

Multiple spinal and cranial meningiomas: A case report and review of literature

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ABSTRACT

Though meningiomas are common neoplasms of the nervous system, the occurrence of multiple meningiomas in different neuraxial compartments is rather rare. We report a case of a 62-year-old female who presented with spastic paraparesis in both lower limbs, and was found to have multiple homogenously enhancing tumors of dorsolumbar spine. Cranial magnetic resonance imaging (MRI) revealed multiple bilateral supratentorial meningiomas. She underwent multiple level laminectomy and total removal of spinal tumors after which power improved in both lower limbs. Histopathology revealed psammomatous meningiomas. Only around 19 cases of multiple cranial and spinal meningiomas have been reported, of which, only five cases have more than one spinal meningioma. The implication of the incidence of such multiple meningiomas in the same patient with relevance to investigations and decision making are discussed along with a brief review of literature of cases with multiple spinal and cranial meningiomas.

Key words: Adult, craniospinal, meningioma, multiple, psammomatous

Introduction

Meningiomas are the most common primary non-gliar brain tumors and comprise 13-19% of all primary intracranial neoplasm.^[1] Spinal canal meningiomas account to about 25% of all spinal cord tumors.^[2] Multiple spinal meningiomas are rarer than multiple cranial meningiomas.^[3] Multiple meningiomas occurring in different neuraxial compartments are distinctly rare, with only 19 well-documented cases reported in world literature,^[4] of these, only six cases had more than one spinal tumor. Occurrence of such multiple meningiomas of spinal and cranial distribution in absence of neurofibromatosis is quite rare.

Case Report

A 62-year-old female presented with backache, pain radiating to both lower limbs, and difficulty in walking. On examination, she had grade 3 power at all joints in both lower limbs and

increased tone. She had no history of seizures, headache, or any other symptom attributable to intracranial pathology. There were no multiple hypopigmented macules, neurofibromas, or other stigmata of neurofibromatosis. Magnetic resonance imaging (MRI) of the dorsolumbar spine was performed [Figure 1], which revealed four well defined, oval, intradural extramedullary, homogenously enhancing solid lesions at D7-8, D11-12, and at D12-L1 levels causing cord compression at all levels. An initial diagnosis of multiple thoracolumbar meningiomas was made, and the patient was subjected to a contrast enhanced MRI of the brain. MRI of the brain revealed multiple extra axial, well defined, homogenously enhancing lesions in the right frontal and left parietal region [Figure 2]. A final diagnosis of multiple cranial and spinal meningiomas was made. The patient underwent multiple level laminectomy and complete removal of all tumors from dorsolumbar region. Post operatively, the patient's power improved and she was able to walk with support after a week. A contrast enhanced MRI of the spine showed complete removal of tumors and decompression of cord [Figure 3]. Histopathology revealed psammomatous meningioma [Figure 4]. Despite our detailed counseling, the patient and their relatives steadfastly refused surgery for intracranial tumors. Patient was discharged on antiepileptics and is under close follow-up.

Discussion

Multiple meningiomas are defined as at least two spatially separated meningiomas occurring at the same time, or more than two meningiomas arising sequentially from two clearly distinct regions.^[5]

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Figure 1: Contrast enhanced MRI spine showing multiple well defined enhancing lesions

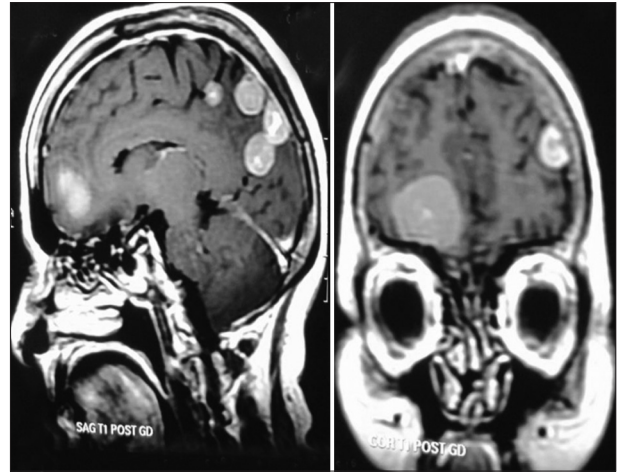


Figure 2: Contrast enhanced MRI brain showing multiple enhancing lesions in both cerebral hemispheres

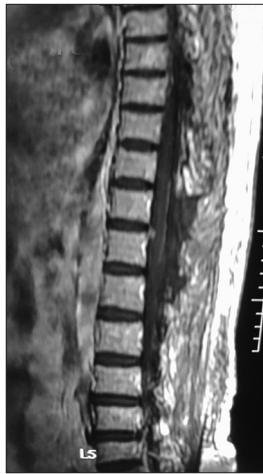


Figure 3: Post operative contrast enhanced MRI spine showing complete removal of meningioma

