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Original Research Article

A review of Pott's Disease

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Abstract

Pott's disease is an eponym for tuberculosis of the spine as described by Percivall Pott in 1779. It is a destructive disease of the spine caused by Mycobacterium tuberculosis, one of the oldest diseases of mankind and a major cause of deaths worldwide. It is a dangerous disease with associated severe complications such as Pott's paraplegia, spinal deformities and severe abscesses. It accounts for half of the skeletal tuberculosis and a disease of global concerns with highest incidence in developing world. The diagnosis is often late and difficult in Nigeria. In northern Nigeria the presentation of a young adult with chronic back pain and angular kyphosis with or without abscess or neurological deficit is often suggestive of Pott's disease. The gold standard for laboratory diagnosis is the biopsy for confirmatory acid-fast bacilli in culture and sensitivity using Lowenstein - Jensen medium or BACTEC radiometric culture. Magnetic resonance imaging is the gold standard for radiological diagnosis of Pott's disease. The main stay of treatment is medical including the use of evidence- based multi-anti-tuberculosis drugs and bracing. Surgery is supplementary. Early diagnosis and treatment are key to successful management. The aim of this review was to highlight the burden of tuberculosis, describe surgical pathology, presentation, diagnosis and options of treatment of Pott's disease in a developing country.

Keywords: Pott's disease, back pain, angular kyphosis, presentation, diagnosis, treatment, outcome, Nigeria

Introduction

Tuberculosis (TB) is one of the oldest disease of man. It remains a disease of significant public health importance. It has assumed a global epidemic status with over 2 billion infected worldwide. Majority of the cases are in the developing nations and the incidence is increasing worldwide due to high rate of HIV – TB, multiple drug resistant strains of tuberculosis and high rate of immigrants from developing countries to developed countries [1-2]. There are over 10 million new cases of TB worldwide and over one million deaths annually. The disease commonly affects the lung, but involves extra pulmonary structures in 10 – 15% of cases. Of the extra pulmonary tuberculosis (EPTB) skeletal TB is second to only lymph nodes involvement [1-2].

TB of the spine (Pott's disease) accounts for 50% of the skeletal tuberculosis. Pott's disease is a dangerous disease with associated severe complications such as Pott's paraplegia, spinal deformities and severe abscesses [1-10].

Pott's disease

Pott's disease is the eponym for spinal tuberculosis as described by Percivall Pott in 1779 in his monogram [3], though the disease was first reported by Hippocrates (100–300 BC), a disease as old as man is endemic worldwide [1-3]. It is a destructive spine disease caused by *Mycobacterium tuberculosis* (MBT) which is a tubercle bacilli. It accounts for half of the skeletal tuberculosis (STB) and a disease of global concerns with highest incidence in developing world, a common cause of non – traumatic paraplegia [1-10]. Therefore, early diagnosis and treatment are key to

