There were numerous references to spinal cord ependymomas,[9,10] but none causing what amounts to distant intracranial false-localising signs consequent to oedema. This case highlights the need to look further afield when presented with the scenario of clinical features of a brainstem lesion with only oedema apparent on cranial imaging. It indicates the need to include cervical imaging well below the foramen magnum in these circumstances.

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Multiple schwannomas of the sciatic nerve☆

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Summary Schwannomas are rare benign tumours of nerve sheath cells of neural crest origin. Often these tumours are solitary and encapsulated. Multiple schwannomas can arise from the peripheral nervous system including cranial nerves, spinal roots, the brachial and lumbar–sacral plexus or major peripheral nerves. We report an extremely rare case of schwannomatosis of the sciatic nerve in a young female and include a comprehensive literature review. Treatment options are discussed.

Journal of Clinical Neuroscience (2003) **10**(3), 391–393

doi:10.1016/S0967-5868(03)00021-3

Keywords: multiple schwannomas, schwannomatosis, sciatic nerve

Received 26 October 2002
Accepted 26 November 2002

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☆There were no sources of support or grants of any kind used in the production of this case report and review.

INTRODUCTION

Single schwannoma, a rare benign tumour of nerve sheath cells of neural crest origin, is the most common of all peripheral tumours.[1] Multiple schwannomas of the peripheral and central nervous system are, however, uncommon. Multiple schwannomas may arise from the cranial nerves, spinal roots, the brachial and lumbar-sacral plexus or major peripheral nerves. These tumours are often solitary and encapsulated. The treatment for solitary schwannoma is to enucleate from the nerve of origin and this can be performed with minimal loss of function in that nerve.[1] We report a very rare case of schwannomatosis found along the right sciatic nerve with no associated neurofibromatosis.

CASE REPORT

An 18-year-old female asthmatic ballet dancer presented with right leg pain. There was no preceding history of trauma. The pain started in the right buttock area with radiation down the back of her leg to the base of her foot, progressive over an 18-month period. She had weakness of her thigh extensors and ankle plantar flexion. Multiple opinions were sought. CT and MR scans of her lumbo-sacral spine were normal. Head CT and MR scans were normal. The patient had never noticed any subcutaneous lumps of nodules. There was no history of tinnitus, hearing loss or vestibular problems. There were no *café-au-lait* spots and ophthalmic examination revealed no visible evidence of cataracts. There was no family history of schwannomatosis or NF2 (neurofibromatosis Type 2, formerly "central neurofibromatosis").

Two weeks prior to presentation there was worsening of her leg pain and she developed weakness of ankle dorsiflexion at 4-/5. Nerve conduction studies showed a mono-neuropathy of the sciatic nerve with slowed conduction along the entire length of the sciatic nerve. MR scans of the gluteal and thigh region demonstrated a generalised increase in diameter of the sciatic nerve with multiple lesions on her sciatic nerve (Figs. 1 and 2). There was loss of right gluteal muscle mass. Her superior and inferior gluteal nerves were not affected with tumour, but there was presumably compression neuropathy of these nerves from the grossly enlarged sciatic nerve.

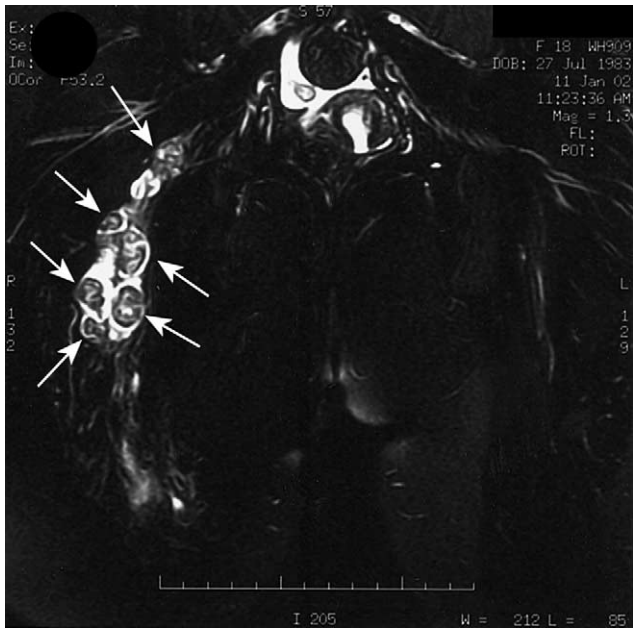

Fig. 1 T2WI coronal MRI of the right sciatic nerve: note the multiple lesions.

