Pulmonary Meningeomatosis - A Case of an Occult Dissemination

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Abstract

Introduction: This paper aims to report a case of a female patient with a recurrent primary tumor and immunohistochemically proved multiple metastases of atypical meningioma in the lungs.

Case Outline: A 67-year-old female presented with sudden and rapid vision loss and severe tremor. A neuro magnetic resonance revealed a mass lesion at the occipital and superior sagittal sinus 11 x 6 x 5 cm with interhemispheric propagation, massive perifocal edema, compression of venous circulation and surrounding brain structures. A standard chest X-ray revealed multiple space occupying lesions in both lungs and chest multislice computed tomography showed that the largest mass (8 x 4 cm) was positioned in the right lung without mediastinal lymphadenopathy. After video-assisted thoracoscopy procedure and immunohistochemistry, the patient was diagnosed with recurrent primary meningioma and pulmonary metastatic atypical meningioma. Due to the patient’s poor general condition and inoperable multiple pulmonary nodes, palliative radiotherapy was ordained.

Conclusions: Extracranial metastases of meningiomas should be considered in the cases of malignant or recurrent primary meningioma. Consistent follow-ups along with examinations of the chest, abdomen and bone are necessary in order to prevent and timely treat metastatic meningioma.

Introduction

Meningiomas are among the most frequent tumors affecting the central nervous system in adults (over 30% of all intracranial tumors). The annual incidence of meningiomas is 6 per 100,000 population and they occur two-fold higher in females with the highest incidence being observed after the fifth decade of life [1-7]. Meningiomas usually manifest as non-malignant solitary intracranial neoplasms located at the skull base or over the convexity of the brain and affixed to the inner layer of the dura mater [8,9]. It is possible to categorize them based on their dural site of origin, the involvement of surrounding tissues and their histological grading [8,9]. More precisely, the World Health Organization (WHO) makes a distinction between three different types of meningiomas based on tumor differentiation and/or mitotic activity: benign (WHO Grade I), atypical (WHO Grade II) and anaplastic/malignant (WHO Grade III) [9]. There are 15 histological subtypes that are differentiated depending on

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the common morphological appearances and the mitotic index that is assessed by the number of mitosis per 10 high power microscopic fields (HPF): 9 subtypes have been classified as WHO Grade I, while WHO Grade II and WHO Grade III contain 3 subtypes each (Table 1) [10]. Multiple meningiomas are present in up to 10% of cases.

<table>
<thead>
<tr>
<th>WHO grades</th>
<th>Subtypes</th>
</tr>
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<tbody>
<tr>
<td>I</td>
<td>meningothelial, microcystic, fibroblastic, secretory, transitional, lymphoplasmacyte-rich, psammomatous, metaplastic, angiomatous, clear cell (spinal)</td>
</tr>
<tr>
<td>II</td>
<td>chordoid, clear cell (intracranial), atypical</td>
</tr>
<tr>
<td>III</td>
<td>papillary, rhabdoid, anaplastic</td>
</tr>
</tbody>
</table>

Table 1: Histological subtypes of meningiomas.

Cancer that metastasizes to the lungs is condition that poses a sever threat to human life and it develops when primary cancer in another area of the body spreads to the lung [4,5]. The lungs are the second most frequent site of metastases stemming from extrathoracic malignancies. Lung metastases are quite common, occurring in 20 to 55% of advanced cancers [4, 5]. While almost any cancer can spread to the lungs, some are more likely than others to do so. The most common types of cancer that metastasize to the lungs include: bladder cancer, breast cancer, colon cancer, kidney cancer, melanoma, ovarian cancer, pancreatic cancer, prostate cancer, etc [4,5].

Metastatic meningiomas are very rare and the overall reported incidence is between 0.1% and 0.2% [11-13]. Moreover, previous studies have showed that the metastatic rate of atypical meningioma is 5% [11-13]. Although very rare, distant metastases most frequently appear in the lungs, followed by the liver, lymph nodes and bones [14-16]. Pale clinical picture may obscure long-term dissemination, and loaded radiographic finding may further complicate the diagnosis in these cases.

