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Case Report

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A Peculiar Concurrent Presentation of Spinal Tuberculosis In A Diagnosed Case of Neurofibromatosis Type 1 – An Atypical Case Report

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Abstract

Spinal tuberculosis is a prevalent ailment observed in the Asian demographic, which can result in neurodeficit if not addressed during the initial phases. Neurofibromatosis is a hereditary disorder that results in the manifestation of dystrophic bony characteristics accompanied by the development of multiple spinal tumors. This study presents a unique case of a young female who had previously been diagnosed with Neurofibromatosis. Additionally, she exhibited symptoms of tubercular spondylodiscitis, resulting in kyphotic deformity and paraparesis. The present case report discusses the challenges encountered during the pre-operative and intra-operative management of a patient presenting with the co-occurrence of neurofibromatosis and tuberculosis of the spine. Despite the advancements in imaging modalities, such as magnetic resonance imaging, and surgical techniques, the prompt diagnosis and management of spinal tuberculosis with Neurofibramatosis remains complex and demanding. The objective of this review is to examine the diagnosis, preoperative planning, and treatment of spinal tuberculosis in a patient of Neurofibromatosis type 1, utilizing studies that possess adequate literature, lucidly presented outcomes, and defensible conclusions.

Keywords: Spinal Tuberculosis; Decompression; Potts spine; Neurofibromatosis type 1

Introduction and Background

Infective spondylodiscitis is a medical condition characterized by inflammation of the intervertebral disc and contiguous vertebral bodies seen in endemic areas due to a pathogen called Mycobacterium tuberculosis. Spinal TB accounts to almost 50% of all extrapulmonary TB[1]. This condition typically manifests in the thoracolumbar region, although it may also occur in other regions of the spinal column. Spinal tuberculosis is a disease that is subject to constant evolution due to changing disease patterns, advancements in imaging, and treatment protocols. The emergence of drug resistance also poses a significant challenge in managing this disease. Neurofibramatosis on the other hand is a condition affecting the neural crest cells that is described as a spectrum of complex illnesses with likely hamartomatous origins that affect the neuroectoderm, mesoderm, and endoderm. The condition results in the development of non-malignant neoplasms on or beneath the dermis, as well as on the peripheral nervous system and other anatomical structures[2]. Neurofibromatosis (NF) follows an autosomal dominant inheritance pattern and is characterized by distinct clinical manifestations such as hyperpigmented spots, neurofibromas, Lisch nodules, skeletal and bony anomalies, and a predisposition to neoplastic growths[3]. Neurofibromatosis comprises two distinct subtypes, namely type 1 and type 2. The most prevalent type is NF 1,

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which exhibits a greater incidence of spinal tumors[4].

This study presents a unique case in which a patient with a history of NF1 was found to have spinal TB resulting in spinal cord compression and paraparesis.

Case Report

A 32-year-old female patient presented to our clinic with severe mid-back pain of 10-day duration. She reported gait imbalance since the last 2-3 days, which had progressively worsened. On examination, she had severe tenderness in the mid dorsal spine at levels D7-D10, with weakness of MRC grade 3 in both ankle dorsiflexion and MRC grade 4 knee extension. Sensations were preserved at all dermatomes, and proprioception was normal. She also reported a positive history of constitutional symptoms including, weight loss and evening rise in temperature for the past 2-3 months. Past medical history and examination revealed that the patient was a known case of Neurofibromatosis Type 1, with features such as cafe-au-lait lesion on skin, but was otherwise asymptomatic. Patient was unable to walk due to buckling of knee and required support to walk more than 10 feet.

Investigations

Routine laboratory investigations including blood count and biochemistry were normal, but the total leukocyte count was raised to 22,000. Diagnostic imaging in the form of Magnetic Resonance Imaging (MRI) (Fig 1)and plain X-ray dorso-lumbar spine(Fig 2) was carried out, which showed presence of an infective disease process at D7, D8, D9, and D10 levels. There was an evidence of fluid collection in the pre- and paravertebral regions, with intraspinal anterior epidural extension noted at the D8-D9 level causing severe spinal cord compression.(Fig 3) There was also a non-compressive finding of a multiple meningoceles in the left neural foramina of D6-D11 levels with the largest meningocele measuring 7*6.5*5.3 cm in the left paravertebral region.(Fig 4) The possibility of tuberculosis was suspected, and a clinico-pathological correlation, including diagnostic aspiration of the prevertebral fluid collection, was recommended.

