SPINE An International Journal for the study of the spine, Publish Ahead of Print

DOI: 10.1097/BRS.0000000000002733

Whorling-Sclerosing Variant Meningioma of the Spine: Surgical Management and Outcome of an Extremely Rare Case

1. Keyvan Eghbal, M.D. – Department of Neurosurgery – Shiraz University of Medical Sciences, Shiraz, Iran (Email: keivaneghbal@gmail.com).

2. Amirreza Dehghanian, M.D. – Department of Pathology – Shiraz University of Medical Sciences, Shiraz, Iran (Email: amirreza_d@yahoo.com).

3. Fariborz Ghaffarpasand, M.D. – Department of Neurosurgery – Shiraz University of Medical Sciences, Shiraz, Iran (Email: ghafarf@sums.ac.ir).

Corresponding Author:
Fariborz Ghaffarpasand, M.D.
Neurosurgery Chief Resident
Department of Neurosurgery office, Namazi hospital, Namazi square, Shiraz, Iran
Postal Code: 7193711351
Cellphone: +98-917-3095214
Tel/Fax: + 98-713-6474259
Email: fariborz.ghaffarpasand@yahoo.com

The manuscript submitted does not contain information about medical device(s)/drug(s).

No funds were received in support of this work.

No relevant financial activities outside the submitted work.

ABSTRACT

STUYD DESIGN: Case Report

OBJECTIVE: To report surgical resection of multiple ventral cervical and thoracic lesion being diagnosed as whorling-sclerosing variant meningioma (WSM) and subsequent stabilization of cervicothoracic spine.

SUMMARY OF BACKGROUND DATA: WSM is a rare histopathology variant of meningioma which is mostly reported in brain and only one case of cervical spine WSM has been reported. Because of the rarity of WSM, there is a paucity of information regarding the clinical, radiological and histopathologic characteristics.

METHODS: A 57-year-old lady presented with progressive quadriparesis and myelopathy since 10 days. The spine magnetic resonance imaging (MRI) revealed multiple ventral cervical and thoracic lesions which were hypointense in T2-weighted images with heterogeneous and scant enhancement with gadolinium. The patient underwent anterior cervical corpectomy and gross total resection of the lesion along with 360-degree fixation in the first stage. The thoracic lesions were also resected totally through laminectomy and right lateral extracavitary approach followed by pedicular screw fixation.

RESULTS: The patient improved muscle powers and spasticity and is not independent (Frankel D). the histopathology examination revealed hypocellular lobulated sclerotic mass with whorling pattern of growth in the sclerotic part suggestive of WSM.

CONCLUSIONS: The spine multiple WSM is an extremely rare condition which has a malignant behavior and it seems that it is spread by CSF seeding. Surgical removal is hard and requires experience and appropriate equipment.

Key Words: Whorling-Sclerosing Meningioma; Spine; Surgical Resection; Spinal Instrumentation

Level of Evidence: 4

BACKGROUND

The whorling-sclerosing variant meningioma (WSM) was first described in 1989 by Davidson and Hope in children as a histological subtype of the meningioma being previously described.[1] Because of the rarity of this variant of meningioma, there is a paucity of information regarding the clinical, radiological and histopathologic characteristic.[2-10] The WSM of the spine is very rare and the clinical course is not well-known. Only one case of cervical intramedullary WSM has been previously described. We herein report a case of multiple cervical and thoracic WSM which was successfully managed surgically.

CLINICAL PRESENTATION

A 57-year-old lady presented to our clinic with progressive weakness and paresthesia of both upper extremities followed by gate problems since 10 days prior to presentation. She had previous history of ischemic heart disease and had undergone coronary artery bypass grafting (CABG) 5 years earlier. In physical examination, the muscle powers of the upper and lower extremities were 4/5 and the deep tendon reflexes (DTR) were hyper. The Hoffman reflex was positive and the Babinski was upward. The patients had spastic gate and urinary retention. Magnetic resonance imaging (MRI) of the whole spine was performed revealing four ventral lesion in cervical and thoracic spine. The lesions were isointense and hypointense in T1 and T2-weighted MRI, respectively (Fig. 1A) and had heterogeneous and scant enhancement after injection of gadolinium (Fig. 1B). in the first stage, we decided to operate the cervical lesion. The patient underwent anterior corpectomy of C4, C5 and C6 and microscopic total resection of the lesion was performed. anterior fixation with mesh cage and cervical plate was also performed. The lesion was reddish and firm, sticking hardly to the dura. We also performed

posterior cervical fixation to provide the stability of the cervical spine. After the first operation the patient revealed her muscle powers and the myelopathy and spasticity resolved within 2-months after the operation. After 9 months of follow-up, she developed paraparesis with muscle powers of 1/5 along with myelopathy and urofecal retention. The patient underwent second operation to remove the thoracic lesions. Bilateral laminectomy of the thoracic spine was performed and the lesions were exposed through right lateral extracavitary approach. The lesion was fleshy and reddish lesion with a wide dural base ventrolateral to the thoracic spinal cord compressing the cord both centrally and laterally (Fig. 1C). The lesions were totally resected the subarachnoid space were completely open postoperatively (Fig. 1D). We also performed pedicular screw fixation of the thoracic spine connected to the previous cervical construct (Fig. 1E). Postoperatively the patient improved the muscle powers, spasticity and sphincter function during the 6-month follow-up. Currently, she walks with aid and performs the routine daily activities independently (Frankel D). Microscopic evaluation of the tumor showed a hypocellular lobulated sclerotic mass with whorling pattern of growth in the sclerotic part (Fig 2A). At the periphery of the tumor there was a rim of small monomorphic cubidal cells with clear cytoplasm without any atypia or mitosis (Fig. 2B, 2C). Immunohistochemical evaluation demonstrated negative staining for EMA, cytokeratin, Olig-2 and S-100 (Fig. 2D) and immunoactivity for progesterone receptor (PR) in the tumoral cells located at the periphery (Fig. 2E). Ki-67 indexing was 4% which was low (Fig. 2F). The IHC was found to be highly positive (Fig. 2G). According to the histologic features and the immunohistochemical profile the diagnosis of WSM was made. The pathologic evaluation of the cervical and thoracic lesion revealed the same results. In a 1-year follow-up the patient had regained her muscle powers (upper extremity: 5/5 proximal and 4/5 distal; lower extremity: 5/5 proximal and distal). The urinary and bowel functions are normal

