Benign Metastasizing Meningioma

C. S. Pramesh¹, Avanish P. Saklani¹, Gouri H. Pantvaidya¹, Anil A. Heroor¹, K. N. Naresh², Sanjay Sharma¹ and Ramakant K. Deshpande¹

Departments of ¹Surgical Oncology and ²Pathology, Division of Thoracic Surgery, Tata Memorial Hospital, Mumbai, India

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Meningiomas represent about one sixth of all primary neoplasms of the central nervous system. They rarely metastasize outside the intracranial compartment. There are no clear criteria to identify the subset of aggressive tumors which recur locally or metastasize. Histological tumor grade is the most important predictor of recurrence or metastases. We report an unusual case of recurrent metastasis in an extrapleural location and review the literature. Our patient developed recurrent thoracic metastases from an intracranial benign meningioma after a disease-free interval of 8 years. She was successfully managed by wide excision of the metastasis and is currently asymptomatic.

Key words: meningioma – benign – metastasis

INTRODUCTION

Meningiomas are benign neoplasms that arise from the intracranial and spinal meninges and their dural extensions. They constitute 14–19% of all central nervous system neoplasms. They are considered as benign neoplasms as they generally do not metastasize, are not invasive and are usually cured by surgical resection. However, they have a potential to become more aggressive and invade the brain and/or calvaria or to metastasize outside the central nervous system (CNS). The local recurrence rates even after complete resection vary from 9 to 32%. There are no definitive criteria to predict local recurrence or metastases in meningiomas. About one in 1000 meningiomas metastasize. In view of the slow-growing nature of these metastases and their good prognosis after resection, surgery is the treatment of choice.

CASE REPORT

A 29-year-old woman presented with unilateral right-sided headache of 18 months duration and blurred vision for 15 days. She also had an unstable gait and upper motor neuron facial nerve palsy. A computed tomography (CT) scan revealed an extradural tumor in the posterior cranial fossa (Fig. 1). Complete excision of the tumor through a right occipital craniotomy was done. The histopathology of the tumor was a benign psammomatous meningioma. The patient had complete remission of all symptoms and remained asymptomatic. Eight years later, she presented with a 1-month history of cough, chest pain and haemoptysis. She had no neurological complaints. A CT scan of the chest revealed an extrapleural mass in the left thoracic cavity. There was no mediastinal lymphadenopathy. An exploratory thoracotomy with excision of an 18 × 16 × 8 cm mass with the fourth and fifth ribs was performed. Histopathology revealed a psammomatous meningioma, metastasizing from the intracranial tumor. One year later she again developed chest pain. A plain radiograph of the chest revealed two well defined opacities in the right mid zone with convex inner margins abutting the right lateral chest wall, suggestive of an extrapleural mass. A CT scan of the thorax revealed a large mass in the right thoracic cavity suggestive of a recurrence (Fig. 2). A routine metastatic workup revealed no evidence of tumor elsewhere. We excised the mass along with the sixth and seventh ribs and the adjoining segment of the lung. We performed a primary closure of the chest wall with the overlying muscles for cover. The patient had no postoperative complications. Histopathology showed spindle meningothelial cells arranged in fascicles infiltrating the lung (Fig. 3). Immunohistochemistry showed it to be vimentin and epithelial membrane antigen (EMA) positive and cytokeratin negative, suggestive of metastatic meningiomas. All the cut margins of the resection were free. We have given the patient postoperative radiotherapy. She is now asymptomatic 1 year after surgery.
DISCUSSION

Meningiomas constitute 14–19% of all primary neoplasms of the central nervous system. These are usually non-invasive, do not metastasize and hence are perceived as benign tumors. Radical surgery is the treatment of choice. Although they are typically slow growing, extra axial benign tumors, about 0.1% of meningiomas metastasize (1). The malignant nature of a meningioma is usually defined by its ability to metastasize and histologically by high cellularity, typical and atypical mitosis, necrosis, poor differentiation and infiltration of the underlying brain (2). Nuclear pleomorphism is the only reliable criterion for predicting malignancy.

