Surgical Neurology International

OPEN ACCESS

For entire Editorial Board visit : http://www.surgicalneurologyint.com

Editor: **James I. Ausman,** MD, PhD University of California, Los Angeles, CA, USA

Case Report

Meningiomatosis restricted to the left cerebral hemisphere with acute clinical deterioration: Case presentation and discussion of treatment options

Victoria Ohla[1,2], Christian Scheiwe[1]

[1]Department of Neurosurgery, University of Freiburg, Breisacherstr. 64. 79106 Freiburg, [2]University of Essen, Hufelandstrasse 55, 45147 Essen, Germany

E-mail: *Victoria Ohla - Victoria.Ohla@uk-essen.de; Christian Scheiwe - Christian.Scheiwe@uniklinik-freiburg.de
*Corresponding author

Received: 12 December 14 Accepted: 28 January 15 Published: 20 April 15

This article may be cited as:
Ohla V, Scheiwe C. Meningiomatosis restricted to the left cerebral hemisphere with acute clinical deterioration: Case presentation and discussion of treatment options. Surg Neurol Int 2015;6:64.
Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2015/6/1/64/155447

Abstract

Background: True multiple meningiomas are defined as meningiomas occurring at several intracranial locations simultaneously without the presence of neurofibromatosis. Though the prognosis does not differ from benign solitary meningiomas, the simultaneous occurrence of different grades of malignancy has been reported in one-third of patients with multiple meningiomas. Due to its rarity, unclear etiology, and questions related to proper management, we are presenting our case of meningiomatosis and discuss possible pathophysiological mechanisms.

Case Description: We illustrate the case of a 55-year-old female with multiple meningothelial meningeomas exclusively located in the left cerebral hemisphere. The patient presented with acute vigilance decrement, aphasia, and vomiting. Further deterioration with sopor and nondirectional movements required oral intubation. Emergent magnetic resonance imaging (MRI) with MR-angiography disclosed a massive midline shift to the right due to widespread, plaque-like lesions suspicious for meningeomatosis, purely restricted to the left cerebral hemisphere. Emergency partial tumor resection was performed. Postoperative computed tomography (CT) scan showed markedly reduction of cerebral edema and midline shift. After tapering the sedation a right-sided hemiparesis resolved within 2 weeks, leaving the patient neurologically intact.

Conclusion: Although multiple meningeomas are reported frequently, the presence of meningeomatosis purely restricted to one cerebral hemisphere is very rare. As with other accessible and symptomatic lesions, the treatment of choice is complete resection with clean margins to avoid local recurrence. In case of widespread distribution a step-by-step resection with the option of postoperative radiation of tumor remnants may be an option.

Key Words: Meningioma, multiple meningioma, meningiomatosis, meningothelial meningioma

Access this article online

Website: www.surgicalneurologyint.com

DOI: 10.4103/2152-7806.155447

Quick Response Code:

INTRODUCTION

Meningioma is one of the most frequent adult primary brain tumor accounting for 15% of intracranial tumors and 30% of all central nervous system tumors originating from the meningeal coverings of the spinal cord and the brain. In 1938, the term of multiple meningiomas was coined by Cushing *et al.*[7] who described a patient suffering of several meningiomas at different locations. Multiple meningioma is therefore defined as at least two spatially seperated meningiomas occuring simultaneously or more than two meningiomas arising sequentially from two clearly distinct regions.[23] Confluent meningiomas or clusters of meningiomas are referred to as diffuse meningiomatosis, which is considered to be an extreme form of multiple meningioma.[12] They occur with an incidence of approximately 2%[2,6,15] before and up to 5.9–10.5% after the advent of CT and MRI scans.[15,18]

Until now, this rare entity and its clinical features are not well understood.