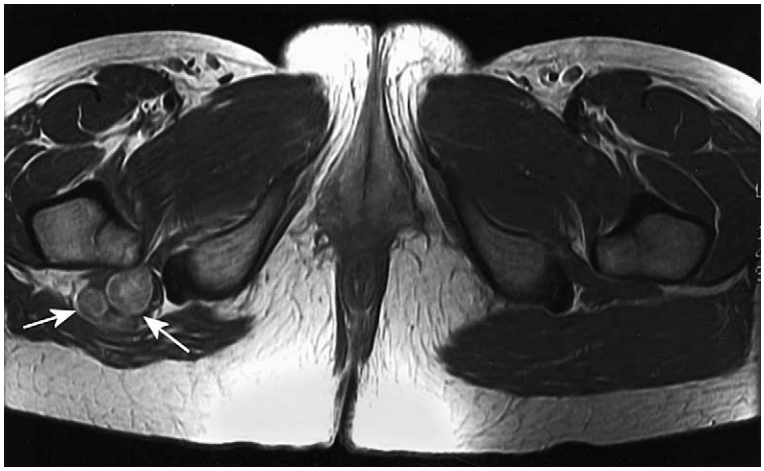

Fig. 2 T1WI axial MRI of the right sciatic nerve: the lesions appear to be well encapsulated – consistent with schwanommatosis rather than neurofibromatosis.

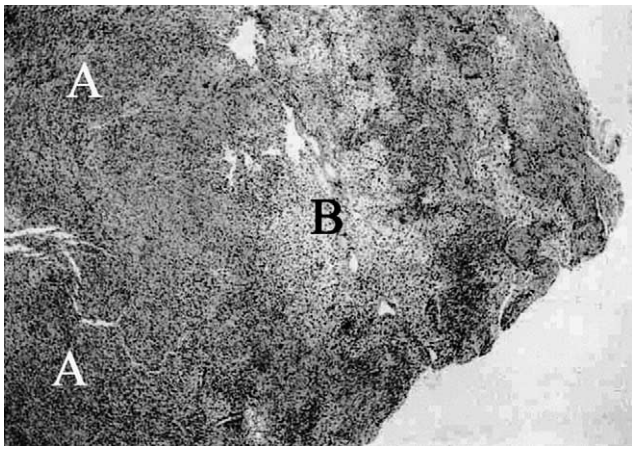

Fig. 3 Microscopic appearance (H&E stain, 10× magnification): note the regions of Antoni A (A) and Antoni B (B) consistent with the diagnosis of schwanomma.

Under general anaesthesia in the prone position, a standard sciatic nerve exposure was performed. On inspection, there were multiple firm nodules along the sciatic nerve. The largest lesion was identified in the limits of our exposure. As the lesions were well encapsulated, it was decided intra-operatively to enucleate the largest lesion for histopathological diagnosis. Histopathological study revealed the classic appearance of schwannoma (Fig. 3).

The post-operative course was unremarkable with hospital discharge on day 2. The patient reported a moderate improvement in pain, however little improvement in motor function. Multiple factors contributed to the patient's decision to receive conservative follow-up treatment with an exercise regime and pain management medication. Factors included: (1) possible further neurological deficits from radical enucleating of schwannoma along sciatic nerve, (2) the patient's belief that she will improve and (3) the patient's belief that she is able to control pain with pain medication.

DISCUSSION

"Neurilemmomatosis" was introduced by Shishiba et al.[2] to describe a distinct clinicopathological disease of multiple schwannomas without manifestations of neurofibromatosis of von Recklinghausen's disease. They described multiple schwannomas of the cutaneous (intradermal) type. Several articles subsequently reported examples of multiple cutaneous schwannomas.[3,4] Many of these early reports included patients with acoustic neuromas in addition to cutaneous schwannomas.[2,4] One of the 4 cases in the series by Shishiba et al. and 1 of 2 cases reported by Purcell was associated with bilateral acoustic neuromas. Cases associated with the bilateral acoustic neuromas, which have previously been reported to be schwannomatosis, would be classified as NF-2 after the NIH Conference statement.[5] The current diagnostic criteria for NF-2 includes diagnosis of bilateral eighth nerve tumours.

Several other reports of schwannomatosis without manifestations of NF-1 or NF-2 have been described.[6,7] We describe a patient with schwannomatosis localised to her right sciatic nerve only with no other stigmata of NF1 or NF2. She presented with 18 months history of leg pain. Prior to the leg pain, she was on high doses of steroids for her asthma. Steroids have some anti-inflammatory effects, which may have masked her leg pain. Her pain appeared after she decided to cease the steroids for her asthma.