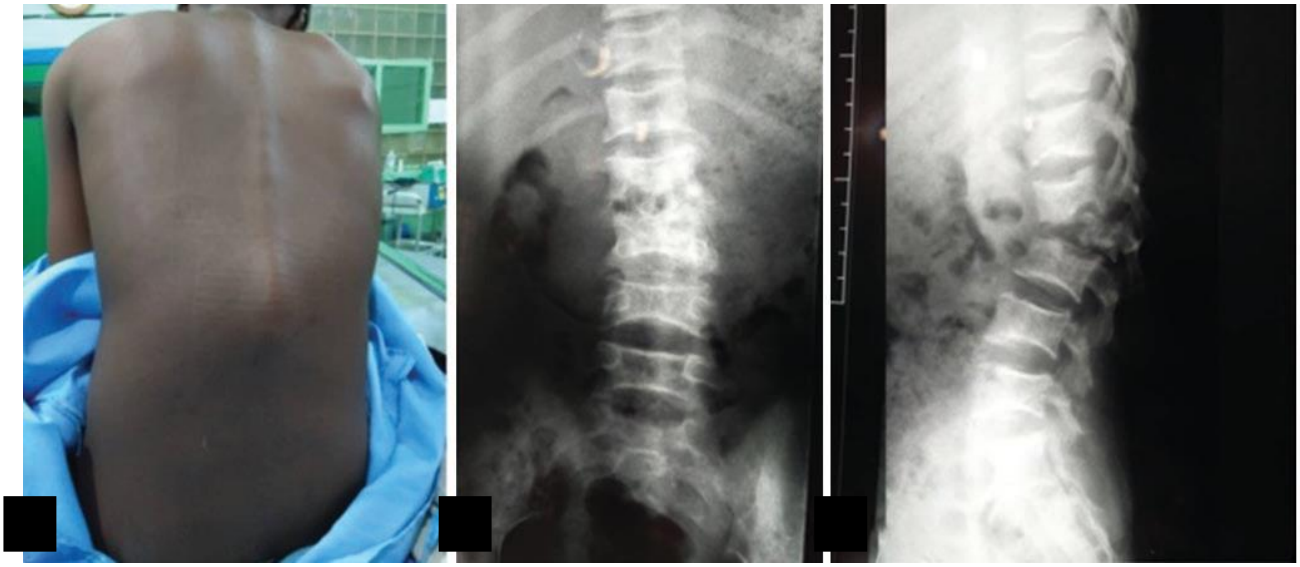

Figure 1: Common clinical features of Potts disease seen in Nigeria: (a) A painful gibbus in a 16 – year – young lady at the thoracolumbar region and (b and c) plain x-ray of the same lady showing complete collapse and disappearance of L2 vertebral body and angular kyphosis with no neurological deficit [1]

Figure 2: Microscopic view of *Microbacterium tuberculosis* (available at www.slideshare.net)

successful management. The clinical and radiological features of the disease are as described in Fig. 1.

Mycobacterium tuberculosis

MBT, the causative organism of Pott's disease, is a rod-shaped, non-spore forming, fastidious aerobic weakly Gram-positive bacterium classified as acid – fast bacilli [1-2] because, once stained, the bacilli cannot be decolorized by acid alcohol. The acid fastness is due to the high content of mycolic acids and other cell – wall lipids. Microscopic views of the organism is as shown in Fig. 2.

MBT cell wall and antibiotics

The mycolic acids are lined to the underlying aribinogalactans and peptidoglycan resulting in very low permeability of the cell wall, thus reducing the effectiveness of most antibiotics. The structure of the MBT cell wall is shown in Fig. 3.

MBT colony and antibiotics

MBT exist in four colony types and this necessitates the adoption of multiple drug therapy in keeping with the varying levels of effectiveness of antibiotic among the colony types. The choice of antitubercular therapy (ATT) based on the colony type is shown in Table 1.

In this era of HIV and hepatitis the injectable form of ATT, streptomycin has been replaced with oral ethambutol to reduce the risk of transmission of infection from needle prick.

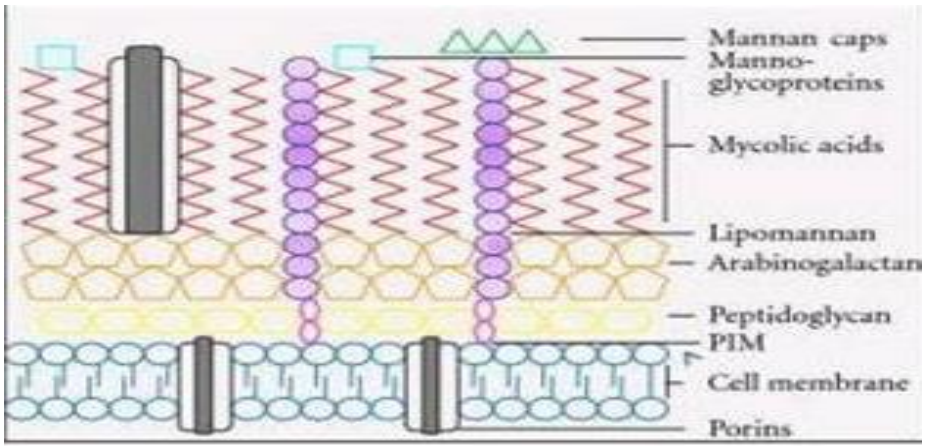

Figure 3: The structure of Microbacterium tuberculosis cell wall (available at www.slideshare.net)

Table 1: The bases for choice of multiple ATT

Types	Colony	Effective Antibiotics
1.	Extracellular rapidly dividing bacilli	Isoniazid / Ethambutol / Streptomycin
2.	Extracellular slowly dividing bacilli	Rifampicin
3.	Intracellular slowly dividing bacilli	Pyrazinamide
4.	Dormant colony	No effective drug and this explains reason for relapse

Predisposing factors to Port's disease

The common predisposing factors to Pott's disease are malnutrition, poor sanitation, overcrowding, close contact with TB patients, multiple pregnancy and immunodeficiency status. These are common state in most developing countries including Nigeria [1, 2, 4-9]. The disease has become a worldwide phenomenon owing to high state of HIV – TB cases, multiple drug resistant strains of MBT and high rate of migrants from developing countries to developed nations [1, 2].

Surgical Pathology

Pott's disease is a secondary or tertiary form of TB. The spread is haematogenous with characteristic caseous granuloma with less toxin and biofilms production [1, 2]. Blood supply of the vertebrae favors involvement of bones on each side of the disc with relative disc sparing in adults. A typical tuberculous granuloma is described in Fig. 4 and the vertebral blood supply are shown in Fig. 5.

