

Original Article

MRI Imaging of Dumbbell Shaped Intra Dural Extramedullary Spinal Schwannoma

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Abstract - Schwannomas are the most typical and frequent occurring intradural extramedullary tumors. These are well-encapsulated tumors that persist as cystic masses. In general, their origin is from the dorsal sensory root. Most neurinomas (about 3/4%) are solely intradural extramedullary tumors; about 1/7th have been found to be extradural. Still, more than 15% of the total have attained a "dumbbell" shape comprising both the intra- and extradural space. Neurinomas infiltrate the cervical and thoracic areas in about 3/5 % of cases and the remaining about 2/5 % in the lumbar area. Only Less than 1% exist inside the cord. Schwannomas are solitary tumors and never established in clumps. CT has been of immense use to critically expose bone erosion in its vicinity based at the foraminal canal, simultaneously associated with vertebral body scalloping. Calcifications are uncommon. On MRI imaging, neurinomas reveal isointense to the cord in about 7/10 % of the cases, while the remaining about 1/5th % reveal as moderately hypointense on T1-WI. On T2-WI, 95% of these lesions are hyperintense. Out of the total schwannomas, about 2/5 % have a cystic component. Hemorrhage has been depicted only in about 1/10 % of cases. Almost all schwannomas enhance intensely and homogeneously in contrast studies. A more heterogeneous pattern is prevalent in cystic conditions. Giant schwannomas have been critically noticed at the lumbar level and depicted to be almost indistinguishable from myxopapillary ependymoma or hemangiopericytoma. Neurofibromas are ill-defined and often persist as multiple tumors, and have never been found to be completely encapsulated. Generally, MRI is unable to differentiate between a schwannoma and a neurofibroma, especially when the tumor is solitary. In neurofibromatosis type 1, multiple plexiform neurofibromas have been peculiarly encountered. These are iso-/hyperintense on T2-WI with a "target" aspect (hyperintense rim and low/intermediate center). Enhancement is typically mild. Specifically, Malignant degeneration may occur in a few cases, especially in the typical case of neurofibromatosis type 2.

Keywords – MRI images, Spinal.

1. Introduction

Schwannoma is an extraordinary tumor originating from the sensitive tissue that enshields nerves, called the nerve sheath. These tumours develop from a type of peculiar cell called a Schwann cell, and therefore relevant exact name of the said tumor is derived. All Schwannomas are generally not cancerous. The most prevalent benign schwannoma is an acoustic neuroma. It may trigger deafness as the same appears on a nerve called the 8th cranial nerve, which accounts for hearing. Cranial Nerves: any cranial nerve may be involved, except CN I and CN II, which lack sheaths composed of Schwann cells. The most frequent involved cranial nerves are sensory nerves. However, associated with pure motor nerves may not be found to be involved as usual; these include CN VIII (acoustic neuroma), often the superior portion of the vestibular nerve (most common) in more than about 9/10% of the cases is followed by CN V, CN VII and Jugular foramen (CN IX, X, XI)^{1,2,3,9,11}.

Abundant of the schwannomas are solitary (about 9/10%) and sporadic. However, there is close proximity to NF2. Multiple schwannomas have been a peculiar

characteristic of NF2. About 1/5th of the solitary schwannomas persist in patients with NF2. The schwannomatosis process comprises multiple schwannomas without simultaneous involvement of cranial nerve VIII generally persists^{2,12,14}.

Pathologic variants include:

- Conventional (most frequently available).
- Melanotic schwannomas (involving dense melanin pigment).
- Cellular schwannomas (predominantly Antoni A tissue devoid of Verocay bodies).
- Plexiform schwannomas (remain benign, unlike plexiform neurofibromas).^{13,15}

Because acoustic schwannomas are responsible for causing damage to the fibers of the inner ear's nerve, irritation and similar ear-related symptoms have been frequently observed. Schwannomas also reveal changes in other nerves, and the noticed symptoms have been found to be associated with the specific nerve. The symptoms alter proportionate to the acquired intensity^{4,6,8,16}.



