

Tethered Cord Syndrome with Syrinx in A Nigerian Adult Female: A Case Report

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ABSTRACT

BACKGROUND Tethered cord syndrome is a diverse clinical entity secondary to any of a heterogeneous group of spinal disorders that typically occurs in children, but occurs as a rare pathologic entity in adults. The occurrence of tethered cord and spinal cord syrinx in an adult Nigerian is quite rare.

METHODS: The case record of the patient in review and literature search on the subject of tethered cord syndrome and spinal cord syrinx.

RESULTS: A 35 years female Nigerian, admitted into the department of internal medicine of the University of Port Harcourt teaching hospital with one month history of progressive weakness of the lower limbs associated with gradual loss of sensation in legs, urinary incontinence but no fecal incontinence or back pain. There was no history of trauma or chronic cough. Examination revealed reduced power in both lower limbs with loss of peripheral sensation, pain and light touch beginning at the level of L3, power in upper limbs was normal and the cranial nerves were intact. Urogenital and Musculoskeletal systems examination were normal. The respiratory and gastrointestinal systems were all normal. There were no cutaneous stigmata (hypertrichosis). Laboratory investigations showed normal hematological indices. Urinalysis was also normal. An impression of acute transverse myelitis was made. Magnetic resonance imaging showed tethered cord and syrinx in the lumbar region.

Conclusion: Tethered cord association with syringomyelia in adults is rare, although with the use of modern imaging tools like MRI, the diagnosis of TCS is no more as rare as it is thought and should be sought for in the appropriate clinical setting.

Keywords: Tethered cord; Syringomyelia; MRI; Adult; Nigeria.

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INTRODUCTION

Tethered cord syndrome is a diverse clinical entity characterized by symptoms and signs which are caused by excessive tension on the spinal cord due to impaired ascent of the spinal cord^{1,2}. This may be secondary to any of a heterogeneous group of disorder e.g spinal lipomas, lipomatous filum, and split cord malformations. It typically occurs in children, but it is a rare pathologic entity in adults^{1,2}.

The association of syrinx and tethered cord has been documented and there is a suggestion that syrinx formation can result from tethering leading to neurological deficit We report a case of a Nigerian female adult with tethered cord and syrinx presenting with neurological deterioration diagnosed by magnetic resonance imaging (MRI)

CASE REPORT

A 35 years old woman was admitted to the department of internal medicine of the University of Port Harcourt teaching hospital with a complaint of progressive weakness of the lower limbs of 1 month duration. There was associated gradual loss of sensation on both legs but no associated pain. There was associated urinary incontinence but there was no fecal incontinence or back pain. There was no history of trauma or chronic cough.

Physical examination revealed a young woman that was not pale or jaundiced. No peripheral lymphadenopathy or pedal oedema. Examination of the central nervous system revealed power of grade I on both lower limbs with loss of peripheral sensation beginning at the level of L3, sensory perception at lower limbs were also altered with reduced pin prick sensation. Power in upper limbs was grade 5. The cranial nerves were intact. Urogenital system showed wet perineum but normal vulva and vagina. Musculoskeletal system did not show scoliosis. The respiratory and gastrointestinal systems were all normal. There were no cutaneous stigmata (hypertrichosis). Laboratory investigations showed normal haematological indices. Urinalysis was also normal.

An impression of transverse myelitis was made to exclude tethered cord syndrome. Plain radiographs of the thoracic and lumbosacral spines were all normal. Magnetic resonance imaging of the lumbosacral spine demonstrated tethered cord and syrinx at lumbar region (See figs 1 and 2). Due to the unavailability of spinal neurosurgery in our centre, the patient was referred to the University college hospital Ibadan, but preferred travelling to the United Kingdom for further management.