Pre Operative Planning And Treatment

Given the presence of paraparesis and an infectious focus exerting pressure on the spinal cord at the D7-D10 level, it was determined that a surgical intervention involving decompression and stabilisation, as well as drainage of the epidural abscess, was warranted. A computed tomography (CT) (Fig 5)scan was performed to assess the viability of the pedicles, as pedicle destruction is a characteristic feature of Neurofibromatosis Type 1. The computed tomography scan indicated the complete destruction of the pedicles on the right side of the D8-D11 vertebrae and on the left side of the D9-D11 vertebrae. The pedicles located both above and below were observed to be dystrophic and narrow.

A computed tomography (CT) scan was performed, utilising a three-dimensional (3D) reconstruction, in order to preoperatively plan the trajectories of screws and determine the appropriate size of screws that could be utilised for pedicle screw fixation.

Following preoperative planning, the patient underwent decompression and fixation surgery through a conventional midline posterior approach, which involved transpedicular decompression of the spinal cord and drainage of the epidural

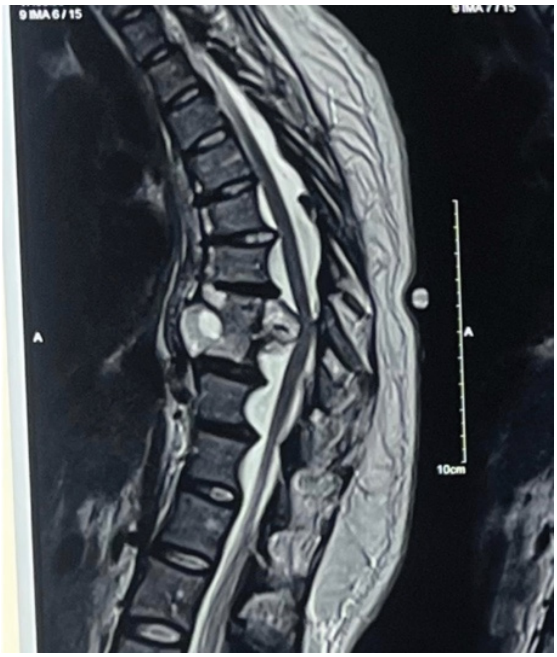

Figure 1: Plain MRI T-2 Image showing involvement of D10-D11 levels majorly

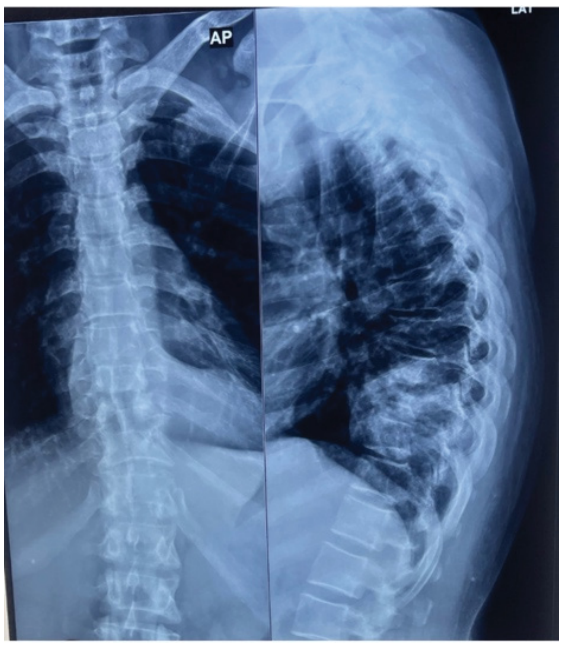

Figure 2: Plain Xray radiograph pf Dorsal spine in Anterio-posterior and Lateral view

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abscess. Pedicle screw fixation was performed from the sixth thoracic vertebra to the first lumbar vertebra. However, the right-sided pedicles were omitted from the eighth to the eleventh thoracic vertebrae, and the left-sided pedicles were omitted from the ninth to the eleventh thoracic vertebrae due to the presence of dystrophic pedicles.(Fig 6).

According to the reports, the patient was diagnosed with Multidrug-resistant Tuberculosis based on a positive gene xpert report. Following the operation, the patient was subsequently referred to a Direct Observed Treatment Schedule (DOTS) for further medical management of Tuberculosis.