General symptoms of schwannomas reveal critical adversities such as mild to severe headache, difficulty in hearing and walking, dizziness or vertigo, frequent falls, ringing in the ear (tinnitus) and numb face.

Severe symptoms with life-threatening conditions seen are- vision alteration, inadequate balancing, difficulty in speaking and walking, frequent falls, and loss of consciousness even for a short period resulting in confusion^{5,7,17,20}.

2. Results

2.1. Case Report

A 32-year-old male patient presented to the medicine outpatient department with chief complaints of numbness, tingling sensation and weakness in left upper and lower limbs for the last 2 years. Weakness was progressive in nature.

On examination, the patient was found to have exaggerated DTRs, Decreased touch & vibration senses, increased muscle tone and reduced motor power in the left upper and lower limbs (UL=3/5LL= 4/5) - suspected UMN lesion.

2.2. Mri Brain And Cervical Spine With Contrast





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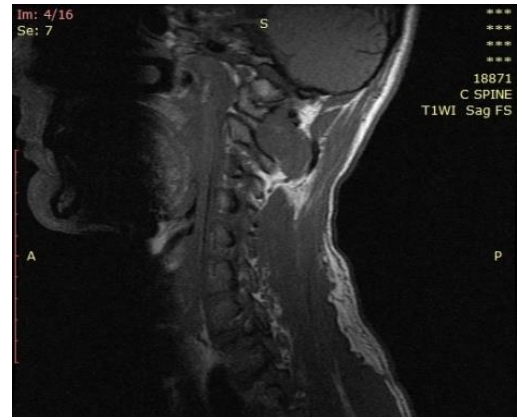
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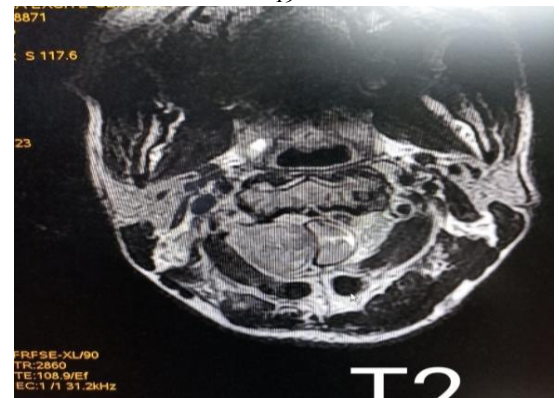
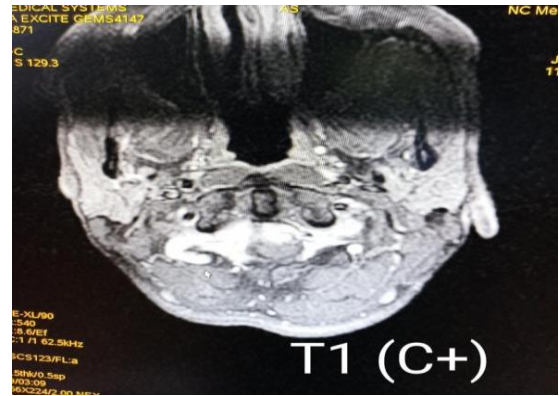
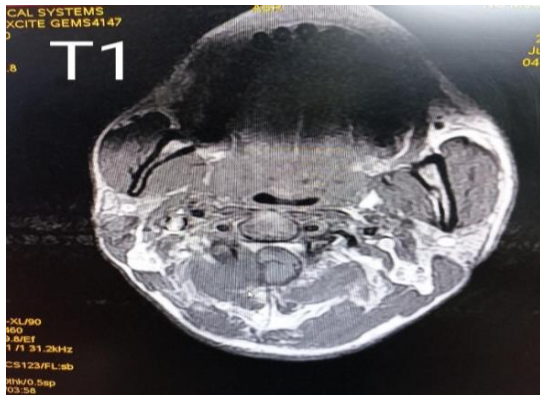
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2.3. Well-defined Intradural Extramedullary Lesion Arising From Nerve Root Causing Mass Effect Over Spinal Cord As Described Above –Likely Neurogenic Tumor.