There are two forms of the Pott's disease:

a) Intradural involvement which is rare and
b) Extradural involvement which is common and consist of four types:
   i. Paradiscal which is common and occurs by arterial spread is associated with wedge compression collapse of the adjacent vertebral bodies and angular kyphosis (Fig. 6a).
   ii. Central which affects the vertebral body and occurs by venous spread through the Batson's valveles plexus of veins. This presents often as vertebral plana and disc involvement is late, making mitotic lesion a strong differential diagnosis at the early stage of the disease (Fig. 6b).
   iii. Anterior, is a subperiosteal disease from arterial spread with a scalloping lesion on the arterior part of the vertebral body to form a tumor – like abscess mass behind the aorta with a pseudo aneurysmal features (Fig. 6c).
   iv. Posterior, is an appendicular lesion from arterial spread with atypical abscess formation (Fig. 6d).

Pott's disease is truly a chronic spondylodiscitis and vertebral osteomyelitis.

Sequale of Pott's Disease

The sequale of Pott's disease are **caseous** abscess of the spine, deformity, mechanical instability cord compression and neurological deficit [1, 2, 9]. Massive abscesses of the cervical spine commonly present as retrophargeal abscess, but can present in the anterior and posterior triangles of the neck or in the axilla as the abscesses track along the plane of least tissue resistance following neurovascular planes around the neck. The retropharyngeal abscess can produce dysphagia, hoarseness of voice and respiratory stridor. In the thoracic spine the abscesses present commonly as fusiform paravertebral swelling radiologically, but may track along the neurovascular planes to present as anterior chest wall mass or present as mediastinal mass or pass through the opening in the diaphragm to present as retroperitoneal mass. In the lumbar spine the abscesses may present

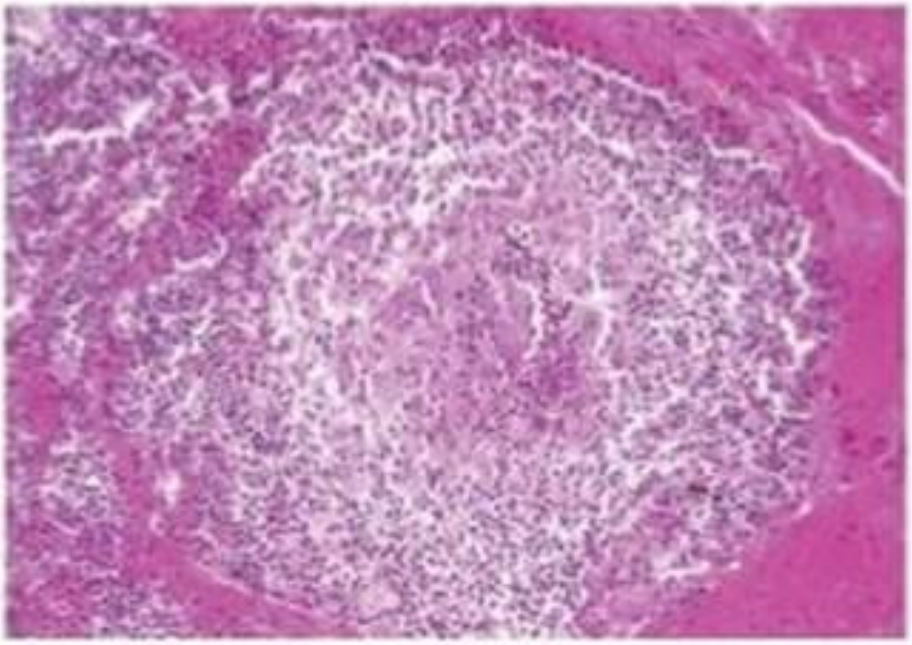

Figure 4: The microscopic structure of tuberculous granuloma showing typical central necrosis with scattered giant cells surrounded by lymphocytes and histiocytes (available from www.slideshare.net)