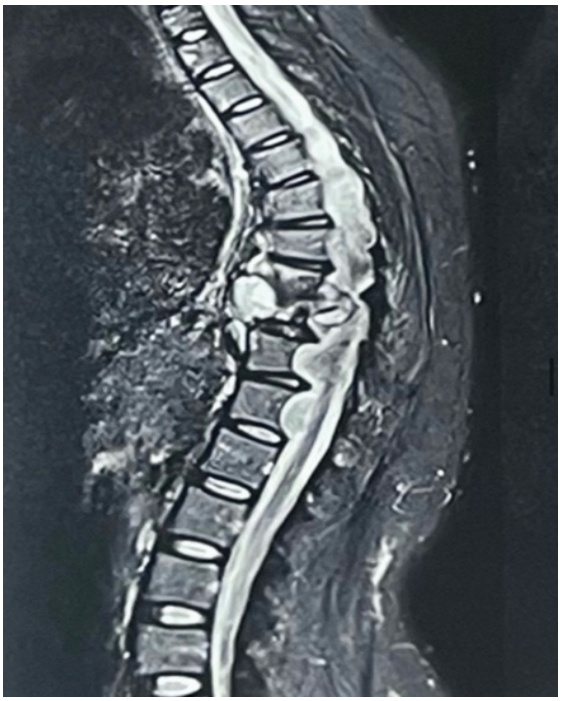

Figure 3: MRI D-L spine showing extension of the epidural abcess into above and below levels from the involved vertebrae

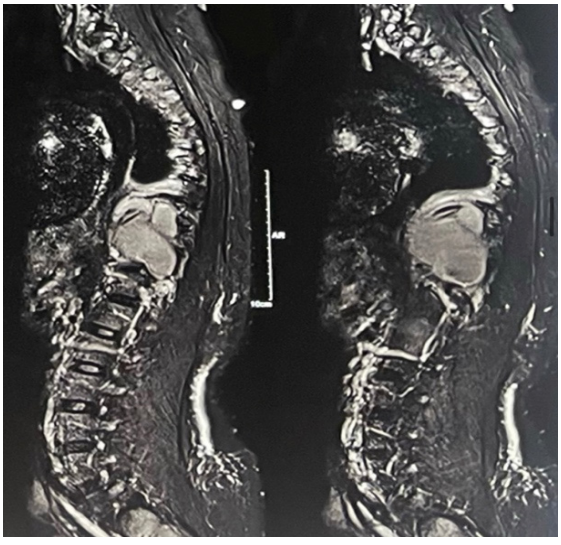

Figure 4: MRI films showing the paraspinal meningocele measuring 7*6.5*5.3 cm

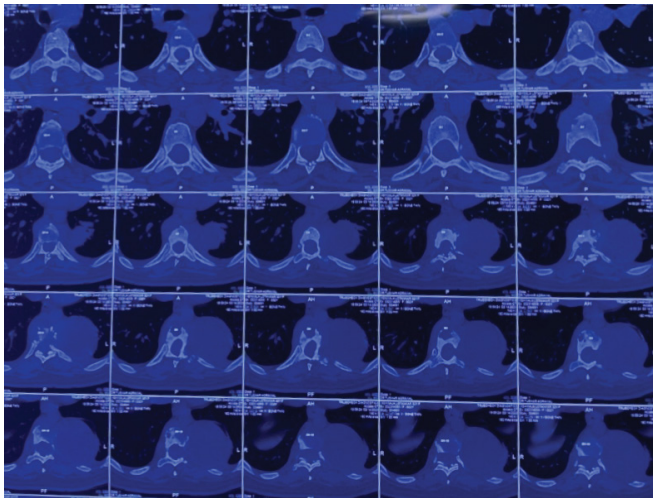

Figure 5: Axial Computed Tomography images showing dystrophic pedicles at multiple levels