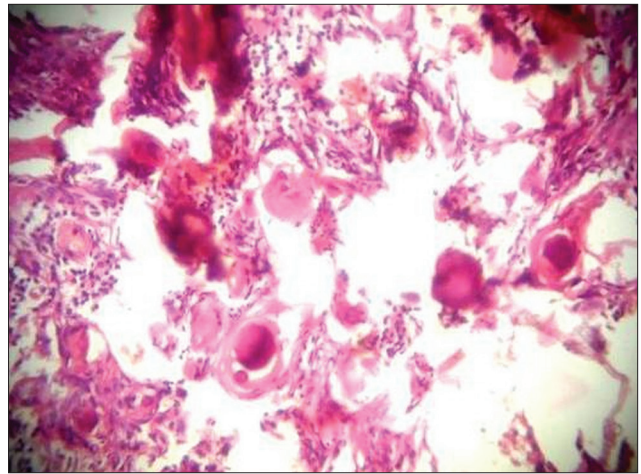


Figure 4: Histopathology showing psammomatous meningioma

The incidence of multiple intracranial meningiomas in the post-CT era has been reported to be between 5.4 and 8.9%. The majority of these are located in a hemispherical distribution.^[4,6] Multiple spinal meningiomas are rarer than multiple cranial meningiomas. Multiple meningiomas occurring in different neuraxial compartments are distinctly rare, with only six cases having multiple spinal and multiple cranial meningiomas [Table 1]. The previous oldest reported case was a 50-year-old male, thus making our case the oldest reported. Four of the six cases were aged 35 years or less and all except one that was a female.

The pathogenesis of multiple meningiomas can be explained in two ways, either these tumors arise independently as evidenced by the histological and cytogenetic differences between multiple tumors from the same patient, or a single transforming event occurs and the original clone of cells spreads throughout the meninges in the formation of multiple, clonally related tumors.^[1,6]

The relationship between isolated multiple meningiomas and neurofibromatosis is unsettled.

Meningiomas are found in about half the patients with Neurofibromatosis type 2 (NF2) and sporadic meningiomas often have somatic mutations in the NF2 gene. NF2 is often the underlying disease in young people who present with meningioma, but adults with multiple meningiomas and no other signs of NF2 are usually not considered to be at high risk for NF2.^[11]

Of the intracranial meningiomas, one percent are multiple, usually in neurofibromatosis. The most common locations are: Falx and parasagittal, convexity, sphenoid, and olfactory groove.^[12] In the spinal canal, the preferred location of meningiomas is at the thoracic level followed by the cervical region, and finally, the lumbar region.^[2]

The clinical feature is characterized by a motor deficit, varying from a slight impairment to paralysis, with pyramidal liberation signs, sphincter disturbances and signs of funicular or radicular impairment. Ordinarily, the symptomatology is restricted to a root or is associated to several sensitive and motor neurological signs.^[2]

Table 1: List of reported cases of multiple meningiomas with more than one spinal meningioma

Year reported	Author	Age/sex	Cranial location	Spinal location	Histopathology
1973	Sedzimir <i>et al.</i> ^[7]	13/m	Tuberculum sellae	Cervical ^[3] Dorsal ^[4]	Cranial-not resected Spinal-psammomatous
1992	Roda <i>et al.</i> ^[8]	50/m	Suprasellar ^[1] Falcine ^[1]	Dorsal ^[2]	Cranial and spinal- meningotheliomatous
2003	Bhatoe H S ^[4]	35/f	Multiple supra and infra tentorial	Dorsal ^[2]	Cranial-transitional, meningothelial Spinal-fibroblastic, atypical
2011	Stachowicz-stencel T <i>et al.</i> ^[9]	13/f	Multiple supra and infra tentorial	Lumbosacral multiple	Cranial-psammomatous Spinal-meningioma
2011	Shukla <i>et al.</i> ^[10]	13/f	Left frontoparietal	Dorsal ^[2] Lumbar ^[3]	Spinal-predominantly meningotheliomatous
2011	Present case	62/f	Multiple bilateral supratentorial	Dorsal ^[3] Lumbar ^[2]	Spinal-psammomatous

Number in superscript in author column indicate the relevant reference number, number in parentheses in cranial/spinal column indicate number of tumors excised

The radiological features are similar to isolated meningiomas. Well described features include well defined extra axial lesions that enhance uniformly on contrast administration, enhancement of the dural attachment and little or no perilesional edema.

Operative management of multiple meningiomas occupying both cranial and spinal compartments poses special problems. A decision is made regarding which lesion, or lesions should be removed initially keeping in mind that histologically, each of these tumors may be a different variant of meningioma. Small or asymptomatic tumors may be followed-up with serial imaging.

The majority of multiple meningiomas (80-90%) are benign and classified as World Health Organization (WHO) grade 1.^[6] Regarding the cases of spinal canal multiple meningiomas, the psammomatous type predominate. Usually, multiple meningiomas are circumscribed and show slow growth and have a good post surgical prognosis. On the other hand, the meningiomas that occur in younger patients might have an aggressive behavior and an unfavorable prognosis.

Conclusion

Although meningiomas are one of the most common tumors encountered in neurological practice, multiple meningiomas still remain a rare entity. Although the incidence of multiple meningiomas may not be sufficient to warrant a complete evaluation of the brain and spine in all cases where a solitary meningioma is diagnosed, features that should alert the surgeon include early age of onset, female sex, multiple spinal tumors, and presence of neurofibromatosis. When silent meningiomas are discovered in a neuraxial compartment different from the one causing the symptoms careful decision making is needed taking into consideration the presenting features, age, sex, tumor biology, associated diseases and patients expectations

from surgery. Regardless of the eventual management decision, such patients must be kept under close follow-up.

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