysphagia (difficulty in swallowing) or asphyxia (lack of oxygen)
- Instability of the cervical spine

Contraindications

Vertebral collapse of a lesser magnitude is not considered an indication for surgery because, with appropriate treatment and therapy compliance, it is less likely to progress to a severe deformity.

The duration of treatment is somewhat controversial. Although some studies favor a 6- to 9-month course, traditional courses range from 9 months to longer than 1 year. The duration of therapy should be individualized and based on the resolution of active symptoms and the clinical stability of the patient.

Isoniazid

Highly active against *Mycobacterium tuberculosis*; has good GI absorption and penetrates well into all body fluids and cavities.

Adult

300 mg PO qd; alternatively, 15 mg/kg IV qd

Pediatric

10 mg/kg PO qd

Rifampin

For use in combination with at least one other antituberculous drug; inhibits DNA-dependent bacterial but not mammalian RNA polymerase. Cross-resistance may occur.

Adult

10 mg/kg PO qd; not to exceed 600 mg/d

Pediatric

10-20 mg/kg PO qd; not to exceed 600 mg/d

Pyrazinamide

Bactericidal against *M tuberculosis* in an acid environment (macrophages); has good absorption from the GI tract and penetrates well into most tissues, including CSF.

Adult

15-30 mg/kg PO qd

Pediatric

Not established

Ethambutol

Has bacteriostatic activity against *M tuberculosis*. Has good GI absorption. CSF concentrations remain low, even in the presence of meningeal inflammation.

Adult

15-25 mg/kg PO qd

Pediatric

15-25 mg/kg PO qd

Not recommended for young children because of difficulty monitoring vision

Streptomycin

It is bactericidal in an alkaline environment. Because it is not absorbed from the GI tract, must be administered parenterally. Exerts action mainly on extracellular tubercle bacilli; only about 10% of the drug penetrates cells that harbor organisms; enters the CSF only in the presence of meningeal inflammation. Excretion is almost entirely renal.

Adult

15 mg/kg IM qd; not to exceed 1 g/d

Pediatric

20-40 mg/kg IM qd; not to exceed 1 g/d

Diagnosis of Pott's disease is challenging and misdiagnosis is common, contributing to the development of potentially severe neurologic deficits. Knowledge of recently recognized risk factors for extrapulmonary TB indicates that regardless of risk factors it is crucial to maintain a high index of suspicion in all such cases. Pott's disease will remain a significant health risk in the near future.