They are known to occur more frequently in women and elderly people. As multiple meningiomas can be associated with other neoplasms such as neurofibromatosis, the distinction between true multiple meningiomas and those which should be considered as a special variant of von Recklinghausen's disease is not always clear-cut. Furthermore several case reports on familal meningiomatosis[4] in patients without neurofibromatosis have been published prior to the National Consensus Statement on Neurofibromatosis in 1987,[1,4] which would nowadays be considered to have neurofibromatosis. It is therefore difficult to estimate the prevalence of meningiomatosis unrelated to NF2 but it is assumed to be very seldom.[16]

Nevertheless, though being a slow growing tumor[19] with a growth rate comparable to incidentally found solitary meningiomas,[27] meningiomatosis potentially may lead to sudden decompensation and death[10] and therefore requires special attention.

CASE REPORT

A 55-year-old right-handed female was transferred to our center because of acute progressive vigilance decrement. There was no family history of malignant tumor or neurofibromatosis. At time of admission to our department, the patient was already intubated, unconscious but moving all extremities spontaneously. Immediate MRI [Figure 1] was performed disclosing tumor masses involving multiple areas of the frontal and parietal lobes of the left cerebral hemisphere, the left posterior horn and infiltrating the superior sagittal sinus almost leading to upper and lower brain herniation. Based on radiographic criteria, massive meningeomatosis was suspected and after adequate antiedematous therapy, emergent debulking was initiated.

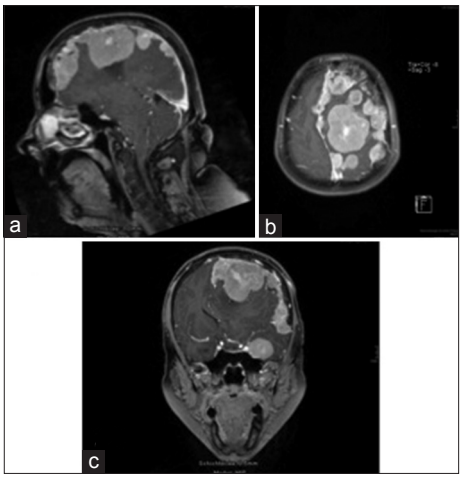

Figure 1: Preoperative T1-weighted MRI with contrast enhancement in (a) sagittal, (b) axial and (c) coronal view, showing massive midline shift to the right due to widespread, plaque-like lesions purely restricted to the left cerebral hemisphere

The patient was placed under general endotracheal anesthesia and positioned supine with the head turned to the contralateral side. A frontoparietal craniotomy was performed, followed by successive tumor removal paramedial of the falx, at various locations of the frontal lobe, frontal pole, and parietal lobe. Residual tumors remained medial and lateral to the sphenoid bone as well as in the temporal lobe and were planned to be resected in a second step.

Intraoperative histopathological frozen-section analysis indicated a leptomeningeal differentiated tumor. Immunohistochemical studies were performed staining positive for Vimentin and epithelial membrane antigen (EMA). Pathological examinations showed syncytial and epithelial cells as well as indistinct cell borders and multiple psammoma bodies. In addition, the tumor cells were often found in characteristic whorled arrangements. Based on the histopathological features and classic immunostaining, the diagnosis of meningothelial meningeoma, World Health Organization (WHO) grade I was made. Due to delayed postoperative arousal of the patient, a CT was performed featuring a reduction of the midline shift from 18 to 9 mm. Antiedematous therapy was therefore continued. On the 6th postoperative day, the patient slowly regained consciousness, showing spontaneous movements in her left extremities but delayed reactions on her right-sided extremities with a hemiparesis of muscle strength grade 2/5. After extubation on day 6, the patient rapidly improved, demonstrating only minor deficits on her right side of her body (muscle strength grade 4/5). The patient was transferred to a rehabilitation facility on day 11.

Only a mild right foot dorsiflexion deficit (muscle strength grade 5−/5) was noticed at a follow-up visit 3 months later. Subsequent MRI scans with and without gadolinium showed calcified frontal and occipital lesions as well as the known left residual posterior horn, temporoparietal and sphenoid wing meningiomas [Figures 2 and 3]. Due to the space-occupying effect of the left temporopolar and sphenoid wing tumors, a second step resection was performed 3 months after the initial surgery. A pterional craniotomy was performed and total microsurgical resection of the temporal tumors achieved. Postoperative evaluation showed no new deficits. Histopathological examination again revealed a meningothelial meningeoma, WHO grade I.

