



Multiple Intracranial Meningioma in a Woman with Headache

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ABSTRACT

Meningioma is one of the most common primary brain tumors in adults. The literature indicates a higher prevalence of multiple meningiomas in women, which might be attributed to the hormonal factors, such as expression of progesterone receptor.

Here we report a 58-year-old woman with a 4-year history of headache, which was aggravated during the few months before her admission. The patient was diagnosed with multiple meningiomas based on her brain magnetic resonance imaging. Therefore, she underwent surgical excision of the tumor and had no complications or signs of recurrence in the site of surgery, after a 3-year follow-up.

Although multiple meningioma is not frequent, it does not differ from solitary tumors in terms of prognosis and management. Our experience shows that complete removal of the tumors is not possible in all cases of multiple meningioma and resection of the largest tumors results in favorable outcomes.

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Introduction

Meningiomas are benign tumors originating from arachnoidal cap cells of the leptomeninges. The annual incidence of meningioma is 5.5 per 100000 people and accounts for 13-26% of all the primary intracranial neoplasms in adults (1,2).

Multiple meningiomas of the central nervous system are only 1-2% of all meningiomas. Multiple meningiomas are relatively rare tumors that accompany either peripheral or central types of neurofibromatosis (i.e., NF-1 and NF-2). In neurofibromatosis several small neoplasms spread under the pachymeninges as a manifestation of neurofibromatosis. In addition, meningiomas are detected in about 50% of neurofibromatosis type 2 patients characterized by absence of the NF2 gene on chromosome 22 encoding the merlin tumor suppressor (3-5).

The World Health Organization (WHO) classifies meningiomas according to the histological findings. The categories include WHO grade I meningioma (~90%) which is a slow progressing benign

tumor, WHO grade II meningioma (6-8%) manifested as atypical cells with increased cellularity and mitotic activity, and WHO grade III meningioma (2-3%) known as anaplastic or malignant type with a high probability of recurrence. However, even the benign tumors can grow larger and cause life-threatening complications due to a mass effect on adjacent brain tissue or involvement of main cerebral arteries (4,6).

Meningiomas are common, extra-parenchymal, and benign tumors of central nervous system that usually present with symptoms resulting from mass effect. Consequently, clinical presentation usually depends on the location of the tumor producing neurological signs and symptoms, epileptic fits or symptoms of augmented intracranial pressure. Many of these cases can be asymptomatic leading to these tumors be detected at post-mortem studies. However, they may contribute to significant surrounding edema, probably associated with their cellular morphology and capability of

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releasing vasoactive substances. The severity of edema is not related to the size of the tumor and may cause life-threatening increase in intracranial pressure (7,8).

Radiologic imaging is the main method for pre-operative diagnosis and evaluation of meningioma. It should be noted that computed tomography (CT) and magnetic resonance imaging (MRI) are preferred. Proton magnetic resonance spectroscopy is a noninvasive technique that enables further assists and improvements in diagnosis of brain lesions, including meningiomas (9-11). Surgical resection is the standard treatment for meningiomas and endovascular embolization is usually performed before surgery to reduce tumor vascularity. When the surgeon is unable to resect the tumor completely, radiation therapy and hormonal therapy are indicated (12).

Resection of meningioma depends on the location and size of the lesion and surgery often results in complete tumor excision and symptoms relief. The choice treatment for meningioma is maximum safe surgical resection, in which the normal adjacent tissue remains intact while the tumoral tissue is excised completely. However, for meningiomas of skull with involvement of the cavernous sinus, petroclival region, or medial temporal fossa, complete excision of the lesion is often not possible.

These interventions can lead to significant complications, such as the risk of cranial nerve deficits. As a result, alternative therapies such as fractionated stereotactic radiotherapy or stereotactic radiosurgery are applied in the adjuvant and primary setting. Advantages and disadvantages of surgical removal of meningiomas should be evaluated considering the potential risks and benefits.

