**Operative approach to spinal schwannomatosis**

***Introduction***

Schwannomas are benign tumors of Schwann cell origin and are the most common tumor of peripheral nerves, and posterior fossa masses. They are slowly growing, encapsulated peripheral nerve sheath tumors, which arise eccentrically from their parent nerve, with the nerve fibers splayed along their surface (as distinct to [neurofibromas](https://radiopaedia.org/articles/neurofibroma?lang=us) which arise within the nerve). Most schwannomas are solitary (90%) and sporadic. However, the presence of multiple schwannomas in a single patient suggests a genetic predisposition to tumorigenesis and possibly an association with one of the Neurofibromatosis (NF) syndromes. Schwannomatosis and neurofibromatosis type 2 (NF2) are both characterized by the development of multiple schwannomas but represent different genetic entities. Whereas NF2 is caused by mutations of the *NF2* gene, schwannomatosis is associated with germline mutations of *SMARCB1* or *LZTR1*.

We present a case of multiple schwannomas of the spine related with schwannomatosis, and describe the radiologic, clinical and intraoperative findings that clarify the trait of schwannomatosis.

***Aim***

Patients with severe neuropathic pain from multiple schwannomas, should undergo systemic review and brain imaging for differential diagnosis of NF type 2. Unlike NF type 2, schwannomatosis shows favourable outcomes, and differential diagnosis with appropriate imaging study is absolutely needed. Gross total resection (GTR) is usually curative for patients with sporadic tumors. For patients with NF2, there is a high incidence of new tumor formation.  
The assessment and diagnosis of spinal schwannomatosis always begins with eliciting a thorough history and physical examination. The aim is to emphasize on precise physical examination that may help create a differential diagnosis and subsequent final diagnosis with the aid of imaging diagnostic modalities.

***Clinical presentation/ Materials and methods***

A case of 66-year old woman admitted with severe left leg and axial back pain (VAS 8). On physical examination she had left L5 and S1 radiculopathy and paraesthesia, peroneal and tibial paresis 4/5 and weakened Achilles tendon reflex. Bowel and bladder dysfunction were excluded.

MRI showed four enhancing intradural-extramedullary masses from T12 to L3 level, biggest of which is at level L2/L3 occupying the whole diameter of the vertebral canal.

Patient was operated in prone position. Central laminectomies from Th12 to L3 were performed followed by microsurgical GTR of lesions. All 4 masses originated from a single nerve root – L1. Because of their infiltrating manner, L1 nerve root rhizotomy also performed. Facet joints were preserved and stabilization was not required.

***Results***

Improvement of patient’s overall condition and neurological status was achieved without any motor deficit.

The complaints and neurological deficits at the time of admission resolved postoperatively. There were no unexpected complications.  
  
***Conclusion***

Schwannomas are benign encapsulated neoplasms of Schwann cells.

Whether or not to perform GTR and sacrifice the functionally important root remains a critical decision for surgeons. In our patient after sacrificing L1 nerve root there were no additional neurologic deficit. We ground our decision on literature review, patient’s clinic and imaging diagnostic.

Similar approach was carried out by other surgeons in the past. Kim et al. reported 31 cases that involved the functionally important root (C5-T1 or L3-S1) in which GTR was achieved by sacrificing the root. They reported a neurologic deficit rate of 23%, but that the observed deficit was not functionally debilitating in any of the cases.

**Operative approach to spinal schwannomatosis**

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We present a case of multiple schwannomas of the spine relevant with schwannomatosis, and describe the radiologic, clinical and intraoperative findings that clarify the trait of schwannomatosis.

***Aim***

Patients with severe neuropathic pain from multiple schwannomas, should undergo systemic review and brain imaging for differentiate schwannomatosis from NF type 2.   
The aim is to emphasize on precise physical examination that may help create a differential diagnosis and subsequent final diagnosis with the aid of imaging diagnostic modalities, which on the other hand helps on pre- and intraoperative decision-making.

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MRI showed four enhancing intradural-extramedullary masses from T12 to L3 level, originating from a single nerve root (L1).

GTR of all 4 masses was achieved. Because of their infiltrating manner, L1 nerve root rhizotomy also performed. Facet joints were preserved and stabilization was not required.

***Results***

Improvement of patient’s overall condition and neurological status was achieved without any motor deficit.

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Schwannomas are benign encapsulated neoplasms of Schwann cells.

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