There were numerous references to spinal cord ependymomas, 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.

REFERENCES


Multiple schwannomas of the sciatic nerve

J. Huang BS(MED), R. Mobbs BS(MED) MB BS, C. Teo MB BS MD FRACS

Center for Minimally Invasive Surgery, The Prince of Wales Private Hospital, Division of Neurosurgery, NSW, Australia

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 lumbo-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.

© 2003 Elsevier Science Ltd. All rights reserved.


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

Keywords: multiple schwannomas, schwannomatosis, sciatic nerve

Received 26 October 2002
Accepted 26 November 2002

Correspondence to: C. Teo, Suite 3, Level 7, POWP Hospital, Barker Street, Randwick, 2031, Australia.

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

© 2003 Elsevier Science Ltd. All rights reserved.

Fig. 1 T2WI coronal MRI of the right sciatic nerve: note the multiple lesions.
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. 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. Many of these early reports included patients with acoustic neuromas in addition to cutaneous schwannomas. 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. 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. 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. 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. found germ-line mutation in patients who presented with schwannomatosis who subsequently developed other 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. Shank et al. presented a case with 4 to 6 schwannomas in the right ulnar nerve. Ogose et al. 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. 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. 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. It was also noted that patients without neurofibromatosis almost always present with a
solitary lesion.\textsuperscript{13} Patients with schwannoma in their lower limb peripheral nerve(s) may present with plantar foot pain. The foot pain caused by peripheral nerve schwanna can be wrongly diagnosed as tarsal tunnel syndrome.\textsuperscript{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.\textsuperscript{16} 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 schwanna surgery is simple enucleation of tumour without damaging the nerve.\textsuperscript{1,6} 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.\textsuperscript{17} suggested conservative follow-up arises when there are schwannomas at multiple sites along the tumour maybe indicated for an infiltrating tumour. The question 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.

**REFERENCES**