Age is an important factor that needs to be considered in these patients and life expectancy is short for the old patients. Therefore, the lesion is rarely threatening for these patients and their poor general health condition might cause the surgery to result in various complications in this group. However, radical excision of the lesion is still the first choice of treatment for aged people who are socially active and are in good general health condition. This approach, application of new imaging methods, and intraoperative use of advanced equipment make the duration of operation shorter, in addition to facilitating the post-operative monitoring and intensive care. All the mentioned factors contribute to lower risk of mortality and morbidity in this age group (13-15).

Case Report

The patient was a 58-year-old woman, with the chief complaint of headache since 4 years ago, which was aggravated during the last few months

before her admission. She had no history of nausea or vomiting and in her past medical history, she had nothing important but addiction to opium. Moreover, she denied any familial history of neurofibromatosis or other similar disorders.

She was first managed in an outpatient service then was referred to our center. Her neurological examination was normal with no focal signs and she was in full conscious state with a Glasgow Coma Scale of 15. Furthermore, All the cranial nerves were found to have normal function. The patient did not show hyperreflexia in deep tendon reflex examination and Muscle force exam revealed normal muscle force (5/5) in all four extremities.

The results of MRI showed 12 foci of intracranial tumors in both infra- and supra-tentorial regions, including convexity, parasagittal, falicine, cerebellopontine angle, tentorial, and clinoidal with mass effect and a shift in the midline to the left side (Figures 1 and 2). Almost all the tumor foci were located in the right hemisphere of the brain or midline region. Therefore, the patient was operated under general anesthesia. A bicoronal incision was made and unilateral frontotemporoparietal osteoblastic flap was removed. The dura was opened with sphenoid ridge base and the two cortical tumors in the convexity region were excised and sent for histopathological examination.

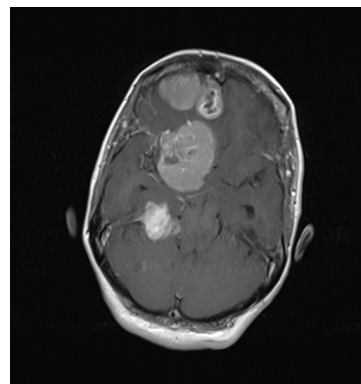


Figure 1: Brain magnetic resonance imaging with contrast, axial view; Meningiomas in the right frontal convexity, parasagittal, right cerebellopontine angle, and right sphenoidal ridge with mass effect.



Figure 2: Brain magnetic resonance imaging with contrast, sagittal view; multiple meningioma in various locations.

Another tumor was found in the right parasagittal region, which was evacuated and the tumor at the right side ridge region was approached under microscopic condition. The remaining tumors were small and were not excised. Following total debulking of the tumors, the dura base was coagulated. Repair of dura was followed by pericranium graft, hemostasis, flap fixation, and repair of skin.

The patient was evaluated postoperatively with an axial brain CT scan that showed no sign of new pathologic lesion or hematoma (Figure 3).

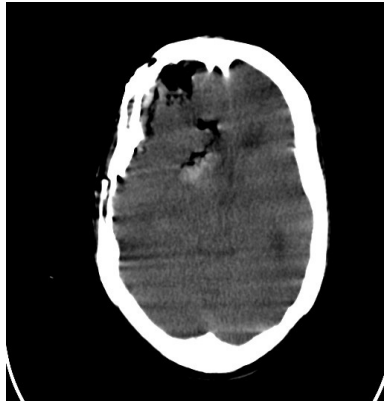


Figure 3: post-operative brain computed tomography scan after removal of meningioma in right frontal convexity, parasagittal, and right sphenoidal ridge.

Postoperative order included dexamethasone intravenous (IV) 8 mg q8h, cefazolin IV 1 mg q6h, gentamycin IV 80 mg TDS, and methadone subcutaneous/intramuscular on demand. The patient was admitted to the intensive care unit after the operation. In clinical examination, she had mild left hemiparesis post-operation (4/5 motor force in upper and lower limbs in the left side).