Fig.1 T1W sagittal section MRI of the lumbosacral spine showing syring formation as hypointense tubular area and the distal spinal cord that ends at the level of L4 vertebra



Fig. 3 T2W sagittal section of lumbosacral MRI demonstrates syring formation in the spinal cord as hyper intense tubular area



Fig.2 T1W parasagittal section MRI of the lumbosacral spine showing syring formation in the lower thoracic spinal cord

DISCUSSION

Tethered cord syndrome (TCS) is a pull-induced functional disease of the spinal cord with its caudal part anchored by an inelastic structure³. Garcean first described the 'filum terminale syndrome' in 1953 in 3 patients⁴. In 1976, Hoffman and colleagues introduced the term 'tethered spinal cord' to describe the symptoms of their patients with elongated spinal cord and a thick filum terminate⁵. In 1981 Yamada⁶ and others widened the stretch induced functional disorder of the spinal cord to patients with other anomalies. Myelomeningocele, lipoma, lipomyelomeningocele, diastematomyelia, meningocele manque and dermoid sinus were included in this class only when parallelism between the oxidative metabolic changes and neurologic status exists. There was TCS was initially taught to be a paediatric problem but in some patients the diagnoses is not established until symptoms manifest in adulthood. This is the case with our patient who had associated syrinx. We do not know which preceded the other however; the increasing frequency in the diagnosis of TCS may well be due to better imaging and clinical recognition of this syndrome⁷.

TCS occurs in 0.05 to 0.25 of 1000 births⁸. It affects both sexes and all races and ethnic groups. Our patient is a Nigerian and of black race. Behavioral symptoms TCS manifest as chronic low back pains, progressive scoliosis, foot

deformities, numbness and loss of sensation on the legs or feet, awkward gait, weakness in legs or feet, unequal growth in the legs or feet, progressive loss of control of bladder and bowel functions (incontinence), and urinary tract infections. Our patient presented with weakness of the legs, loss of sensation and urinary incontinence.

Clinical signs of TCS include fatty tumors below the skin, hairy patches, spots of increased pigmentation, dimples that may indicate dermal sinus tracts and skin tags. None of these signs were seen in our patient.

The pathology involves an impairment of longitudinal movement of the spinal cord especially the conus medullaris which subsequently leads to chronic local ischaemia. In addition, the fillum terminate may be tight, thickened and sometimes shortened. An intradural lipoma, intradural scar formation or other lesions can also lead to conus fixation¹⁰.

Diagnosis is most readily achieved by MRI. Plain lumbosacral radiograph may show spinal bifida. MRI would delineate the localization of the conus, the thickness of filum terminalis (< 2mm is normal), posterior displacement of the conus and filum^{11,12}. MRI demonstrated conus terminalis that terminated distal to the level of L2/L3 intervertebral disc as well as syrinx formation in our patient.

Few cases of TCS coexisting with syrinx have been reported^{13,14} and findings suggested that the tethered cord preceded the syrinx formation.

Hsu et al¹³ reported a case of syringomyelia with associated aortic coarctation and tethered cord syndrome, where serial imaging showed significant reduction in thoracic syrinx after the repair of the coarctation and release of tethered of cord, indicating a possible cause effect relationship between syringomyelia and tethered cord.

Erkan et al¹⁴ reported thirty two cases of tethered cord with associated terminal synomyelia in the United States of America and noted that the frequency of sacral tethering was 40.6%, with the intramedullary paracentral position of the syrinx being 75%. Our patient had lumbar tethering with a central syrinx.

Erkan et al¹⁴ concluded that radiological significant terminal syringomyelia affects the clinical presentation of the tethered cord syndrome. Our patient declined?? surgery, so we could not monitor resolution of clinical symptoms and the syrinx. Management of TCS in adults may involve surgical detethering in those that are asymptomatic and decompression in those with associated syrinx¹⁴.

CONCLUSION

Tethered cord association with syringomyelia in adults is rare, although with the use of modern imaging tools like MRI, the diagnosis of TCS is no more as rare as was thought. There is a need to include TCS as an important differential diagnosis in patients that present with lesion or disease of the spinal cord and MRI is recommended as an important diagnostic work-up in these patients.

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