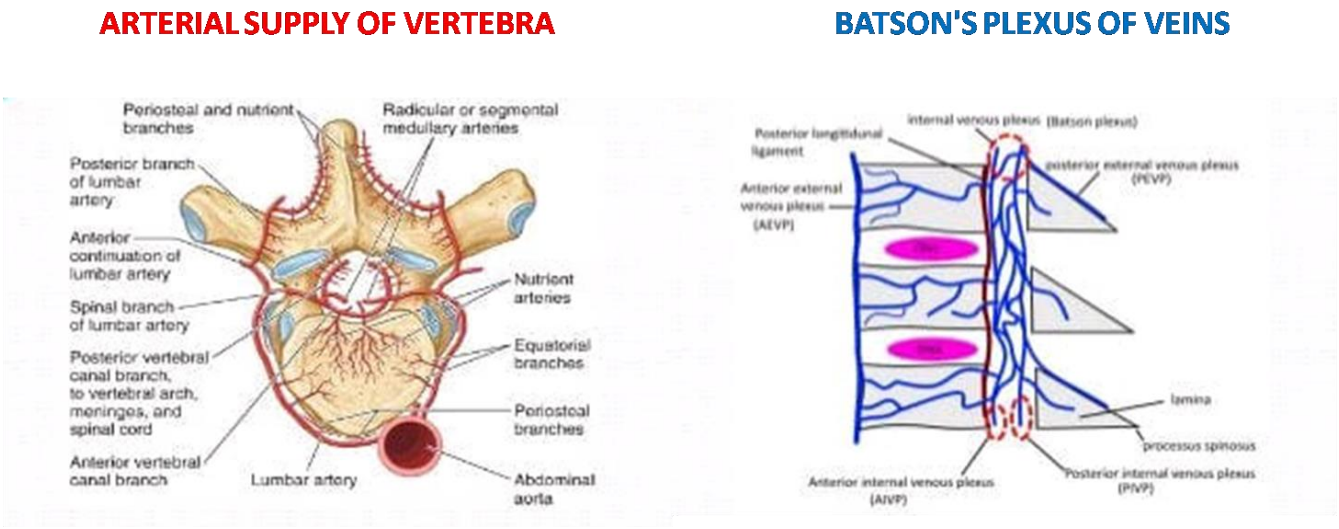

Figure 5: Vertebral blood supply. (available from www.slideshare.net)

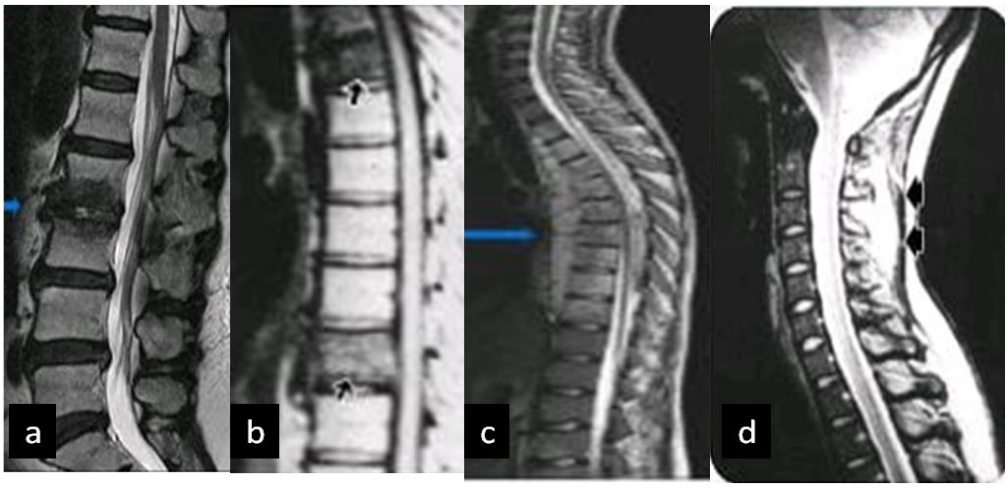

Figure 6: Magnetic resonance imaging – T2 weighted – showing various types of spinal tuberculosis (blue and black arrows): (a) paradiscal (b) central (c) anterior (d) appendicular spondylodiscitis. Images derived from www.slideshare.net

in the Petit's triangle as lumbar abscess or pass along the plane of psoas muscle to present as psoas abscess and cold abscess of the hip joint causing pseudo – flexion deformity of the hip or follow neurovascular plane to present as cold abscess in the femoral canal, perineum and popliteal fossa [1, 2].

The spine deformity and mechanical instability may result from spine at risk signs in children, panvertebral or bilateral facetal involvement, multifocal contignous disease involving more than 3 vertebral, vertebral collapse of more than 50%, kyphosis more than 30 – 40 degrees and anteroposterior or lateral translation [1, 2, 10].

The cord compression and neurological deficit may result from mechanical causes such as granulation tissue, sequestrum, prolapsed disc and extradural or intradural tuberculoma. The non – mechanical causes include pachymeningitis, thrombophlebitis and endarteritis [1, 2].

The thoracic spine is the common region of involvement in Pott's disease in about 42% of cases. This is followed by Lumbar spine in 26%, thoracolumbar spine in 12%, cervicodorsal spine in 5% and Lumbosacral spine in 3% of cases [1].

Complications

The complications of Pott's disease include Pott – Paraplegia, cold abscesses and sinuses, kyphosis and instability, secondary infections, amyloidosis and fatality [1, 2].

Classifications

There are several classifications of Pott's disease, but for a sense of direction on best form of treatment to institute the disease is simply classified as complicated and uncomplicated. The complicated cases would require surgical treatment and for the uncomplicated cases medical treatment suffices [1- 3].