Schwannomatosis was previously thought to be a distinct, non-hereditary condition. However, MacCollin et al.[8] suggested that schwannomatosis might be due to segmental mutation of the NF2 gene or other schwannoma-related genes. Evans et al. showed that linkage analysis in families with schwannomatosis was consistent with involvement of the NF2 gene. Honda et al.[9] found germ-line mutation in patients who presented with schwannomatosis who subsequently developed others signs of NF2.

Single schwannoma is a rare benign tumour of nerve sheath cells, but it is the most common of all peripheral tumours. Multiple localised schwannomas confined to a deep, major nerve in a single extremity is rare. Lewis et al. described a patient with 12 tumours along the median and ulnar nerves.[10] Shank et al.[11] presented a case with 4 to 6 schwannomas in the right ulnar nerve. Ogose et al.[12] presented a case series with 4 patients all with multiple schwannomas arising from peripheral nerves in a single extremity. MacCollin also presented a series with 3 patients having multiple tumours limited to a single limb.[6] Most patients in this series presented with pain. Pain was relieved post-surgical removal of tumour.

Schwannoma is an encapsulated, slow growing nerve sheath tumour. Neurofibroma does not possess a true capsule.[1] Schwannoma is the most common of all peripheral nerve tumours. On clinical exam, it is often mobile side to side, but fixed along the nerve: painful paraesthesia in the dermatome of the nerve of origin, similar to Tinel's sign, may be present.[1] It was also noted that patients without neurofibromatosis almost always present with a

solitary lesion.[13] Patients with schwannoma in their lower limb peripheral nerve(s) may present with plantar foot pain. The foot pain caused by peripheral nerve schwannoma can be wrongly diagnosed as tarsal tunnel syndrome.[14,15]

Distinction between schwannomas and neurofibromas can be made histologically and radiologically. Histological features of schwannoma may include areas of compact bundles of Schwann cells (Antoni type A) or loose matrix of oval cells (Antoni type B) (Fig. 3). Antoni A areas show greater cellularity in schwannomas compared to neurofibromas. S-100 immunostaining is particularly prominent and uniform in cellular areas of the schwannomas, whereas neurofibromas tend to be variable in staining of cells for the S-100 protein.[13] This characteristic is also useful when differentiating schwannomas from fibrosarcoma and leiomyosarcoma. T2-weighted MRI may show peripheral hyperintense rim with central low intensity. This is the "target pattern" which is characteristic of schwannoma on contrast-enhanced T1-weighted and T2-weighted images.

The principle of schwannoma surgery is simple enucleation of tumour without damaging the nerve.[1,16] Partial excision of the tumour may be indicated for an infiltrating tumour. The question arises when there are schwannomas at multiple sites along the nerve. Yamamoto et al.[17] suggested conservative follow-up treatment for their patient as she had minimal symptoms. Our patient is different and poses a challenging management problem. The initial operation was for histological diagnosis and relief of pain symptoms. A multi-disciplinary approach was taken which included consultation with the radiation oncologist and the pain management specialist. We planned for a conservative follow-up regime. If there is worsening pain or hemiparesis, further enucleation of tumour will be considered. Further imaging studies would be repeated every 3 to 5 years as it is still possible for the patient to develop bilateral vestibular schwannomas. We believe it is unlikely for the patient to have NF2, but we would not completely exclude the diagnosis.

Many papers support and disagree with the concept that schwannomatosis is a distinct clinical entity. The pathogenesis of schwannomatosis is still unclear. It is unclear whether all cases of multiple schwannomas are variants of NF2 or a distinct clinicopathological entity. It is hoped that with long-term follow-up and further genetic analysis, we will come to understand the pathological basis for schwannomatosis. At present in Australia, genetic testing for NF2 is a second line investigation and only performed in individuals after confirmation of a clinical diagnosis of NF2 rather than being a diagnostic investigation.

CONCLUSION

We present a rare case of schwannomatosis partially involving the right sciatic nerve of an 18-year-old girl. The extent of the involvement of the schwannoma nodules was confined to that single nerve. We believe that schwannomatosis without familial history, imaging or clinical evidence of NF1 or NF2 may represent a distinct clinicopathological entity.

ACKNOWLEDGEMENTS

Thanks to Medical Illustration Unit at The Prince of Wales Hospital for preparation of diagrams and to Catherine Hyam for her assistance in all administrative work.

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