The differential diagnosis of meningiomas includes hemangiopericytoma, hemangioblastoma, solitary fibrous tumor, sarcoma and choroid neoplasms (3). Hemangiopericytomas which were previously grouped under meningothelial tumors have now been reclassified under mesenchymal, non-meningothelial tumors. They have the classical staghorn pattern at low magnification and a distinctive turbulent pattern at higher magnifications. Although they are enucleated surgically with ease, they are aggressive, have a high rate of local recurrence and tend to metastasize to distant sites. The new World Health Organization (WHO) classification of tumors of the nervous system lists 15 histopathological variants of meningioma (3). Tumors of meningothelial cells are subdivided into benign meningiomas of WHO grade I (including eight different histopathological subtypes and metaplastic variants), atypical meningiomas of WHO grade II, anaplastic meningiomas of WHO grade III and rare meningioma subtypes of WHO grade II or III (3). Atypical meningiomas have a mitotic rate of four or more mitoses per 10 HPFs, have increased cellularity, small-cell population with high nucleus:cytoplasmic ratio, prominent nucleoli, patternless growth and foci of spontaneous or geographic necrosis. They are associated with a significantly higher rate of recurrence (3).

Tumor grade is the most important histological property to determine the likelihood of recurrence. The proliferation associated antigen Ki-67 (MIB-1) is an index of mitoses and labeling indices above 5–10% suggest a higher chance of recurrence. However, the variability of this reaction between laboratories pre-empts the use of an increased MIB-1 index as a single diagnostic criterion for a WHO grade II (3). Meningiomas, like leiomyomas, may metastasize despite a benign appearance and the metastasis itself may also be benign. Extracranial metastases in cases of meningioma are, however, rare. Only anecdotal case reports of extracranial metastatic meningiomas have been reported in the medical literature. The common sites of metastasis in order of occurrence are the lung, liver, lymph nodes and bone.

Ectopic meningiomas are very rare and four mechanisms have been suggested for their occurrence: (a) direct extension from an intracranial lesion, (b) distant metastasis from an intracranial meningioma, (c) origin from arachnoid cells within the sheaths of cranial nerves and (d) origin from embryonic nests of arachnoid cells (4). The previous existence of a CNS neoplasm, the non-contiguous site of metastasis and the close histological resemblance point towards the second mechanism. The time interval from diagnosis of the primary to the occurr-

Figure 1. CT scan of brain showing intracranial meningioma.

Figure 2. CT scan of thorax showing a smooth hyperdense lesion in the right hemithorax.

Figure 3. Photomicrograph showing spindle-shaped meningothelial cells arranged in fascicles (H&E stain, original magnification ×10).
rence of the metastasis after complete control of the primary is variable and ranges from predating the primary tumor to 19 years after treatment of the primary (5).

Meningiomas are generally slow growing and have a good prognosis. In addition, the long DFI, the absence of other sites of recurrence and the resectability of the metastasis prompted us to perform a second metastatectomy in our patient. Regular follow-up is necessary to assess the long-term results and survival in such cases. Radiotherapy, when used as a primary modality of treatment, is ineffective in meningiomas. However, when used as an adjuvant, it decreases local recurrence and time to recurrence after complete or subtotal resection (6). Although not for meningiomas, it has been conclusively proven that resection of solitary metastasis in a well-controlled primary improves survival.

Survival rates after diagnosis are highly variable and range from 10 days to 24 years (7). Except for radiotherapy, the role of other adjuvant modalities remains under trial. The role of ifosfamide is currently being investigated. About 80–90% of meningiomas are positive for progesterone receptors (8). This may explain their predominance in women. It also raises the possibility of using endocrine manipulation as an adjuvant to prevent recurrence. The role of mifepristone, an antiprogesterone agent, which has shown some response, is under study.

CONCLUSION

Meningiomas are generally slow-growing benign tumors. They rarely become more aggressive and recur locally or metastasize. If the metastasis is solitary and the disease-free interval is long, surgical resection of the metastasis is recommended in an effort to improve survival.

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References