Staging

There are several staging methods, but the modified Frankel/American Spinal Injury Association (ASIA) staging is used to predict severity of injury and outcome of treatment. This stages motor & sensory cord function following injury from A to E. Stage A represents severe motor and sensory injuries with poor outcome and stage E represents normal motor and sensory function. The staging is summarized as follows [1, 2]:

A – Complete motor and sensory deficit
B – Complete motor but partial sensory deficit
C – Intact sensory but useless motor < 3
D – Intact sensory but useful motor > 3
E – Normal motor and sensory functions

Clinical Features

The clinical features of Pott's disease are variables. Back pain is the common complaint in patients with Pott's disease. This is followed by cold abscess, deformity and features of neurological deficits (Fig. 7).

Diagnosis

The diagnosis of Pott's disease is difficult and late especially in Nigeria. The combination of clinical, radiological and laboratory findings suffice. When in doubt, start therapeutic trials, though current practice is evidence based from microbiological and histopathological results of biopsy.

Laboratory Diagnosis

The gold standard for laboratory diagnosis is the biopsy for confirmatory acid-fast bacilli (AFB) in culture and sensitivity using Lowenstein & Jansen (L&J) culture media – this take about 4 weeks for the result to be ready, but the BACTEC radiometric culture takes about 2 weeks only. The classical granuloma on histopathological assay is typical of Pott's disease.

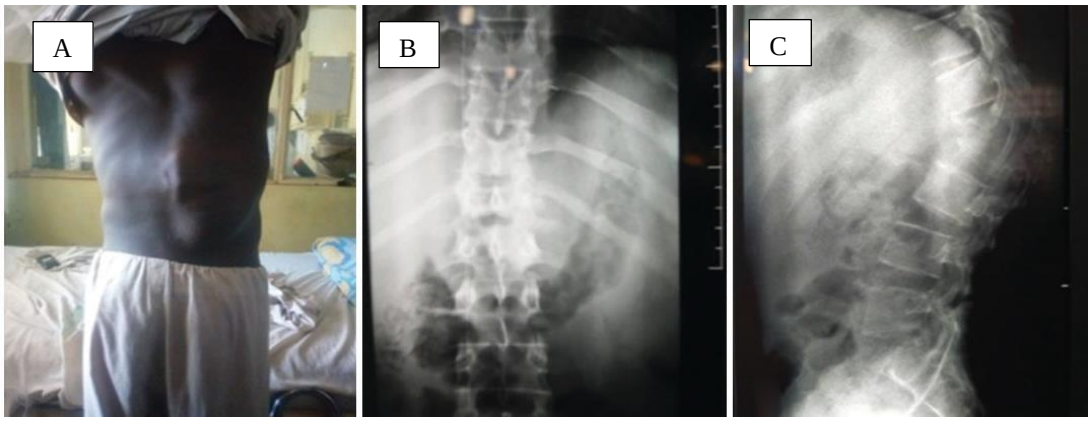

Figure 7: Common clinical features of Pott's disease in Nigeria: (a) a painful thoracolumbar gibbus in a 20 – year – old man with paraspinal cold abcess extending to the right lumbar triangle and paraparesis (b and c) plain x-ray of the same man showing wedge collapse fracture of L2 vertebral body and angular kyphosis [1].

Microscopy for AFB in Zeihl-Neelsen (Z & N) stain is often helpful in diagnosis. The use of commonly employed Mantoux text is limited by so many factors and is considered not reliable, but other immunological assay such as Interferon Gamma Release Assays (IGRAs) are more reliable and this includes: The QuantiFERON – TB GOLD PLUS and LIOFeron TB / LTBI. The molecular assays such as Gen Xpert MTB / RIF which takes about 90 mins and the new generation Xpert MTB / RIF Ultra are more sensitive and specific tests. However, the haematological assays of erythrocytes sedimentation rate (ESR) and polymerase chain reaction (PCR), though nonspecific are useful in monitoring the outcome of treatment [1, 2].

Imaging

Imaging techniques for diagnosis of Pott's disease are useful in the assessment of the extent of the lesions, guide collection of samples and assist in defining the best treatment strategy. The imaging modalities employed in the diagnosis of Pott's disease include the following: Plain X – ray, computerized tomography (CT) scan, magnetic resonance imaging (MRI) and fluorodeoxgylglucose - positron emission tomography (FDG – PET). MRI is the gold standard of the imaging techniques in the diagnosis of Pott's disease.

Plain X – ray

The lateral standing plain X – ray is used in the assessment of the kyphotic angle. There are two standard methods of measurement of kyphotic angles: The Dickson and MRC used in the measurement of Kyphotic angle are as shown in Fig. 8.

Plain X – ray is also use in the assessment of the 'Spine at risk' signs in children. These include separation of facets, retropulsion, lateral translation and toppling signs seen in severe deformity and instability in children with Pott's disease necessitating surgery (Fig. 9).