2.3.1. DDs

- Schwannoma
- Neurofibroma

MRI Findings

There is evidence of well defined lobulated dumbbell-shaped intradural extramedullary mass lesion of approximately 26x22x25.1 mm (CCxAPxML) noted at the right anterolateral aspect which appears to arise from the C1 nerve root extending from the C1 to C2 vertebral level and laterally extending up to the nerve root foramina on the right side. This lesion occupies most of the cord, causing displacement and compression. This lesion displays an isointense signal to the cord on T1 mixed signal on T2 and STIR with intratumoral cystic areas. On Contrast, intense enhancement was seen except for intralesional cystic areas. The enhancement is also seen along the right side's nerve sheath at the posterior aspect. The underlying cord shows a hyperintense signal on T2 consistent with edema.

3. Discussion and Conclusion

Common imaging features of schwannomas are as under:

3.1. On CT

Isodense to hypodense on CT. On viewing contrast images, dense contrast enhancement is visualised. Small tumours reveal homogeneous enhancement, and larger tumours may appear as a heterogeneous enhancement. Bone remodelling may also be visualised in its vicinity.^{4,18,19}

3.2. On MRI

Cystic and fatty degeneration are most prevalent. Haemorrhage has been observed only in about 1/20% of the cases. Calcification is rarely observed. In a few cases, peripheral arachnoid cysts may be closely associated with scalloping. Peritumoural oedema can be well visualised.^{21,22,24}

3.3. Typical Signal Characteristics

T1: isointense or hypointense.

T2: hyperintense.

T1 C+ (Gd): intense enhancement duly observed.

The larger the schwannoma is, the more displays of heterogeneity owing to cystic degeneration or due to haemorrhage.

3.4. MRI Signal Characteristics

3.4.1. On T1 images

3/4% are isointense, 1/4% are hypointense.

3.4.2. On T2 images

> 19/20% are hyperintense. Schwannomas could have blended signal intensity on T2.

A hyperintense rim and central area of low signal show a particular target sign that can be visualised in neurofibromas and schwannomas. The same may result from a dense central area of the collagenous stroma.^{20,23}

3.4.3. On T1 C+ (Gd) images

virtually 100% enhance. Heterogenous enhancement accompanying areas of the low signal is highly characteristic of a neurofibroma.

Differential Diagnosis of Schwannoma/Neurofibroma

D/D	IMAGING FINDINGS
Meningioma	Broad dural-based reveals dural tail sign. Often located posterolaterally (compared to nerve sheath tumors specifically located anteriorly), neural exit foraminal widening is less frequently seen with meningiomas than nerve sheath tumors.
Paraganglioma	Has been generally visualised as a well-circumscribed mass inferior to the conus medullaris. Flow voids have been typically observed at the surface within the tumor nodule. Hemorrhage is most frequent, leading to a "cap sign" on T2 weighted images. The characteristic "salt-and-pepper" appearance of neck and skull base paragangliomas can also be visualised.
Myxopapillary ependymoma	Generally persist in the conus medullaris, and filum terminale hemorrhage is most frequent.
Intradural extramedullary metastases	Persists as peculiar and an important differential for multiple neurofibromas. Cord edema can be observed with a more dreadful disease, particularly if there is an intramedullary component. "Sugar coating" of the spinal cord and nerve roots can be well visualised.^{25,26}
Perineural root sleeve cyst	CSF density on CT; CSF intensity on T1 and T2 weighted images; no contrast enhancement has been detected.
Epidermoid or dermoid	Epidermoid similar in intensity to CSF on T1 and T2 weighted images have been critically found to be non-enhancing. However, a thin rim of contrast enhancement may be visualised as diffusion restriction on DWI.
	Dermoid presence of fat is suggestive of dermoid and generally occurs in younger patients (<20 years), and associated dermal sinus may exist.
Intradural disc herniation	No enhancement was observed.