Due to the subtotal removal, subsequent follow-up with MRI is scheduled in 6 months, with option of adjuvant radiation in case of radiographic progression.

DISCUSSION

Though meningiomas are ordinarily seen in the field of neurosurgery, true meningiomatosis occurs with an incidence of only 2%[2,6,15] in surgical series while the incidence seen in autopsy series varies between 8.2% and 16.9%.

They were first described in 1889 by Anfimow and Blumenau[3] and classified as a discrete nosological entity by Cushing and Eisenhardt in 1938[7] who clearly distinguished the condition from diffuse meningiomatosis, neurofibromatosis, and recurrences. An increased incidence of solitary meningioma in females is a well known fact that is attributed to hormonal influences such as progesterone.[21] Sheehy *et al*.[22] furthermore presented a case study of 10 patients with multiple meningiomas in which all patients were females. A similar observation was made by Domenicucci *et al*.[8] in whose cohort 13 out of 14 patients with multiple meningiomas were females. We therefore concluded a preponderance of multiple meningiomas in females, although studies of female sex hormone receptors are rare and further analysis is needed. The pathogenic factors related to the development of multiple meningiomas are unknown and various theories have been reported.

As most meningeomas are histologically benign, a spontaneous dissemination through the venous system or seeding after surgery is very unlikely.[13,26] On the one hand a noncontiguous spread of original clones of cells via the cerebrospinal fluid (CSF) space or throughout the meninges resulting in the formation of multiple and clonally related tumors has been debated.[9,24] Different molecular analysis have shown that multiple meningiomas had a loss of the same copy of chromosome 22 or an inactivation of the same X chromosome, strengthening the theory that the tumor arise from the same clone cells.[11,14,20,29]

On the other hand, the hypothesis of multicentric neoplastic foci states an origin from multiple sites in which the tumors develop independently under stimulation of a supposed tumor-producing factor. This opinion is supported by Butti *et al*.,[5] who excluded cell migration through the subarachnoid space as a pathogenetic factor in multiple meningiomas after evaluating eight cases of multiple meningiomas treated over a 13-year period and believes that these tumors are due to inherent multicentricity of the dural foci and arise independently.[21]

This theory is shared by Stangl *et al*.,[24] who investigated 39 tumors in 12 patients by analyzing the entire coding region of the NF2 gene and evaluating altered fragments by DNA sequencing.[24] As the majority of multiple meningioma patients with NF2 gene mutations exhibited the identical DNA alterations in the NF2 gene, a monoclonal origin of MM was concluded. Similar results were found by Zhu *et al*.[29] who demonstrated the loss of the same copy of chromosome 22 as well as a common unmethylated allele at the AR locus in his cohort of eight

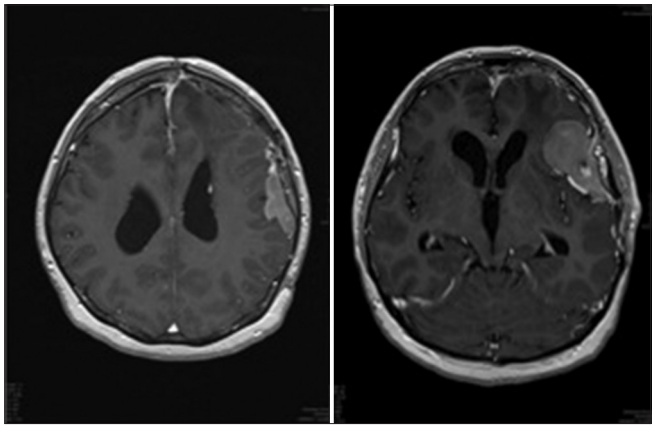

Figure 2: Follow-up MRI 3 months postoperatively, showing complete resolution of midline shift in axial T1 view