Case report

In April 2018, a 67-year-old female was admitted to University Hospital Medical Center Bezanijska kosa, Belgrade, Serbia and presented with sudden and rapid vision loss and severe tremor. A neuro magnetic resonance imaging (NMRI) revealed a mass lesion at the occipital and superior sagittal sinus 11 x 6 x 5 cm with interhemispheric propagation, massive perifocal edema and compression of venous circulation and surrounding brain structures. Moreover, intracranial hemorrhage was not observed. Meanwhile, the patient complained of fatigue, dyspnea and chest pain. A standard chest X-ray revealed multiple space occupying lesions in both lungs (Figure 1) and chest multislice computed tomography (MSCT) showed that the largest mass (8 x 4 cm) was positioned in the right lung without mediastinal lymphadenopathy (Figure 1).

The patient’s medical history revealed that in 2003 the patient had been diagnosed with meningioma in the occipital region. Even though it had been completely surgically resected, the tumor relapsed in the same region in 2009, 2012 and 2015. The patient underwent surgical resection and Gamma Knife radiation. During the period between 2003-2018 no intra- or extracranial distant metastases of primary tumor were diagnosed or treated. Therefore, based on the medical history, along with the clinical and radiographic findings, the patient was diagnosed with recurrent meningioma. Percutaneous needle biopsy of the selected lesion in the right lung was conducted for further histopathological investigation. A histopathology report of the examined tissue confirmed distant pulmonary metastases of the atypical meningioma (WHO Grade II). Additional bronchoscopy was negative. Due to poor general condition of the patient and the presence of multiple metastatic inoperable tumors in the lungs, the doctor’s committee decided to treat the patient with antiedematous therapy and palliative radiotherapy of the endocranium (total dose of 15 Gy in 3 session).

One month after the initial hospitalization the general
condition of the patient improved and she was referred to the Clinic for Chest Surgery, Military Medical Academy in Belgrade for additional surgical diagnostic procedures of the pulmonary changes by means of video-assisted thoracoscopy (VATS). VATS was conducted under general anesthesia by experienced thoracic surgeons. Thereby, two nodes in the right lung were completely resected and used for further immunohistochemical analysis. The list of tumor markers and stains analyzed by immunohistochemistry was presented in Table 2.

<table>
<thead>
<tr>
<th>Analyzed markers</th>
<th>Finding</th>
</tr>
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<tbody>
<tr>
<td>Cytokeratine AE1/AE5</td>
<td>Negative</td>
</tr>
<tr>
<td>Carletin</td>
<td>Negative</td>
</tr>
<tr>
<td>Chromogranin A</td>
<td>Negative</td>
</tr>
<tr>
<td>CD 34</td>
<td>Negative</td>
</tr>
<tr>
<td>CD 56</td>
<td>Positive</td>
</tr>
<tr>
<td>CD 57</td>
<td>Negative</td>
</tr>
<tr>
<td>Epithelial membrane antigen</td>
<td>Positive</td>
</tr>
<tr>
<td>Estrogen receptor</td>
<td>Negative (Score 0)</td>
</tr>
<tr>
<td>Ki-67</td>
<td>Positive in 26%</td>
</tr>
<tr>
<td>Progesterone</td>
<td>Positive in 80% (Score 7)</td>
</tr>
<tr>
<td>S-100</td>
<td>Positive</td>
</tr>
<tr>
<td>Synaptophysin</td>
<td>Negative</td>
</tr>
<tr>
<td>Vimentin</td>
<td>Positive</td>
</tr>
</tbody>
</table>

Table 2: The list of tumor markers and stains analyzed by immunohistochemistry.

In summary, the tissue samples were positive for vimentin, epithelial membrane antigen, S-100, CD 56 and progesterone with a total Allred score 7 (Figure 2).

Additionally, Ki-67 labeling index was 26%. A comparative analysis of histopathological specimens from primary meningioma diagnosed in 2003 and metastatic pulmonary atypical meningioma diagnosed in 2018 showed the same morphological changes (Figure 2). All these findings were highly confirmative for the histopathological characteristics of atypical meningioma (WHO Grade II) in the analyzed specimens.