Pathological examination of the excised supratentorial tumor specimen revealed a meningothelial tumor composed of spindle cells with round to oval vesicular nuclei and intranuclear pseudo-inclusion bodies surrounded by eosinophilic cytoplasm with whirling pattern. No glial tissue was seen and no sign of mitosis or necrosis was present. In addition, Scattered Psammoma bodies were observed. The findings were suggestive of conventional type meningioma.

After a 3-year follow-up, the intact meningiomas had no change in size and contour. The patient developed no new neurological deficit or symptoms, except occasional headaches and her seizures were completely controlled with phenytoin. Left hemiparesis was improved after one year.

Discussion

Although meningioma is a relatively prevalent tumor, true meningiomatosis is not that common and is seen only in 2% of surgeries involving meningiomas. However, its prevalence is reported to

be higher in autopsies, comprising 9% of all meningiomas (16,17). The pathogenic and etiologic factors associated with the development of multiple meningiomas are not clearly known yet. However, it is well established that the incidence of solitary meningioma is higher in females, which could be attributed to the hormonal factors, especially the excessive amounts of progesterone (17,18).

According to many reports that indicate the higher incidence of multiple meningioma in women, a higher preponderance of female gender is observed in multiple meningioma cases with a male: female ratio of about 1:9 (17). Furthermore, the recent studies indicated that progesterone receptors are found to have higher expression rate in multiple meningioma cases, compared to the solitary ones (17). Therefore, hormonal therapy seems to be a valid and promising treatment in these cases.

Although meningiomas are benign by nature and do not disseminate through venous or lymphatic systems, spread of the cells via the meninges or cerebrospinal fluid (CSF) is thought to be the underlying mechanism for formation of multiple tumors. Molecular analyses of the tumors in several studies demonstrated the same chromosomal or genetic disturbances in all meningioma clones (19). On the other hand, there is another hypothesis stating that not all neoplastic lesions come from a single locus in a patient with multiple meningiomas. Consequently, each of the tumors grows independently under the influence of oncogene factors (16).

According to the reported cases of multiple meningioma, rapid progression of the tumor is an unlikely event. Most studies have reported slow-growing foci of tumor with the patient being in good condition, which is in line with our case who only complained of headache and had no complication. It is generally accepted that asymptomatic meningiomas should only be followed-up with imaging techniques. However, rapidly progressing tumors need urgent treatment (20).

In the presented case, meningiomas were located in both infratentorial and supratentorial regions with the majority being located in the latter. Larger tumors, which were excised, were located mostly in the convexity and parasagittal regions. In our case, the number of intracranial meningiomas (12 meningiomas) is in the highest rate when compared with other literatures. This is somewhat similar to the previous reports by Huang and Dumenicucci who reported both the cerebral and cerebellar convexity, as well as parasagittal site as the main locations of the meningiomas (17,21).

This might be associated with the origin of the tumors, supporting the hypothesis that multiple foci of the tumors in multiple meningioma

patients are disseminated through CSF flow and tend to involve the convexity region of the brain. Moreover, almost all of the tumor foci in our patient were located in one hemisphere of the brain. This is consistent with the findings of previous reports indicating that all the tumors were located in one intracranial space (16).

Management can be very difficult in patients with multiple meningioma. A brain MRI is usually enough for preoperative diagnosis. However, a brain CT angiography might be requested to determine the accurate location of the tumor foci and their relations with other intracranial components, namely sinuses, arteries, and veins (18).

Treatment with progesterone antagonists can be a valid option in management of these patients, especially those with higher expression of progesterone receptors (22). In addition, Complete surgical excision of all tumor foci would be an ideal option. Nevertheless, it might not be feasible in all patients due to the size and location of the tumor foci. In the current case, we only excised the large masses that were responsible for the major abnormalities in imaging studies.

Conclusion

In conclusion, unlike the solitary meningiomas, multiple meningiomas are not frequently encountered. However, they do not differ from solitary tumors in terms of prognosis and management, except that complete removal of the tumors is not possible in all cases. As a result, resection of only the largest tumor foci is performed, which was shown to result in favorable outcomes.

Conflict of Interest

The authors declare no conflict of interest.

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