INTERCOSTAL NERVE ROOT SCHWANNOMA PRESENTING WITH PARAPLEGIA: A CASE REPORT OF AN UNCOMMON DIFFERENTIAL DIAGNOSIS

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ABSTRACT

INTRODUCTION: Schwannomas are tumours arising from the Schwann cells that usually form the myelin sheath on the peripheral nervous system. They arise commonly from the Obersteiner-Redlich zone of the posterior (sensory) nerve roots. They may lie entirely within the spinal canal or partly as a “dumbbell” mass passing through the intervertebral foramen and may cause paraplegia. They are known to be an unusual cause of paraplegia. Total excision with or without preserving the nerve results in a good outcome.

CASE PRESENTATION: The patient was a 23-year-old female referred to the Neurosurgery unit with upper back pain and progressive weakness of her lower limbs of spontaneous onset, and a clinical diagnosis of Pott’s disease. There was no previous history of trauma or history suggestive of malignancy. She was preserved with stable vital signs and had paraplegia with classic upper motor neuron lesion signs and sensory level at T3/T4. Screening for metastasis and tuberculosis were negative. Her magnetic resonance imaging revealed a “dumbbell” tumour at the third thoracic (T3) level, suggestive of a schwannoma, passing through the intervertebral foramen with significant spinal cord compression by its intracanalicular component. She had a T3 laminectomy and total excision of the tumour. Her postoperative period was uneventful, and she was discharged from the hospital three (3) weeks after the surgery with lower limbs power of MRC 4/5.

CONCLUSION: Tumours are an important cause of non-traumatic paraplegia. An intercostal nerve Schwannoma may be an important differential diagnosis that may mimic Pott’s disease despite its rarity. Total surgical excision is curative.

KEYWORDS: Paraplegia, Intercostal nerve, Schwannoma, Case report.
INTRODUCTION:
Schwannomas are tumours originating from the Schwann cells that usually form a myelin sheath on peripheral nervous system. They are believed to arise most commonly from a transition zone: The Obersteiner-Redlich zone. This zone represents a transition point along a neve where Schwann cells replace enveloping glial cells. They are slow-growing benign tumours that commonly arise from the posterior (sensory) nerve roots. They lie either entirely within the spinal canal or partly as a 'dumbbell' mass through the intervertebral foramen. Solitary spinal nerve Schwannoma is the most common nerve sheath tumour of the spine. They are most commonly located in cervical and lumbar regions because of the higher density of nerve roots at these sites. Paraplegia may result from thoracic or lumbar spinal cord compression. It has been found that the following may commonly compress the spinal cord: A bone, connective tissue that lines the spinal canal (ligaments), haematoma, tumour, abscess (commonly tuberculosis), and a herniated disc, among others. Spinal cord compressive myelopathies represent a majority of non-traumatic myelopathies in sub-Saharan Africa, and most are due to Pott’s disease. Schwannomas tend to occur in the 30–60 age group with a male-to-female ratio of 1:1. Patients may present with any of these two main symptoms of radiculopathy and neurogenic claudication; worsening sensorimotor loss, and back pain radiating from the tumour level. Typically, they may present with root pain and signs of cord compression may follow. Complete surgical excision is very feasible. Sometimes, the nerve root of origin may be inevitably sacrificed (excised with the tumour). However, the overlap from adjacent nerve roots usually minimises any neurological deficit. We report a rare case of an intercostal nerve schwannoma who presented with paraplegia which has rarely been reported.