**Discussion**

Meningiomas are typically non-malignant tumors which develop slowly and carry a bright prognosis in many cases [6]. Mahmood et al. [18] reported that out of 319 primary intracranial meningiomas that were surgically treated at Henry Ford Hospital (Detroit, Michigan, USA) in the period from 1976-1990, 92% were benign, while 6.26% were atypical meningiomas and 1.7% were malignant.

Surgical treatment is the method of choice when it comes to symptomatic meningiomas as it is usually necessary to relieve the neurological symptoms [19]. When possible, complete surgical excision of primary tumor is highly recommended because it often proves to be curative. Nevertheless, total resection often turns out to be unfeasible. Regarding recurrent tumors or meningiomas that were incompletely resected, radiotherapy, either conventional or stereotactic, is a suitable replacement [19].

The present standard for treating WHO Grade III lesions is a combination of utterly safe gross total resection (GTR) followed by radiotherapy, whereas radiotherapy is not obligatory when it comes to WHO Grade II lesions [20]. Radiosurgery has been proposed as a replacement for conventional surgical treatments and radiotherapy has been suggested for patients who have been diagnosed with intracranial tumors [21]. Previous studies have reported that meningiomas classified as WHO Grade II and treated by means of GTR and radiotherapy showed 5-year progression-free survival rates of 74%-100%, whereas meningiomas categorized as WHO Grade III showed 5-year overall survival rates of 47%-61% when the same therapy was administered [22]. Also, it has been reported that traditional chemotherapeutic agents were not quite successful in treating meningiomas [19].
According to Lee et al. [16], even once complete resection has been performed, the local recurrence rates of meningioma range from 9 to 32%. WHO Grade II and III meningiomas are likely to behave more aggressively and there is a considerably higher risk of their recurrence (29% - 52% WHO II and 50% - 94% WHO III) [6]. Hortobágyi et al. [23] stated that higher histological grades, higher proliferative index, incomplete resection, tumor size (> 50 mm), tumor localization (skull base), irradiation and peritumoral edema are the most significant predictive factors of meningioma recurrence.

Metastatic meningiomas are uncommon and their overall reported incidence is between 0.1% and 0.2%, whereas the incidence of distant metastases is approximately 5% and 43%, respectively [11-13]. In 2015, Frydrychowicz et al. [24] conducted a literature review related to pulmonary metastases of primary meningioma. They reported that the age of the published patients ranged from 17–84 years (mean 53 years); 53% of the patients were women and 47% were men. Also, all published cases underwent craniotomy for the purpose of intracranial meningioma resection and nearly all of the reported meningiomas recurred at least once after the initial surgery.

They have showed that most reported meningiomas involved late pulmonary metastases with their occurrence varying between 2 months and 26 years following intracranial surgery, whereas there were four reported cases of the coincidence of intracranial meningioma and lung metastases [24].

The authors concluded that a high incidence of metastatic spread is directly associated with increasing meningioma malignancy, craniotomy and recurrent primary tumor. Also, they stated that venous sinus invasion could be a frequent mechanism that contributes to a spread of tumor cells [24].

Primary therapy for pulmonary metastatic meningiomas should encompass tumor resection performed by thoracotomy and followed by focal radiation [24]. In our case, the reported patient suffered from recurrent primary tumor for the fourth time.

The patient was treated with palliative radiotherapy due to her poor general condition. Moreover, taking into account the presence of multiple metastatic nodes in both lungs, thoracotomy was not an option in this case.

**Conclusion**

In the case of malignant or recurrent meningioma, extracranial metastases should be taken into consideration. The latency period between the diagnosis of an intracranial tumor and the appearance of metastases varies to a high extent, (i.e. several months to over 20 years). Occult dissemination may lead to the rare picture of pulmonary meningeomatosis. Consistent follow-ups along with examinations of the chest, abdomen and bone are necessary, particularly so once accompanying signs or symptoms appear.

**References**


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