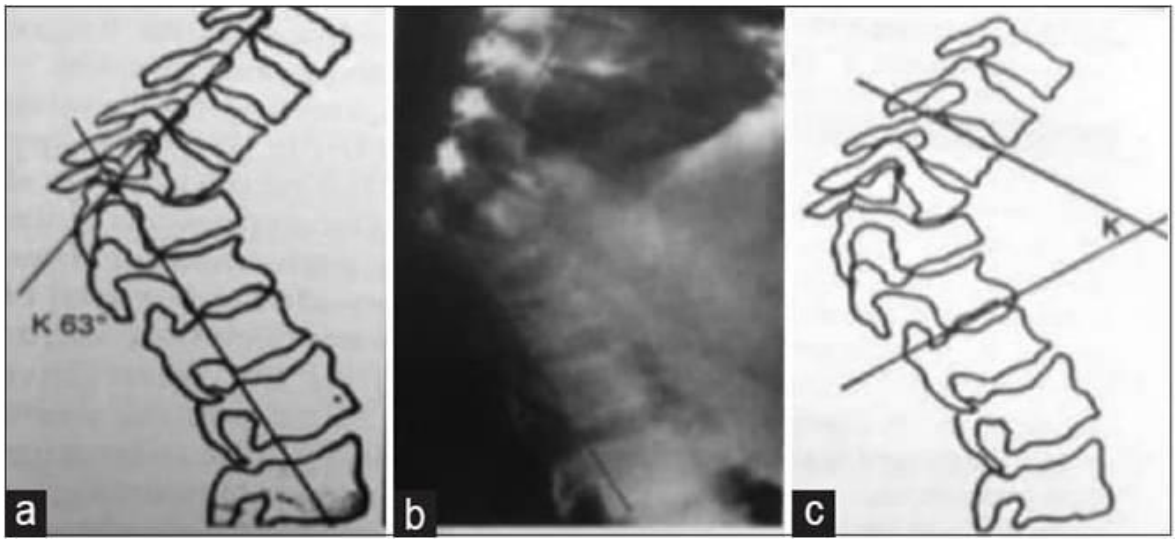

Figure 8: Measurements of kyphotic angle: (a) Dickson method, a line is drawn along the posterior margin of the bodies of the healthy vertebral above and below the site of disease, angle K is the angle of kyphosis; (a) lateral spine radiography used for measuring the kyphotic angle and (c) Konstam and Blesovsky (Medical Research Council) method, is by determining the angle between the upper - end - plate of the normal vertebral proxima to the affected vertebrae and the lower – end - plate of the normal vertebral distal to the affected vertebrae. Angle K increases with increase in the angle of kyphosis. Derived from the internet, www.slideshare.net [1]

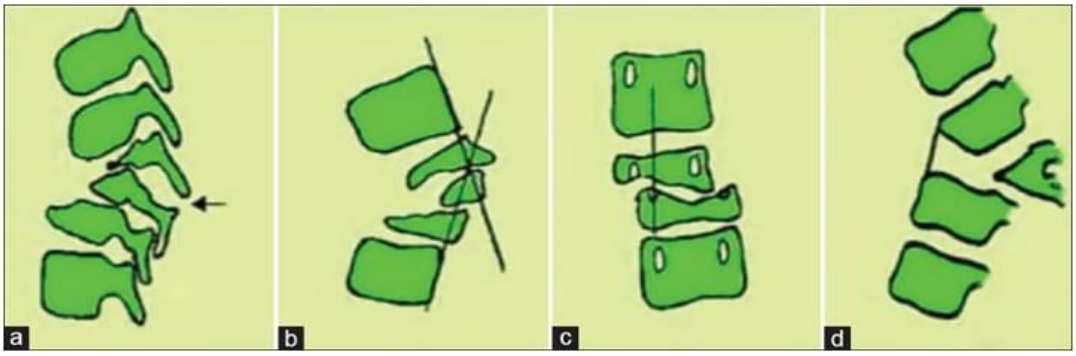

Figure 9: Spine at risk sign in children; (a) separation of facets joint (b) retropulsion (c) lateral translation (d) toppling sign. These signs of spinal instability in children are indications for surgical intervention in children with Pott's disease. Derived from internet, www.slideshare.net [1]

CT and MRI

CT scan help in early review of vertebral osteolysis and kyphosis. MRI which is the gold standard in early diagnosis of Pott's disease allows for early detection of inflammatory oedema, extent of abscess and visualization of spinal cord lesion. Owing to cost constraints these diagnostic tools are used in a few selected cases and in majority of cases we rely on plain x-ray films for the assessment of patients, culminating in late diagnosis and treatment.

FDG -PET / CT

FDG – PET / CT may be superior to MRI in the detection of Pott's disease, but expensive and not a readily available tool in Nigeria [1].

Differentials

The differential diagnosis of Pott's disease includes the followings: pyogenic spondylodiscitis / osteomyelitis, atypical degenerative disc disease, other granulomatous diseases, multiple myeloma, eosinophylic granuloma, metastatic spine diseases. It is important to emphasis that in the differential diagnosis between metastatic diseases and infective diseases of the spine, it is common to say 'Bad disc' of infective diseases connotes good news and 'Good disc' of metastatic diseases connotes bad news.

