Recurrent Intracranial Meningioma with Malignant Change and Extracranial Bone Metastasis: A Case Report

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In general, meningiomas are slowly growing benign neoplasms originating from specialized meningothelial cells in arachnoid granulation, but have a tendency to be locally invasive and recurrent. Meningiomas very rarely metastasize outside the nervous system, occurring in less than 0.1%.

We report the CT and MR findings of a case of a solitary benign syncytial meningioma showing recurrent multiple tumors and malignant progression with eventual bone metastasis to rib after six surgical extirpations during six years.

Index Words: Meninges, neoplasms
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Less than 9% of intracranial meningiomas represent multiple lesions and only 1 - 2% are anaplastic or malignant(1, 2). It has been estimated that less than 0.1% of meningiomas metastasize, and both the benign and malignant meningiomas can metastasize to distant sites(3, 4).

This is a case of multiple and malignant intracranial meningiomas metastasizing to rib, which progressed from a solitary benign syncytial one after six times of surgical extirpation during six years.

CASE REPORT

A 48-year-old man presented with several-month history of progressive right hemiparesis and dull headache. CT revealed a large lobular mass of 9 cm in maximal diameter in the left high parasagittal region abutting the falx. The mass was hyperdense and diffusely enhanced with a small area of central low density(Fig. 1a). He underwent his first operation for total removal of the mass and it was found to be a well encapsulated hard mass tightly adherent to the left side of falx and superior sagittal sinus. No evidence of underlying brain parenchymal invasion was noted. Pathologic diagnosis was benign syncytial meningioma(Fig. 1b).

One year and 8 months later, the second operation was performed to remove a recurrent tumor in the same site. Pathologic diagnosis was atypical meningioma. Two more operations were done during the next 3 years to remove the recurrent histologically proven atypical meningiomas.

One year after the 4th operation, the patient was readmitted because of aggravating right hemiparesis. Follow-up CT demonstrated a recurrent cystic mass with two large enhancing nodules, but there was no enhancement in the cyst wall. On MR(Fig. 2a - c), the cyst fluid was hyperintense on both T1-weighted (T1WI) and T2-weighted images (T2WI) suggesting lysed chronic hematoma, and the wall was isointense on T1WI and hypointense on T2WI suggesting to be laden with hemosiderin. The solid nodules showed isosignal intensity on T1WI and slightly high signal intensity on T2WI, and rather heterogeneous contrast enhancement was seen. There was another small flat extraaxial solid mass at right lower frontotemporal convexity having signal characteristics similar to the left parasagittal nodules. The fifth operation was performed to remove the recurrent left parasagittal mass. The cyst fluid was shown to be liquefied darkish hematoma surrounded by a yellowish capsule and underlying brain parenchyma was invaded by the tumor. A histologic examination demonstrated malignant meningioma with atypical meningothelial cells, cellular pleomorphism, frequent mitoses, focal necrosis and invasion of brain, and the capsule contained a lot of hemosiderin (Fig. 2d).

One year later, he was readmitted for tonic-clonic...
seizure and paraparesis. A follow-up CT demonstrated a recurrent tumor invading and penetrating the falx. He underwent his 6th operation to remove the recurrent malignant meningioma involving bilateral parasagittal regions.

One year after the last operation, a follow-up CT revealed local recurrence in the left high parietal region, more enlargement of the right frontotemporal convexity mass with calcifications, and two new growths were seen abutting the superior border of the right tentorium and the left border of anterior falx (Fig. 3). Chest CT revealed the left 4th rib destruction with a heterogeneously enhancing bulky soft tissue mass and a histologic examination demonstrated metastatic malignant meningioma (Fig. 4).

Despite 6 surgical extirpations of the recurrent meningiomas progressing from a typical benign one to an atypical and malignant one, the patient died about 8 years after the initial presentation.

**DISCUSSION**

Although meningiomas usually are considered to be benign and cause compression of adjacent neural tissue, there are cases in which they show local invasion to dura, dural sinus, bone, muscle and brain substance. And local tumor recurrence has been reported to occur in 9% to 32% of cases depending on the follow-up periods (5, 6). Reports suggest that recurrence depends on the incompleteness of tumor excision. It is well known that such local invasion and recurrence are not necessarily indicative of malignancy, although malignant meningiomas account for much higher incidence of local invasion and recurrence.