Schwannoma Vs Neurofibroma

IMAGING FINDINGS ON MRI	SCHWANNOMA	NEUROFIBROMA
TARGET SIGN ON T2	OCCASIONALLY SEEN	COMMONLY SEEN
FASCICULAR SIGN	COMMONLY SEEN	MAY BE SEEN
SURROUNDING PARENT NERVE	PERIPHERALLY LOCATED	CENTRALLY LOCATED
ENHANCEMENT PATTERN	DIFFUSE ENHANCEMENT	CENTRAL ENHANCEMENT
CYSTIC CAVITATION/ CALCIFICATION/NECROSIS	COMMONLY SEEN	UNCOMMON

References

- [1] Al-Ghamadi S, Black MJ, Lafond G, "Extracranial Head and Neck Schwannomas," *Journal of Otolaryngology*, vol. 21, pp.186-188, 1992.
- [2] Tung H, Chen T, Weiss MH, "Sixth Nerve Schwannomas," *Journal of Neurosurgery*, vol. 75, pp. 638-641, 1991. Brogan M, Chakeres DW, "Gd-DTPA-Enhanced MR Imaging of Cochlear Schwannoma," *American Journal of Neuroradiology*, vol. 11, pp. 407-408, 1990.
- [3] Sigal R, D'Anjouard F, David P Et Al, "Cystic Schwannoma Mimicking A Brain Stem Tumor: MR Features," *Journal of Computer Assisted Tomography*, vol. 14, pp. 662-664, 1990.
- [4] Salunke AA, Chen Y, Tan JH, Et Al, "Intramuscular Schwannoma: Clinical and Magnetic Resonance Imaging Features," *Singapore Medical Journal*, vol. 56, No. 10, pp. 555-557, 2015.
- [5] Isobe K, Tominaga S, Tsutomu A, Hiroyuki K, "Imaging of Ancient Schwannoma," *American Journal of Roentgenology*, vol. 183, pp. 331-336, 2004.
- [6] Liu WC, Choi G, Lee SH, Han H, Lee JY, Jeon YH, Et Al, "Radiological Findings of Spinal Schwannomas Anmeningiomas: Focus on Discrimination of Two Disease Entities," *European Radiology*, vol. 19, pp. 2707-15, 2009.
- [7] Wilkinson JS, Mann SA, Robinson CA, Fourney DR, "Giant Cystic Intradural Lumbosacral Schwannoma: Is Stabilisation Necessary?" *Canadian Journal of Neurological Sciences*, vol. 37, pp. 535-8, 2010.
- [8] Kyoshima K, Horiuchi T, Zenisaka H, Nakazato F, "Thoracicdumbbell Intra- and Extramedullary Schwannoma," *Journal of Clinical Neuroscience*, vol. 12, pp. 481-4, 2005.
- [9] Koeller KK, Rosenblum RS, Morrison AL, "Neoplasms of the Spinal Cord and Filum Terminale: Radiologic-Pathologic Correlation," *Radiographics*, vol. 20, pp. 1721-49, 2000.
- [10] Van Goethem JW, Van Den Hauwe L, Ozsarlak O, De Schepper,AM, Parizel PM, "Spinal Tumors," *European Journal of Radiology*, vol. 50, pp. 159-76, 2004.
- [11] Perrin RG, Laxton AW, "Metastatic Spine Disease: Epidemiology,Pathophysiology and Evaluation of Patients," *Neurosurgery Clinics of North America*, vol. 15, pp. 365-73, 2004.
- [12] Flemming DJ, Murphey MD, Carmichael BB, Bernard SA, "Primary Tumors of the Spine, Semin Musculoskelet Radiol," vol. 4, pp. 299-20, 2000.
- [13] Drevelgas A, Chourmouzi D, Boulogianni G, Sofroniadis I, "Imaging of Primary Bone Tumors of the Spine," *Eur Radiology*, vol. 13, pp. 1859-71, 2003.
- [14] Murphey MD, Andrews CL, Flemming DJ, Temple HT, Et Al, "From the Archives of the AFIP, "Primary Tumors of the Spine: Radiologic Pathologic Correlatio," *Radiographics*, vol. 16, pp. 1131-58, 1996.
- [15] Wang K, Allen L, Fung E, Chan CC, Et Al, "Bone Scintigraphy in Common Tumors With Osteolytic Components," *Clinical Nuclear Medicine*, vol. 30, pp. 655-71, 2005.
- [16] Leone A, Costantini A, Guglielmi G, Settecasi C, Priolo F, "Primary Bone Tumors and Pseudotumors of the Lumbosacral Spine," *Rays*, vol. 25, pp. 89-103, 2000.
- [17] Russel DS, Rubinstein LJ, "Pathology of Tumours of the Nervous System," 5th Ed. Baltimore: Williams & Wilkins; vol. 537, 1989.
- [18] Suzuki A, Nakamura H, Konishi S, Yamanoy, "Dumbbell-Shaped Meningioma with Cystic Degeneration in the Thoracic Spine: A Case Report," *Spine (Phila Pa 1976)*, vol. 27, pp. E193-196, 2000.
- [19] Iwanami A, Kobayashi Y, Takano M, Mikami S, Toyama Y, Nakamura M, "Invasive Dumbbell Spinal Meningiomas: Report of Four Cases and A Review of the Literature," *Journal of Orthopaedic Science*, vol. 20, pp. 1148-1154, 2015.
- [20] Hirano K, Imagama S, Sato K, Kato F, Yukawa Y, Yoshihara H, Et Al, "Primary Spinal Cord Tumors: Review of 678 Surgically Treated Patients in Japan, A Multicenter Study," *European Spine Journal*, vol. 21, pp. 2019-2026, 2012.
- [21] Engelhard HH, Villano JL, Porter KR, Stewart AK, Barua M, Barker FG, Et Al, "Clinical Presentation, Histology, and Treatment in 430 Patients with Primary Tumors of the Spinal Cord, Spinal Meninges, or Cauda Equina," *Journal of Neurosurgery*, vol. 13, pp. 67-77, 2010.