Treatment of Pott's disease

The principles of treatment of Pott's disease have evolved from watchful observation to active evidence-based combination therapies of medical and surgical treatment. The changing trends in principles had led to controversies in literatures about the best treatment method, indications, approaches and techniques. Varying degrees in outcome of treatment have been reported between methods; albeit, there is no standard one fit all method [1- 3, 11-16]. However, the middle path regimen reported by Tuli is generally an accepted practice [13].

Pharmacotherapy

In majority of cases uncomplicated Pott's disease are suitable for medical treatments, and multi – anti – tuberculosis drugs are the mainstay of treatment, but with mean increase deformity of 15 degrees and in 3 – 5% final deformity is more than 60 degrees [1- 3, 10, 11]. There are various regimen, but no consensus on the duration and mode of treatment. The common mode are the direct observation treatment system (DOTS) and ambulant form of therapy with or without brace [1, 2, 9].

The problems with medical treatment are multi drug – resistant strains (MDR –TB, a resistance to Isoniazid and Rifampicin), Extensive drug - resistant strains (XDR – TB, a resistance to Isoniazid and Rifampicin along with resistance to any flouroquinolone and at least one parenteral second-line antitubercular drug), Totally drug – resistant strains (TDR – TB, a resistance to all available antitubercular drugs), non – compliance and association with HIV / AIDS making treatment difficult with poor outcome [1 - 3, 11-13].

The multi – anti – tuberculosis drug therapy consists of two phases. There is an initiation phase, which start with 4 first line drugs for two months, comprising of [1 - 3, 12, 13]:

1. Isoniazid, 5 -10 mg/kg for a maximum dose of 300mg per day
2. Rifampicin 10 – 20 mg/kg for a maximum dose of 600mg per day
3. Ethambutol 15 – 25 mg/kg for a maximum dose of 800mg per day
4. Pyrazinamide, 25 – 40 mg/kg for a maximum dose of 1.2gm per day

In the course of treatment, watch out for complications and deal accordingly.

In the continuation phase isoniazid and rifampicin are given for varying length of time based on the physician perdegree and experiences. The followings are the practicable continuation regimens based on the length of therapy [1, 2, 12]:

1. 4 months for the 6 months British regimen
2. 7 months for the 9 months American regimen
3. 10 months for the 12 months WHO regimen
4. 16 months for the 18 months old school regimen.

We believe in the old school regimen and would recommend nothing less than 12 months regimen for the developing nations. The second line drugs include Kanamycin, Amikacin, capreomycin and Levofloxacin. These second line drugs are more toxic and should be used with care. There are also new drugs such as Dalamanid, Bedaquiline, SQ109 and Dutezolid which are on trial for drugs resistant strains [1, 2, 12].

Surgical Treatment

Surgery remains complementary to standard multi – drug therapy. However, controversy remains regarding indications, approaches and modality of instrumentation [1 - 3, 10, 11, 13-16]. It is advisable that first line multi antituberculosis drugs should be administered for two to three weeks before surgery and continued for three months after surgery and thereafter the usual continuation phase is administered. And because of the low production of biofilm by the MBT it is save to use spine implants even in the presence of active infection [15].

The indications for surgery may include neurological deficit, extensive abscesses, severe deformity with or without instability, failure of medical treatment and cosmesis especially among young adults preparing for marriage in northern Nigeria [1]. The main goals of surgical treatment are to achieve adequate decompression, debridement, maintenance and

reinforcement of stability, correction and prevention of deformity. The surgical approach to the spine are anterior, posterior, combined and minimally invasive surgery (MIS).

Anterior approach: The anterior approach was popularized by Hodgson in 1960 [14] which directly deal with the pathology with wide exposure and direct visualization across the midline [1 - 3]. Anterior reconstruction can be performed more efficiently, and quality of debridement and bony fusion is optimal.

Posterior approach: Posterior approach is the best means of achieving reduction and stable saggital alignment overtime. It allows for ease of approach with less learning curve, less morbidity and global reconstruction is made possible by posterior alone approach [1-3,]

Combined approach: Combined approach is indicated in cases with severe instability, severe kyphotic deformities and is vital in osteoporotic bones. It can be done in 1 or 2 stages, but associated with high morbidity, hence not advisable in less severe cases [3]

MIS: The MIS is indicated in high-risk patients with no neurological deficit. It is used either as standalone or in combination with open procedures. MIS is useful as in thorascopic debridement, posterolateral endoscopic debridement and video – assisted surgery for anterior debridement and bone grafting [1-3]

Case illustration

A 16 – year – old lady with painful tuberculosis kyphosis of the lumbar spine. There was no neurological deficit, plain X – ray showed complete collapse and disappearance of L2 vertebral body and angular kyphosis. She had uneventful posterior decompression, stabilization and posterolateral fusion (Fig. 10).