and mild upper extremity paresthesia is present. There is no sign of myelopathy (Frankel E). Currently, the patient can perform all the daily living activities (DLA) independently. The patient and her family provided their informed written consent to publish her case and the images in a scientific publication.

DISCUSSION

The WSM is a rare variant of meningioma which has not been included in the last edition of the WHO classification of the CNS tumors.[11] These lesions have been previously described mostly in brain [1-9,12] and only one case of cervical intramedullary WSM has been previously described.[13] We herein reported a very rare case of multiple cervical and thoracic ventrally located WSM which was surgically managed in several stages during a 1 year period. Our presented case is similar to the case presented previously by Preven et al.[13], Haberler et al.[4] and Elmaci et al.[2] regarding both clinical course and the histopathology.

There are several interesting points in the presented case which should be taken into consideration. First, the lesion was multiple and located ventrally to the spinal cord. The lumbosacral MRI after resection of all the four lesion demonstrating a small budding tumor in the lumbosacral region which bring us to the hypothesis that these lesions are spread through the CSF to the lower areas. We are now following the patient in order to test the hypothesis. Second, the tumor growth rate was significant as the thoracic lesion increased by size significantly during the 9-month between the two surgeries. And third, the consistency of the tumor was severely firm, sticking to the dura and could not be removed with conventional method. We used different methods to remove the tumor including the SONOPET Ultrasonic Aspirator, cutting by knife and cauterization. And fourth, the multiplicity and unusual location of the tumor is another point that

should be taken into consideration. We believe that this pathology should be classified into the WHO classification of the CNS tumors and the grade should be higher than I and lower than III. In conclusion, the spine multiple WSM is an extremely rare condition which has a malignant behavior and it seems that it is spread by CSF seeding. Surgical removal is hard and requires experience and appropriate equipment.

Acknowledgment: We would like to thank the patient and her family who provided us to report her case and also for her patience during the treatment procedure. We would also like to acknowledge the editorial assistance of Diba Negar Research Institute for improving the style and English of the manuscript.

REFERENCES

1. Davidson GS, Hope JK. Meningeal tumors of childhood. *Cancer* 1989;63:1205-10.

2. Elmaci I, Altinoz MA, Sav A, et al. Whorling-sclerosing meningioma. A review on the histological features of a rare tumor including an illustrative case. *Clin Neurol Neurosurg* 2017;162:85-90.

3. Fukushima S, Narita Y, Yonezawa M, et al. Short communication: sclerosing meningioma in the deep sylvian fissure. *Brain Tumor Pathol* 2014;31:289-92.

4. Haberler C, Jarius C, Lang S, et al. Fibrous meningeal tumours with extensive non-calcifying collagenous whorls and glial fibrillary acidic protein expression: the whorling-sclerosing variant of meningioma. *Neuropathol Appl Neurobiol* 2002;28:42-7.

5. Hope JK, Armstrong DA, Babyn PS, et al. Primary meningeal tumors in children: correlation of clinical and CT findings with histologic type and prognosis. *AJNR Am J Neuroradiol* 1992;13:1353-64.

6. Im SH, Chung CK, Cho BK, et al. Sclerosing meningioma: clinicopathological study of four cases. *J Neurooncol* 2004;68:169-75.

7. Kang H, Kim JW, Se YB, et al. Sclerosing Meningioma : Radiological and Clinical Characteristics of 21 Cases. *J Korean Neurosurg Soc* 2016;59:584-9.

8. Kim NR, Im SH, Chung CK, et al. Sclerosing meningioma: immunohistochemical analysis of five cases. *Neuropathol Appl Neurobiol* 2004;30:126-35.

9. Pope LZ, Tatsui CE, Moro MS, et al. Meningioma with extensive noncalcifying collagenous whorls and glial fibrillary acidic protein expression: new variant of meningioma diagnosed by smear preparation. *Diagn Cytopathol* 2003;28:274-7.

10. Farrokhi MR, Ghaffarpasand F, Khani M, et al. An Evidence-Based Stepwise Surgical Approach to Cervical Spondylotic Myelopathy: A Narrative Review of the Current Literature. *World Neurosurg* 2016;94:97-110.

11. Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol* 2016;131:803-20.

12. Masoudi MS, Haghnegahdar A, Ghaffarpasand F, et al. Functional Recovery Following Early Kyphoplasty Versus Conservative Management in Stable Thoracuolumbar Fractures in Parachute Jumpers: A Randomized Clinical Trial. *Clin Spine Surg* 2017;30:E1066-e73.

13. Perven G, Entezami P, Gaudin D. A rare case of intramedullary 'whorling-sclerosing' variant meningioma. *Springerplus* 2015;4:318.