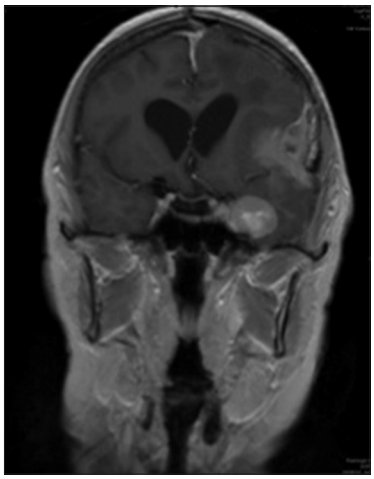

Figure 3: Follow-up MRI 3 months postoperatively, showing in a coronar T1 view with contrast enhancement residual tumors at the lateral and medial sphenoid wing

patients and therefore concluding a monoclonal origin.[29]

In the context of the pathogenesis of meningeal neoplasia Morrison *et al*. could further prove ethylnitrosourea (ENU) to cause meningiomatosis in a mouse model.[17]

Rapid clinical deterioration due to tumor growth and cerebral edema as in the presented case is very rare and requires urgent treatment. In our case, we decided to perform a two-step-operation with resection of the tumors located in the frontal and parietal region causing midline shift and edema first and in a second step (after completing rehabilitation) the tumors of the lateral and medial sphenoid wing in order to avoid an involvement. The reason was to avoid an involvement of the whole hemisphere in a former healthy patient who was not aware of his disease.

The management of a patient with meningeomatosis can be very challenging.

Though MRI is often sufficient, angiography might be indicated preoperatively to determine the status of the sagittal sinus, the location of cortical veins and the relationship of cortical arteries[25] may require selective preoperative embolization of the meningiomas.

A pretreatment with dexamethasone and proton pump inhibitors should occur prior to surgical resection to decrease the amount of edema and therefore reducing the need for retraction. Ideally, complete surgical resection should be opted, however, total resection might not be possible based upon the size and location of the tumors. In this case, only the main symptomatic mass may be removed[28] and radiotherapy should be considered as adjuvant therapy.

CONCLUSION

Although meningeomas are frequently encountered, the presence of meningeomatosis purely restricted to one cerebral hemisphere is seldom. Though its prognosis does not differ from benign solitary meningiomas, open surgery is the gold standard with its aim of complete tumor removal. In emergency cases, a resection of the largest tumors may be useful as a first treatment.