- [22] Schellinger KA, Propp JM, Villano JL, McCarthy BJ, “Descriptive Epidemiology of Primary Spinal Cord Tumors,” *The Journal of Neuro-Oncology*, vol. 87, pp. 173–179, 2008.
- [23] Alorainy IA, “Dural Tail Sign in Spinal Meningiomas,” *European Journal of Radiology*, vol. 60, pp. 387–391, 2006.
- [24] Gu R, Liu JB, Zhang Q, Liu GY, Zhu QS, “MRI Diagnosis of Intradural Extramedullary Tumors,” *The Journal of Cancer Research and Therapeutics*, vol. 10, pp. 927–93, 2014.
- [25] Iwata E, Shigematsu H, Yamamoto Y, Kawasaki S, Tanaka M, Okuda A, Et Al, “Preliminary Algorithm For Differential Diagnosis Between Spinal Meningioma and Schwannoma using Plain Magnetic Resonance Imaging,” *Journal of Orthopaedic Science*, vol. 23, pp. 408–413, 2018.
- [26] Dr Parveen Chandna, Dr Aditya Duhan, Dr Rohtas K Yadav, Dr Nithin Narayanan, Dr Shikhar Gupta, Dr Vasudha, "MRI Imaging of Dengue Encephalitis," *SSRG International Journal of Medical Science*, vol. 6, no. 9, pp. 1-3, 2019.
Crossref, <https://doi.org/10.14445/23939117/IJMS-V6I9P101>.