A Case Report of Malignant Peripheral Nerve Sheath Tumour (MPNST) Which Present as an Acute Traumatic Sciatic Neuropathy

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Abstract Malignant peripheral nerve sheath tumour (MPNST) is a very rare sarcoma and it accounts for around 5% of all sarcomas. We are reporting an interesting case of sporadic MPNST at the proximal sciatic nerve which concealed as a traumatic cause of sciatic neuropathy and foot drop. A 59 year old Caucasian lady presented with a constant paraesthesia and pain at her left hip after she had a fall. On examination, there was marked neurological impairment along the L4, L5 and S1 distribution. A MRI of the pelvis was performed. It revealed a large soft tissue mass at the centre of the left buttock arising from the proximal sciatica nerve. The biopsy result revealed the diagnosis of malignant peripheral nerve sheath tumour (MPNST). She underwent a complete resection of tumour with clear margins of the tumour. A vigilance MRI examination was performed 5 months post-operatively and no evidence suggesting of local recurrences. However, 10 months after the operation a surveillance CT scan was performed and it revealed presence of pulmonary metastases. MPNST is a heterogenous group of tumour arising from the Schwann cells or the perineural cells. The presentation is usually subtle and invariably leads to a late diagnosis of the disease. The prognosis is often poor. The mainstay of treatment is surgical resection. However, despite complete resection, the tumour can recur and metastasis. A continuous paraesthesia and pain along nerve root must raise the suspicion among clinicians about the possibility of extraspinal causes of sciatica.

Keywords MPNST; Malignant peripheral nerve sheath tumour; Sciatica; Foot drop

Introduction

Malignant Peripheral Nerve Sheath Tumour (MPNST) is an extremely rare medical condition (Karr et al., 2006; Nthumba and Juma, 2011). It is often underappreciated as a cause of peripheral nerve compression, especially the involvement of sciatic nerve where spinal pathology and traumatic sciatic neuropathy supersede the diagnosis. In the current published literatures, the first presentation of MPNST is usually a mass which follows nerve symptoms such as pain, paraesthesia and limb weakness (Nthumba and Juma, 2011). However, we are reporting an interesting case of MPNST at the proximal sciatic nerve which concealed as a traumatic cause of sciatic neuropathy and foot drop, where a subtle mass was only detected later.

1 Case

A 59 year old lady who was otherwise fit and healthy presented with a 15 months history of left leg weakness and pain. Initially she had a fall which landed herself on her left hip while walking in the snow. Ever since then, she has been complaining of a constant dull pain around her left buttock. Hip trauma was initially suspected and a hip x-ray was performed. However, it did not show any bony fractures. The pain gradually improved and it was diagnosed as an acute traumatic neuropathic pain secondary to the fall.

However, she fell on her left hip again 15 months later. Subsequently she started having severe pain in the left buttock which radiated to the lower half of her left leg. Hip x-ray was performed again and did not show any fractures. She was therefore referred to the neurosurgical
department with the diagnosis of possible lumbar disc herniation. On examination, she presented with a very obvious left foot drop. Straight leg raising test was 40 degrees for the left leg. There was marked weakness along the L5 and S1 distribution with significant sensory impairment as well as decreased ankle reflex. There was a slight weakness in the L4 muscle innervation as well. However, further MRI scan of the spine did not show any herniation of the disc or external compression of the nerve.

Convinced that it was a sciatica nerve compression, a subsequent MRI of the pelvis was performed. It revealed a large soft tissue mass at the centre of the left buttock which arise from the sciatica nerve at the level of the great sciatic notch. It measured 10.8 cm (supero-inferiorly) × 9.6 cm (transversely) × 7.8 cm (antero-posteriorly). There was no evidence of local invasion into the surrounding tissues. Staging CT scan did not reveal any metastasis of the tumour. Genetic testing was also performed and she did not carry Neurofibromatosis 1 gene.

The biopsy result revealed malignant peripheral nerve sheath tumour (MPNST). The patient underwent a complete resection of tumour with clear margins of the tumour. She was recovering well from the operation without any complications and was commenced on chemotherapy. A vigilance MRI examination was performed 5 months post-operatively with no evidence suggesting of local recurrences. However, patient started being breathless and lethargic. A surveillance CT scan was performed which revealed presence of multiple well defined pulmonary metastasis and a 14 mm subcutaneous nodule in the right anterior abdominal wall.