CASE PRESENTATION:
The patient was a 23-year-old woman, referred to us with upper back pain and progressive weakness of her lower limbs for nine (9) and six (6) months, respectively, with a presumed clinical diagnosis of Pott’s disease. The pain was dull aching and band-like; more on the left, no known aggravating factor, not severe to affect her routines and relieved by taking over the counter analgesics. Difficulty in walking began with sensory changes and sensation of heaviness in the lower limbs, commencing on the left side. It ultimately led to her inability to walk five (5) weeks before the presentation. There was no urinary or faecal incontinence. There is no history suggestive of trauma, infection, malignancy, or comorbidities. She was found to be preserved with stable vital signs. Neurological examination revealed normal upper limbs. Lower limb findings were: average bulk, spastic, paraplegic (Frankel grade A) with upper thoracic sensory level corresponding to T3/T4 dermatome; deep tendon reflex was brisk with ankle clonus and Babinski sign. Her complete blood count, erythrocyte sedimentation rate (ESR) and Mantoux tests were unremarkable. Her thoracic spine magnetic resonance imaging (MRI) findings are as shown below in figures 1 and 2.
Figure 2: (A) An axial, non-contrast, T1 weighted image showing a left-sided “dumbbell”-shaped tumour with intra and extra canal components, the spinal cord pushed and compressed to the right, (B) a contrast study of an axial T1 weighted image showing an enhancing “dumbbell” tumour compressing the spinal cord.

With the clinical and radiological diagnosis of a nerve sheath tumour (Schwannoma), the patient was prepared for an urgent surgical excision of the tumour. Surgery was carried out under general anaesthesia, in the prone position. Tumour was accessed and totally excised after a T3 laminectomy. Tumour was found to be largely solid with a minor cystic area, well defined, extradural, and was easily shelled out in two (2) pieces sparing the nerve. Haemostasis was secured, no accidental dura breach, wound closed in layers and dressed. The postoperative period was uneventful, and she commenced physiotherapy on the third day. The suture was removed on the 10th day, and she made a good recovery. She was discharged home on the 3rd-week post-op with Frankel grade D functionality, which improved to grade E at first follow-up (2nd week after discharge). She was lost to subsequent follow-ups, and therefore postoperative MRI is not available.

The tumour was examined histopathologically and confirmed to be a benign (WHO grade I) schwannoma as shown in figure 3 below.

Figure 3: (A) photomicrograph of the Schwannoma exhibiting foci of hyper cellular and hypo cellular areas (Antoni A and Antoni B), H and E X100. (B) Higher magnification showing varocay body, H and E X200.
DISCUSSION:
Schwannoma has the highest incidence among Nigerians, as reported by Adeolu et al. The index case was a 23-year-old female. The typical age affected is third to sixth decades with equal sex predilection. Her referring clinical diagnosis was paraplegia from Pott's disease. Govind et al. had reported that most cases of Schwannoma in the intercostal site are misdiagnosed on the clinical findings because it can mimic other lesions like spondylopathy and disk herniation. The index patient presented with truncal pains and progressive limbs weakness as red flag signs. These findings are in keeping with reported cases by Selli, Bruneour, and Chikani. In their reports, the patient's presentation was non-specific. However, they noted that pain and motor dysfunction are constant symptoms of spinal cord tumours. The duration of our patient's symptom was nine months. Adeolu had reported that most patients' symptoms' duration exceeds six months. Similarly, the classic clinical presentation of the index case was in concordance with the findings of Kottbuer. However, Kottbuer has additionally found sphincter dysfunction among some of his patients with paraplegia. His findings may be due to the fact that some of his patients had intramedullary tumours. In contrast to his, our patient had no sphincter dysfunction probably because her tumour was entirely extradural in location.

The treatment of choice for intercostal Schwannoma with paraplegia is urgent surgical resection as suggested by Kim and Karikari. She underwent laminectomy in the prone position in order to access the posterior located tumour. It has been found that through laminectomy, almost all posterolaterally located tumours are easily accessed as found by Zong and Lenzi. We found a mixed consistency tumour similar to most findings of Lee.

CONCLUSION:
Spinal cord compression from intercostal nerve schwannomas though not expected, is an important differential diagnosis to consider in patients with paraplegia. Total excision is achievable with an excellent clinical outcome.

REFERENCES:


