Content available at: iponlinejournal.com

IP Indian Journal of Neurosciences

Journal homepage: www.ipinnovative.com

Case Report

Microcystic reticular schwannoma in the lumbar spine

Sonal Tripathi[1],*, Himalina Sangma[1], Anshul Singh[1], Vatsala Misra[1], Tejasvita Singh[1]

[1] *Dept. of Pathology, Moti Lal Nehru Medical College, Prayagraj, Uttar Pradesh, India*

ARTICLE INFO

Article history:
Received 27-05-2020
Accepted 02-06-2020
Available online 20-07-2020

Keywords:
Nerve sheath tumors
Benign neural neoplasms
Myxoid neural masses
Spinal schwannomas

ABSTRACT

Microcystic/Reticular Schwannoma [MRS] is a rare histological variant of Schwannoma which has been reported to mainly affect the gastrointestinal tract. Few cases involving soft tissue and various visceral and salivary glands have also been reported. However, MRS involving the spine is extremely rare with only two cases reported till date. Hereby, we present a case of Microcystic/Reticular schwannoma in a middle aged female who presented with complaints of pain and numbness in both legs. MRI findings were suggestive of an intradural extramedullary neoplasm. Diagnosis of MRS was made based on the typical histopathological features and Immunohistochemical positivity for S100. MRS is a variant of Schwannoma, which differs from the conventional Schwannoma in several aspects. Raising awareness regarding this variant is essential to the budding pathologists to prevent their misdiagnosis.

1. Introduction

Microcystic/ Reticular Schwannoma (MRS) is a rare histological variant of Schwannoma with unique histological features and predilection for visceral organs.

MRS differs from classical schwannoma in certain histomorphological aspects such as lack of hyalinised blood vessels and palisading structures that may hinder in diagnosing this variant of Schwannoma.

MRS of the spine are extremely rare with only two cases reported till date, one in the cervical region[1] and other in the lumbar region.[2] We present here a case of MRS in the lumbar spine, in a 45 year old female to highlight this rare site as well as the unique histological features that are crucial for making an accurate diagnosis of this rare entity. To the best of our knowledge, this will be the third case of MRS in the spine reported worldwide.

2. Case History

A 45 year old female with no medical record complained of continuous severe pain and numbness in both legs since 4 years. The patient was given some medications and physiotherapy for Lumbar disc herniation in a local hospital, details of which were not available. However her condition deteriorated and she developed difficulty in walking, following which she was referred to our hospital. On clinical examination, there was decreased power of bilateral lower extremities and hyperalgesia of both feet. Magnetic resonance imaging (MRI) showed an intradural altered signal enhancing lesion in the spinal cord at L1-L2 level, compressing the conus with intramedullary T2 hyperintense signal in the conus at D12 level. Diffuse disc bulges from L2 till S1 levels indenting the anterior thecal sac without overt nerve root impingement were seen. The findings were suggestive of an intradural extramedullary neoplastic lesion. Following this, Laminectomy was performed, tumor resection was done and was sent to us for histopathological examination.

Four, irregular, grey white pieces of tissue were received, the largest one measured 0.5 x 0.5 cm in size. All were processed.

Histopathological examination showed an encapsulated tumor area with meninges seen close by. Predominantly hypocellular areas (Antoni B) were seen along with

* *Corresponding author*.
E-mail address: sonaltripathi1403@gmail.com (S. Tripathi).

https://doi.org/10.18231/j.ijn.2020.028

numerous microcystic structures interconnected by bland looking spindle cells (Figure 1). The hypercellular areas (Antoni A) were very focal and showed nuclear palisading at some places though well formed Verocay bodies were not seen (Figure 2). Numerous hyalinized blood vessels in a myxoid stromal background were noted (Figure 3). On Immunohistochemistry, the tumor cells showed diffuse S-100 positivity (Figure 4). Therefore, a diagnosis of MRS was finally made. The patient was not given any additional therapy post op and was apparently doing well during follow up.

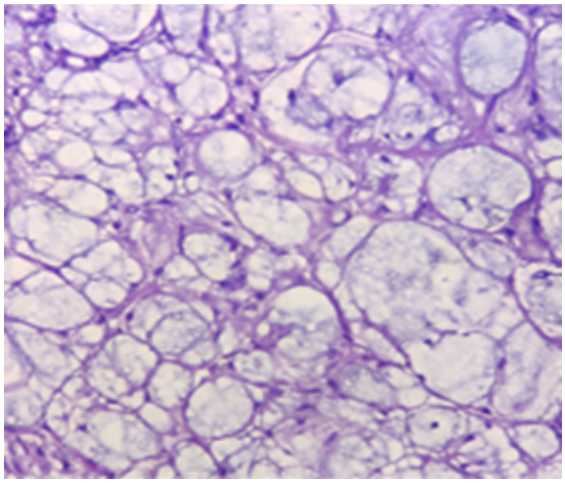

Fig. 1: Tumor cells arranged in microcytic /reticular pattern with a prominent myxoid background

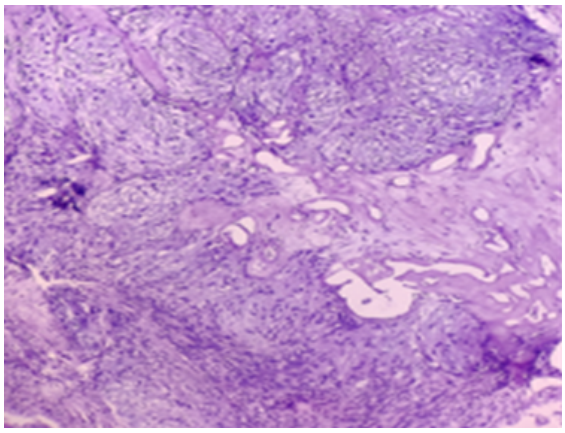

Fig. 2: Focal areas showed nuclear palisading (H and E X40)

3. Discussion

Schwannomas are benign, occasionally painful mesenchymal tumors which are derived from Schwann cells of the neural sheath. They typically occur in the 4th to 6th decades of life and mainly occur in the soft tissues of the

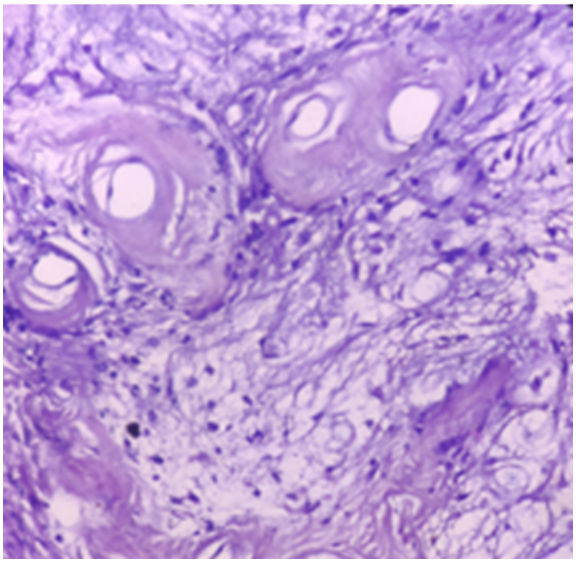

Fig. 3: Numerous hyalinized blood vessels were soon amongst the tumor cells.

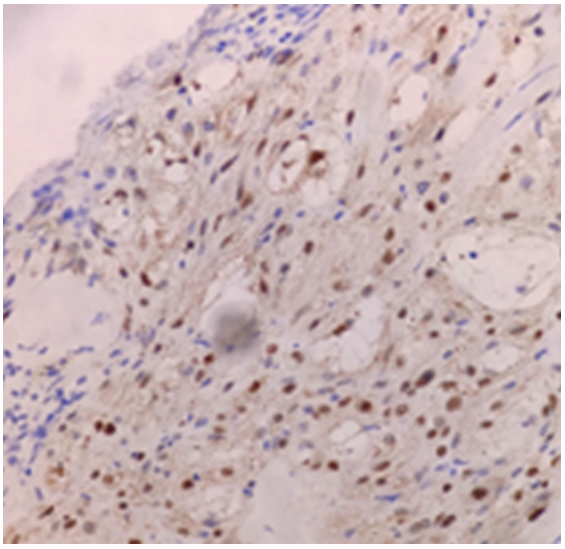

Fig. 4: Tumor cells showed positivity for S100

extremities, head and neck and the trunk. Rare anatomic sites include retroperitoneum and mediastinum. Intracranial schwannomas have predilection for 8th cranial nerve and sensory nerve roots of the spinal cord.[3]

In addition to the classical (conventional) type, different histopathological subtypes have been described. These variants include the ancient schwannoma, cellular schwannoma, plexiform schwannoma, melanotic schwannoma, epithelioid schwannoma, and hybrid forms of schwannomas.[4] However, these histopathological variants carry no prognostic significance.

On histopathological examination, Schwannomas are usually encapsulated and display two different areas in

varying ratios, referred to as Antoni A and Antoni B. Antoni A area refers to a cellular area which contains spindle-shaped cells having ill-defined eosinophilic cytoplasm with wavy basophilic nuclei. Within these, parallel rows of nuclei (Verocay bodies) are frequently seen. Antoni B areas are the hypocellular areas containing more loosely organized components in a myxoid stroma. The presence of hyalinized thick-walled blood vessels is highly characteristic.[5]

MRS is a relatively new and rarer variant of Schwannoma, which has distinct histological features. MRS was initially described in the landmark paper by Liegl et al[6] in 2008 with a predilection for the visceral organs. Subsequently they have also been reported in the soft tissue, as well as in visceral and salivary glands.[2] In the recent years, MRS have been reported in the central nervous system also, with one case in the frontal lobe[7] and two cases in the spinal cord.[1,2]

MRS differs from the classical schwannoma histologically. The spindle-shaped cells are present in a myxoid stroma. However, they may frequently lack the palisading structures and hyalinized thick-walled blood vessels. Cell atypia can be found but nuclear atypia is never seen. Mitosis and necrosis are also rare. Microcystic appearance is present when the mucus composition is relatively few, whereas reticular pattern is formed with major mucus composition.[3]

In the present case, palisading structures as well as hyalinized blood vessels were detected which pointed towards Schwannoma. The presence of microcystic structures in a myxoid background hinted towards a Microcystic/Reticular variant. No cytologic or nuclear atypia was seen and neither any mitosis and necrosis. Immunohistochemically, the tumor cells showed diffuse positivity for S-100 that confirmed the diagnosis.

Our findings were consistent with the findings of Li et al and Liu et al who also reported similar histomorphology in their respective cases of spinal Schwannomas[1,2]

Differential diagnosis of spinal MRS include Neurofibromas with extensive myxoid degeneration[8] and microcystic and myxoid Meningiomas.[8,9]

Lack of capsule, uniform arrangement of cellular components, absence of typical Antoni A and Antoni B regions and inflammatory cells in stroma are useful distinguishing features of a neurofibroma. Moreover they do not show very strong positivity for S100 as seen in schwannomas.[10]

In meningiomas, the cytoplasm of tumor cells are eosinophilic, and the nucleus is round and uniform, with commonly found intranuclear pseudoinclusions [meningothelial cells]. Besides, meningiomas are EMA positive with infrequent and patchy positivity for S-100.[8]

4. Conclusion

MRS is a unique and rare variant of schwannoma with distinct histomorphology. This case is being presented for raising awareness of this rare entity at a rare site to enrich our knowledge and prevent diagnostic pitfalls in future.

5. Acknowledgement

None.

6. Source of Funding

None.

7. Conflict of Interest

None.

References

1. Li BZ, Wang JW, Wei HQ. Microcystic/reticular schwannoma occurring in cervical spine: report of a case with literature review. *Chin J Pathol*. 2010;39:396–9.
2. Liu. Lumbar intraspinal microcystic/reticular schwannoma Case report and literature review. *Med*. 2018;97:39.
3. Antonescu CR, Perry A, Woodruff JM. WHO Classification of Tumours of Soft Tissue and Bone. IARC Press; 2013. p. 170–3.
4. Georgescu TA, Dumitru AV, Oproui AM, Nica AE, Costache D, Patrascu OM. Cutaneous microcystic/reticular schwannoma: case report and literature review of an exceedingly rare entity with an unusual presentation. *Rom J Morphol Embryol*. 2018;59:303–9.
5. Kurtkaya-Yapicier O, Scheithauer B, Woodruff JM. The pathobiologic spectrum of schwannomas. *Histol Histopathol*. 2003;18:925–34.
6. Liegl B, Bennett MW, Fletcher CDM. Microcystic/Reticular Schwannoma: A Distinct Variant With Predilection for Visceral Locations. *Am J Surg Pathol* . 2008;32(7):1080–7.
7. Pearson L, Akture E, Wonderlick J, Fuller G, Zenali M. Microcystic/Reticular Schwannoma of the Frontal Lobe: An Unusual Occurrence. *Case Rep Pathol*. 2017;2017:1–5.
8. Yamamuro K, Seichi A, Kimura A, Kikkawa I, Kojima M, Inoue H, et al. Histological Investigation of Resected Dura Mater Attached to Spinal Meningioma. *Spine*. 2012;37(22):E1398–E1401.
9. Ahuja A, Rahar S, Bharati P, Bhardwaj M, Chaudhary A. Myxoid meningioma: A case report of an extremely rare subtype of metaplastic meningioma. *Indian J Pathol Microbiol* . 2020;63(2):267–9.
10. Min H, Kim K. Differential diagnosis between nasal septal schwannoma and nasal septal neurofibroma. *J Craniofac Surg*. 2017;28:1780–3.

Author biography

Sonal Tripathi 1st Year Junior Resident

Himalina Sangma 2nd Year Junior Resident

Anshul Singh Associate Professor

Vatsala Misra Professor and Head

Tejasvita Singh 1st Year Junior Resident

Cite this article: Tripathi S, Sangma H, Singh A, Misra V, Singh T. **Microcystic reticular schwannoma in the lumbar spine**. *IP Indian J Neurosci* 2020;6(2):141-143.