Of the meningiomas, 1–2% are malignant (2). Histologic criteria for malignant meningiomas remain at least partly uncertain. Greater than 10 mitoses per 10 high-power fields and atypical features are the most reliable indicator of malignant behavior, but that measure is also imperfect. Rapid rate of recurrence, aggressive local invasiveness and the presence of remote metastases are also suggested for determining the criteria of malignancy. But as described previously, the former two have a variable and significant margin of error in predicting malignant biologic behavior. Di Chiro et al. (7) demonstrated that positron emission tomography with fluorine-18-2-fluorodeoxyglucose is reliable for predicting the biologic behavior and recurrence of meningiomas by assessing tumoral glucose utilization.

As in our case, parasagittal meningiomas are known to have greater probability of recurrence and malignant change than those in other locations. Reported CT findings favoring malignancy of meningioma include extensive bone destruction, irregular indistinct tumor margins, deeply penetrating fronds, extensive necrosis, absent or minimal calcification and mushrooming pannus of tumor extending from the globoid part of the tumor. But there are few reports about MR findings of malignant meningiomas (8). MRI of our case showed extratumoral cystic pattern of malignant meningioma after 4 surgeries, and the cyst resulted from lysed chronic hematoma. Cystic meningiomas have two different morphologies: intratumoral and extratumoral cysts. Intratumoral cysts representing tumor necrosis or degeneration show rim enhancement. On the other hand, extratumoral cysts representing reactive arachnoid cyst, direct secretion of fluid by tumor cells, absorption of hemorrhage as in our case, or loculated cerebrospinal fluid show no rim enhancement (9).
Fig. 2. Recurrent cystic meningioma with malignant progression after 4 times of surgical extirpation.

a, b. The cystic component is hyperintense on both T1WI(a, TR/TE=650/25) and T2WI(b, TR/TE=2300/520) suggesting chronic lysed hematoma and the wall(arrows) is hypointense on T2WI(b) suggesting hemosiderin deposit.

c. Post-contrast T1WI shows diffuse enhancement of the masses.

d. Microscopic features show findings of malignant meningioma with sheets of atypical meningotheial cells, cellular pleomorphism, hyperchromatism and frequent mitoses(H&E, X400).

Fig. 3. Recurrent and multiple meningiomas after 6 times of surgical extirpation.

a. Post-contrast CT shows a large recurrent mass in left high parietal region and another small mass is seen abutting left anterior falx.

b. Lower level scan shows two more tumors at right frontotemporal convexity and upper border of right tentorium. Note calcifications in right frontotemporal lesion(arrows)
Fig. 4. Metastatic meningioma.

Post-contrast CT of chest shows destruction of left posterior 4th rib with a heterogeneously enhancing bulky soft tissue mass.

Multiple meningiomas occur in less than 9% of imaged cases and are known to have an identical histologic type in each case(1). Although multiple meningiomas can be associated with neurofibromatosis type 2, the majority of patients have no relation with it. Our patient also showed no other evidences of neurofibromatosis. The loss of genetic material in chromosome 22 has been proposed as a cause of multiple meningiomas.

Distant metastasis of intracranial meningiomas is extremely rare and is said to arise in less than one in 1000(3, 4, 11). The malignancy of a tumor is usually defined by its ultimate biologic behavior, that is, by its propensity to metastasize. But not only the malignant but also the atypical and benign meningiomas have been reported to metastasize via blood stream and cerebrospinal fluid pathway(3, 4, 8, 11). Those benign metastasizing meningiomas could be due to highly proliferative activity of the neoplastic cells. Most common sites of metastases are lung, liver and lymph nodes. The hematogenous spread via the caval circulation following invasion of dural sinuses probably explains the high incidence of pulmonary metastasis(11). Bony metastasis as in our case has been reported to constitute about 20% of all metastases(3, 4). There has been controversy about the relationship between metastatic disease and surgery. Although about 75% of reported cases metastasized after variable number of surgical extirpations, no particular relationship is known to be present between them(12).

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