REFERENCES

1. Neurofibromatosis. Conference statement. National Institutes of Health Consensus Development Conference. Arch Neurol 1988;45:575-8.
2. Andrioli GC, Rigobello L, Iob I, Casentini L. Multiple meningiomas. Neurochirurgia (Stuttg) 1981;24:67-9.
3. Anfimow J BL. Ein Fall multiplier Geschwulste in der Schadelhohle. Neurol Zentralbl 1889;585.
4. Atkinson JL, Lane JI. Frontal sagittal meningioma: Tumor parasitization of cortical vasculature as the etiology of peritumoral edema. Case report. J Neurosurg 1994;81:924-6.
5. Butti G, Assietti R, Casalone R, Paoletti P. Multiple meningiomas: A clinical, surgical, and cytogenetic analysis. Surg Neurol 1989;31:255-60.
6. Castro A, Klug W. Proceedings: Multiple meningiomas. Acta Neurochir (Wien) 1975;31:278-9.
7. Cushing H, Eisenhardt L. Meningiomas, their classification, regional behaviour, life history and surgical end results. Springfield: Ch.C Thomas; 1938.
8. Domenicucci M, Santoro A, D'Osvaldo DH, Delfini R, Cantore GP, Guidetti B. Multiple intracranial meningiomas. J Neurosurg 1989;70:41-4.
9. Heinrich B, Hartmann C, Stemmer-Rachamimov AO, Louis DN, MacCollin M. Multiple meningiomas: Investigating the molecular basis of sporadic and familial forms. Int J Cancer 2003;103:483-8.
10. Karabagli P, Karabagli H, Yavas G. Aggressive rhabdoid meningioma with osseous, papillary and chordoma-like appearance. Neuropathology 2014;34:475-83.
11. Larson JJ, Tew JM Jr, Simon M, Menon AG. Evidence for clonal spread in the development of multiple meningiomas. J Neurosurg 1995;83:705-9.
12. Lazorthes G, Anduze-Acher H, Amaral-Gome F. Les meningiomes multiples, les meningiomatoses, et les meningiomes associes a d'autres tumeurs nerveuses. Neurochirurgie 1960;6:156-60.
13. Levin P, Gross SW, Malis LI, Kirshenbaum AH, Hollin SA. Multiple Intracranial Meningiomas. Surg Gynecol Obstet 1964;119:1085-90.
14. Lomas J, Bello MJ, Alonso ME, Gonzalez-Gomez P, Arjona D, Kusak ME, *et al*. Loss of chromosome 22 and absence of NF2 gene mutation in a case of multiple meningiomas. Hum Pathol 2002;33:375-8.
15. Lusins JO, Nakagawa H. Multiple meningiomas evaluated by computed tomography. Neurosurgery 1981;9:137-41.
16. Maxwell M, Shih SD, Galanopoulos T, Hedley-Whyte ET, Cosgrove GR. Familial meningioma: Analysis of expression of neurofibromatosis 2 protein Merlin. Report of two cases. J Neurosurg 1998;88:562-9.
17. Morrison JP, Satoh H, Foley J, Horton JL, Dunnick JK, Kissling GE, *et al*. N-ethyl-N-nitrosourea (ENU)-induced meningiomatosis and meningioma in p16(INK4a)/p19(ARF) tumor suppressor gene-deficient mice. Toxicol Pathol 2007;35:780-7.
18. Nahser HC, Grote W, Lohr E, Gerhard L. Multiple meningiomas. Clinical and computer tomographic observations. Neuroradiology 1981;21:259-63.
19. Nakamura M, Samii M. Surgical management of a meningioma in the retrosellar region. Acta Neurochir (Wien) 2003;145:215-9; discussion 219-20.
20. Petrella R, Levine S, Wilmot PL, Ashar KD, Casamassima AC, Shapiro LR. Multiple meningiomas in a patient with constitutional ring chromosome 22. Am J Med Genet 1993;47:184-6.
21. Santoro A, Tacconi L, Paolini S, Bristot R, Bastianello S. Multiple meningiomas in different neuraxial compartments. Report of two cases. J Neurosurg Sci 1999;43:211-4; discussion 214-5.
22. Sheehy JP, Crockard HA. Multiple meningiomas: A long-term review. J Neurosurg 1983;59:1-5.
23. Spallone A, Neroni M, Giuffre R. Multiple skull base meningioma: Case report. Surg Neurol 1999;51:274-80.
24. Stangl AP, Wellenreuther R, Lenartz D, Kraus JA, Menon AG, Schramm J, *et al*. Clonality of multiple meningiomas. J Neurosurg 1997;86:853-8.
25. Tuna M, Gocer AI, Gezercan Y, Vural A, Ildan F, Haciyakupoglu S, *et al*. Huge meningiomas: A review of 93 cases. Skull Base Surg 1999;9:227-38.
26. Winkelman NW. Post-operative seeding of the subarachnoid space and ventricles from a meningioma. J Neuropathol Exp Neurol 1954;13:260-6.
27. Wong RH, Wong AK, Vick N, Farhat HI. Natural history of multiple meningiomas. Surg Neurol Int 2013;4:71.
28. Yaşargil MG: Meningiomas, In: Yaşargil MG, editor. Microneurosurgery: Microsurgery of CNS Tumors v. 4B: Thieme Publishing Group; Vol IVB, p. 154.
29. Zhu JJ, Maruyama T, Jacoby LB, Herman JG, Gusella JF, Black PM, *et al*. Clonal analysis of a case of multiple meningiomas using multiple molecular genetic approaches: Pathology case report. Neurosurgery 1999;45:409-16.