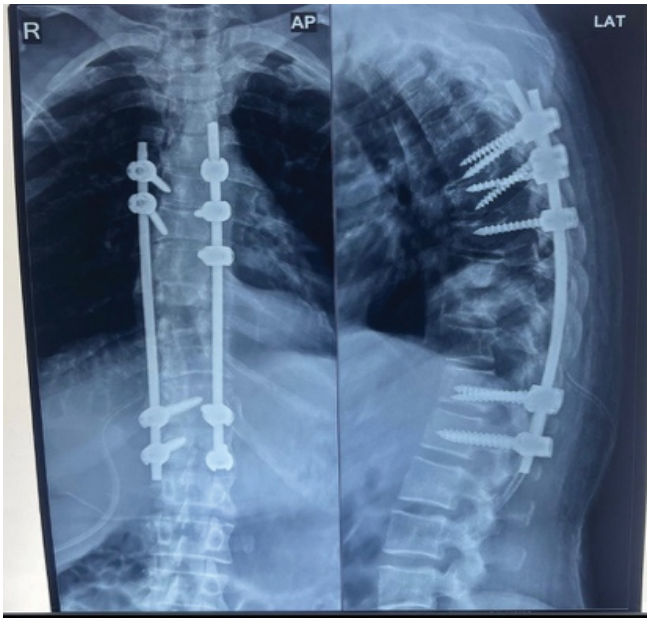

Figure 6: Post operative Xray showing planned pedicle screw instrumentation above and below the Involved levels to provide stabilisation

Discussion

We reported a rare case report of a young 32 year old female who is a known case of Neurofibromatosis Type 1 presenting with constitutional symptoms and paraparesis, managed surgically with decompression and stabilisation. In the literature review we have done there has been no reports of any cases of Potts spine seen in a patient with neurofibromatosis. So the endemicity of Tuberculosis in Indian subcontinent exposes us to rarest of the cases seen in the Tuberculosis spectrum. Southeast Asia is reported to

exhibit the second-highest incidence rate, with a prevalence of 226 cases per 100,000 individuals[5]. Even though surgical treatment is not always advised, literature shows good functional outcomes in patients treated surgically. Few studies also show the clinical features that were observed to be linked with inadequate functional recuperation which encompass indications of spinal cord compression, such as neurogenic bowel and bladder, muscle spasms, and weakness, as well as other aetiology's of cord compression, which included spinal deformity, thoracic spine involvement, vertebral collapse and abscess formation[6,7,8].

The precise prevalence of spinal deformities in individuals with NF1 remains uncertain, with estimates ranging from 2% to 69%[9]. However, scoliosis affecting the thoracic spine is the most frequently observed manifestation[9]. The occurrence of bone destruction and dystrophy is a frequent observation in individuals with NF1. The proposed pathophysiological mechanisms underlying this phenomenon include the localised infiltration or erosion of bone by neurofibromas, primary mesodermal dysplasia, osteomalacia, and endocrine disruptions[10].

Kyphosis leads to a greater degree of neurologic impairment compared to scoliosis due to the development of a pathological spinal flexion. This flexion results in excessive attenuation and deformation of the spinal cord parenchyma, ultimately giving rise to neurological symptoms[11]. Progressive kyphosis is a significant contributor to neurodeficit in cases of spinal tuberculosis also. Considering our case report the involvement of both NF1 with spinal tuberculosis puts our patient in a high risk of progressive neurodeficit and increased risk of paraplegia. The fundamental tenet of surgical intervention involves the execution of intervertebral debridement to achieve surgical decompression, which is subsequently followed by vertebral fixation. Two techniques for fixation, namely autologous bone grafts and titanium cage, have demonstrated remarkable functional outcomes in terms of JOA[12].

The process of segmental instrumentation may present difficulties at times due to the inadequate anchorage points for fixation caused by severely deformed vertebrae. Therefore, preoperative CT analysis is a crucial tool in the planning of a secure surgery with optimal hold of the pedicle screws. Excessive bleeding may be caused by the existence of paraspinal neurofibromas and plexiform venous channels in the soft tissues that envelop the spinal region which may also increase chances of postoperative hematoma formation[13].

To summarise, the condition of Neurofibromatosis presents significant obstacles throughout the process of diagnosis and development of treatment approaches. The presence of a secondary infection caused by Mycobacterium tuberculosis has presented a significant obstacle in devising a treatment regimen that is both safe and efficacious in achieving a successful recovery. The manifestation of progressive deformity is indicative of an imminent state of neurodeficit, which necessitates prompt and timely surgical intervention.

Conclusion

The present case report elucidates an infrequent manifestation of Potts spine with spinal cord compression in association with Neurofibromatosis Type 2. Timely identification and proper preoperative templating of the dystrophic pedicles are necessary in such instances to have a safely planned surgery. The patient underwent a diagnostic aspiration of prevertebral fluid collection and decompression and fixation surgery in order to prevent worsening of neurodeficit.

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None

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