In the surgical treatment of Pott's disease at the thoracolumbar spine, one – stage posterior only approach is advocated as illustrated in fig. 10 above [1-3]. Zhang et al 2013, have also described a one – stage posterior only approach for combined interbody and posterior fusion [16]. Agreed, surgery in healed TB Spine for kyphosis more than 60 degree combined approaches of anterior corpectomy, posterior shortening and instrumentation plus anterior and posterior fusion suffice, but the risk of blood loss is high. There are several options for posterior deformity correction and this include transpedicular decancellation, pedicle subtraction osteotomy, posterior closing wedge osteotomy, posterior vertebral column resection and closing opening wedge osteotomy [1 - 3].

The surgical procedures in the treatment of Pott's disease include, but not limited to the followings:

1. Anterior radical debridement and strut grafting
2. Posterior instrumentation and bone grafting with anterior radical debridement and strut grafting in a single or two stage procedure
3. Extra plural anterior lateral decompression and strut grafting with posterior instrumentation in thoracic or thoracolumbar spine
4. Single stage transforminal decompression and posterior instrumentation and fusion

Decision making for surgery therefore should be individualized with reference to age, location of bony lesion, presence of medical comorbidity, severity of kyphosis, numbers of vertebral levels affected, region of spine involve, and experience and preference of surgeon.

The recent trend is for all posterior global reconstruction in thoracic and lumbar spine. However, anterior debridement and fusion remains the standard of care in lower cervical TB.

The combined procedures become invaluable in patient with large vertebral defects involving 2 – 3 vertebrae, revision surgeries and in thoracolumbar TB.

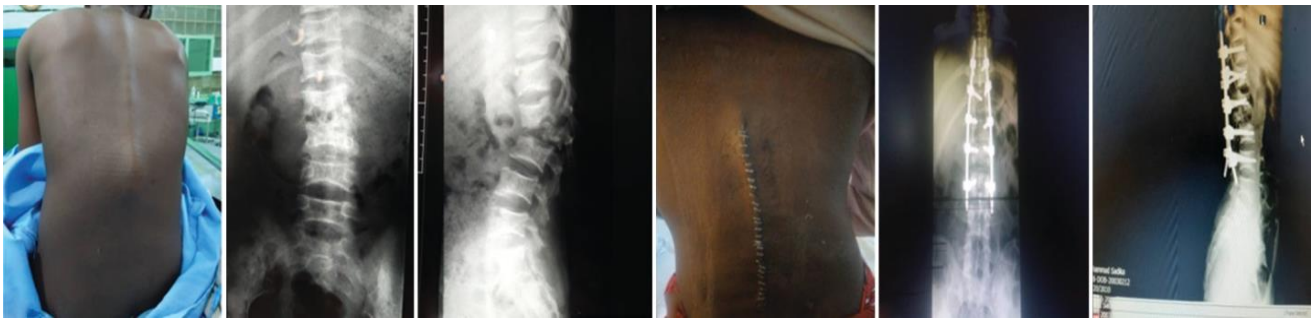

Figure 10: Surgical treatment of tuberculosis kyphosis and instability through a single stage posterior only approach in: (a) A 16-year-old lady with tuberculosis kyphosis of the lumbar spine, (b and c) Plain X-ray of the same lady showing kyphosis due to completer destruction of second lumbar vertebral body, d) Post-operative appearance of the same lady, (e and f) Post-operative plain X-ray of the same lady following decompression, osteotomy for correction of the deformity, posterolateral fusion and stabilisation with pedicle screws and rods construct. The lady was discharged home 2 weeks post-operative period with good cosmesis and intact neurology. Majority of our young patients with tuberculosis spine kyphosis aside pain and interference with marital functions, request for cosmetic surgery in the immediate perimarital period [1].

Conclusion

The burden of Pott's disease is rising in Nigeria owing to increasing cases of tuberculosis. Chronic back pain and angular kyphosis with or without abscess and neurological deficit are the main presentation. Late diagnosis and treatment are the major problems in our settings. The prognosis is good if treated early. Multi antituberculosis drugs remains the main trust of treatment. Surgery is supplementary and when indicated, takes the form of spinal debridement and fusion with or without instrumentation.

List of abbreviations

AFB – Acid-fast bacilli
CT – Computerized tomography
ESR – Erythrocytes sedimentation rate
FDG-PET– Fluorodeoxgylglucose - positron emission tomography
MRI – Magnetic resonance imaging
MIS – Minimally invasive surgery
MBT – *Mycobacterium tuberculosis*
TB – Tuberculosis
WHO – World Health Organization

Declarations

Ethical approval

None provided.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

No conflict of interest associated with this work.

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Contribution of Authors

We declare that this work was done by the authors named in this article and all liabilities pertaining to claims relating to the content of this article will be borne by the authors.

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