Intramedullary Schwannoma

James Watson Kernohan has been recognized as the first neurosurgeon to report an intramedullary schwannoma case in 1952, though Wilder Penfield had already described an intramedullary lesion with schwannomas characteristics in 1932 [1].

Epidemiology

Intramedullary (IM) schwannomas are rare entities representing the 0.3-1% of intramedullary tumors and 1.1% of spinal schwannomas. Beside many theories proposed, their rare occurrence might be related to the absence of Schwann cells into the spinal cord.

To date, approximately around 60 cases of intramedullary schwannoma in patients without neurofibromatosis have been reported in the English literature, of which only eight were children [2].

The male:female ratio for intramedullory schwannomas is 3:1 with a mean age of 40 years old. They are usually single lesions affecting the cervical spinal cord (63%), the thoracic spinal cord (26%), and the lumber spinal cord (11%). They have a slow growth pattern and because of this, the average interval between first symptom and diagnosis is 28.2 months (from 6 months through 20 years) [3].

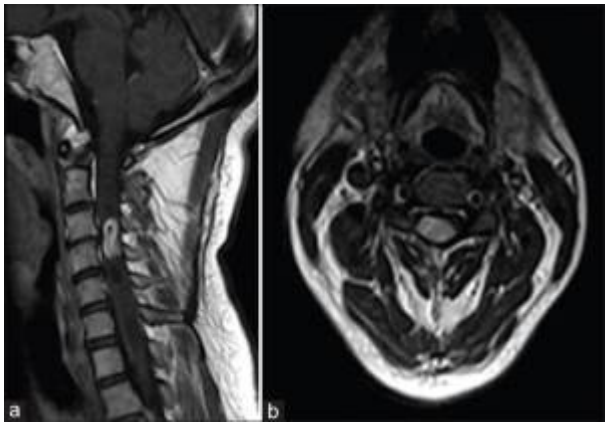
see Thoracic intramedullary schwannoma.

Clinical features

The most described clinical manifestation is the pyramidal syndrome followed by sensitivity complaints and sphincter dysfunction. There are reports of muscular fasciculations as the first symptom [4].

Diagnosis

A tumor located in the dorsolateral spinal cord, causing expansion of the cord, with hypointense areas on T2-weighted and obviously enhancement, should arise suspicion of an intramedullary schwannoma [5].



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Outcome

Intramedullary schwannoma may recur after resection [6].

Case series

The mean age of the total 11 patients at the operation was 50.2 years. The mean duration of the symptoms was 23 months, with limb paresthesia being the most common clinical presentation. The cervical spine was the most common localization level of the tumor in 6 cases. The mean follow-up duration was 49.4 months. Gross total resection (GTR) and subtotal resection (STR) was achieved in 9 and 2 cases, respectively. According to the modified McCormick scale at 6 months postoperatively, 7 cases (63.6%) had improved and 4 cases (36.3%) had unchanged grades. Typical MRI findings of the intramedullary schwannoma included ring-like enhancement, syringomyelia, cystic formation, intramedullary edema, and hemosiderin deposition. Gadolinium enhancement was homogenous in 8 cases (72.7%). The tumor margins were well-demarcated in all cases.

Intramedullary schwannoma should be considered when sharp margins and well-enhanced tumors are present at the cervical spine level and the initial symptoms are relatively mild, such as dysesthesia. When GTR cannot be achieved, STR for tumor decompression is recommended [7].

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8 patients with pathologically confirmed intramedullary schwannomas were reviewed.

There were 6 male and 2 female patients (mean age, 49 years). Tumors were located in the cervical cord (2), the thoracic cord (5), and the cervicothoracic cord (1). Most were in the dorsolateral spinal cord and limited to one side. Expansion of the cord was observed. The majority were hypointense on T1-weighted images and hypo-hyperintense on T2-weighted images. Peritumoral edema was easily found. Gadolinium enhancement was obviously. No recurrence was seen during the follow-up period.

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Case reports

A 52-year-old gentleman presented with a history of gait instability and numbness in bilateral lower limbs. He had clinical signs of myelopathy. His magnetic resonance imaging (MRI) dorsal spine was done that showed an intradural IM lesion at the level of D11, with one differential of ependymoma. Near total resection of lesion was done and histopathology reported it schwannoma.

Preoperative radiologic assessment for IM spinal lesions is difficult and high degree of suspicion should be present when approaching a patient with somatic pain and IM lesion on MRI, keeping in mind one differential of IM schwannoma [9)]

Dhake and Chatterjee described two cases of thoracic intramedullary schwannomas that recurred after primary excision. A 10 years old boy presented with weakness of both lower limbs. Magnetic resonance imaging showed a D10 to D12 intramedullary lesion, which was excised near totally and confirmed to be a schwannoma on histopathological examination. The tumour recurred twice after that and was re operated both times. Another 57 years old lady presented with weakness of both lower limbs and a history of being operated in the past for D9-D10 intramedullary lesion. She was re-operated with total removal of the lesion confirmed to be schwannoma on histopathological examination. Intramedullary schwannoma may recur after resection. [10)].

A 8 year-old female affected by a progressive paraparesis. Neuroradiological investigations showed an oval-shaped mass at the level of T10-T11. The patient underwent surgery, performed under neurophysiological monitoring. The patient was operated on with a complete removal of the lesion. The postoperative course was uneventful.

The clinical, neuroradiological, and intraoperative findings are presented, along with a review of the literature. Despite the number of lesions potentially compressing the spinal cord, IM schwannoma is rare but should be taken into account in the differential diagnosis [11)].

A rare case of thoracic intramedullary tumour in a 28 year old male patient who presented with progressive weakness of both the lower limbs and decreased sensation below D2 dermatomal level. Magnetic resonance imaging revealed an intramedullary lesion from D1 toD7. D1 to D7 laminoplasty and near total excision of the mass done. Total removal was difficult because of the infiltrative nature of the tumour [12)].

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