2 Discussion
Malignant Peripheral Nerve Sheath Tumour (MPNST) is an extremely rare medical condition with an incidence of 1 in every 100,000 population (Karr et al., 2006). It is a heterogenous group of tumour arising from the Schwann cells or the perineural cells (Omezzine et al., 2009). It accounts for 3%–10% of all soft tissue sarcoma (Katz et al., 2009). Almost half of the MPNST are associated with Type 1 Neurofibromatosis or von-Recklinghausen’s disease, an autosomal dominant disease which is associated with inactivation of NF1 gene (Omezzine et al., 2009; Katz et al., 2009; Dimou et al., 2009). It can also be associated with post-radiation. The rest of the incidences are sporadic causes. It is more common between the age group of 20 to 50 years old, with a predisposition to the 4th decade of life (Lin et al., 2007). In patients with neurofibromatosis, the disease will manifest earlier by at least one decade (Geller and Gebhardt, 2006). The mean age for the presentation of the disease in sporadic cases is 39.7 as compared to those with neurofibromatosis with mean age of 28.7 (Karr et al., 2006; Nthumba and Juma, 2011). MPNST can also occur in children with NF-1 mutation (Dimou et al., 2009; Geller and Gebhardt, 2006).

The common site of presentation is usually at the large nerve trunk in the peroneal nerve, sciatica nerve, radial plexus and sacral plexus (Nthumba and Juma, 2011; Geller and Gebhardt, 2006). In patients with neurofibromatosis, it usually develops from deep seated plexiform neurofibroma. It presents with symptoms of nerve compression such as paraesthesia, radicular pain and weakness (Nthumba and Juma, 2011; Rodero et al., 2004). Hence, clinically it is very difficult to differentiate sciatica caused by spinal pathology from MPNST at the proximal sciatic nerve. However, our case report revealed that the patient has been suffering from sciatica for 15 months without complaining of a mass until a later stage. Given the history of trauma accompanied by proximal sciatica and foot drop, it is not uncommon to misdiagnose it as traumatic neuropathy or spinal cause of sciatica.

It has a poor prognosis with a 5 year survival rate stands between 20%–50% (Katz et al., 2009). It is biologically aggressive and has a very high propensity to metastasise. The local recurrence rate and metastatic rate after a complete surgical excision ranges from 40% to 65% and 40% to 68% respectively (Geller and Gebhardt, 2006). MRI is the preferred radiographic method as part of the integral management of MPNST (Wasa et al., 2010). It is very useful in differentiating between benign and malignant peripheral nerve sheath tumour, especially deep seated sarcoma where dissemination
of the tumour in the visceral organs or nerve palsy could occur if biopsies were attempted (Wasa et al., 2010). Although there are certain features on MRI which could be useful in differentiating MPNST from its benign counterpart such as schwannoma or neurofibroma, they are often unreliable (Geller and Gebhardt, 2006). Radiological Gross histogramology and immunostaining is still the mainstay of diagnosis. It is also essential prior to the surgery (Geller and Gebhardt, 2006).

The mainstay of treatment is surgical resection (Katz et al., 2009). There are two surgical approaches; limb sparing surgery (en bloc or radical resection) or limb amputation (Nthumba and Juma, 2011). Limb sparing surgery is favoured over limb amputation. The main aim is to achieve clear margins of resection and to prevent systemic spread with adjuvant chemotherapy (Lin et al., 2007). However, residual prognosis of the limb can be a devastating consequence of limb sparing surgery (Dorsi et al., 2011). Reconstruction of the nerves is not recommended as it does not restore the full functionality of the limb (Dorsi et al., 2011). In some cases, amputation of the limb is inevitable if the tumour has progressed around the vital structures (Dorsi et al., 2011).

Chemotherapy has low sensitivity to the sarcoma. Radiotherapy on another hand has shown to control local recurrences (Karr et al., 2006; Dimou et al., 2009). However, it has no effect on long term survival. It was suggested that radiotherapy should commence as early as possible for intermediate to high grade tumour or low grade tumour after marginal excision (Lin et al., 2007).

3 Conclusion

Extraspinal causes of sciatica alone are extremely rare and it is often underappreciated even though in some cases the disease might prove fatal to the patient. The patient in the case study has a delayed diagnosis for 15 months. Even though surgery has achieved a complete resection with clear margins of resection, the patient developed a secondary metastasis in the lung months after the surgery despite her receiving ongoing chemotherapy. The clinical diagnosis of sciatic MPNST remains difficult especially if the clinical presentation suggest of other common diseases. An extraspinal cause should take into considering in the management of sciatica pain and footdrop. A late detection of malignant tumour could prove to have poor prognosis.

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