



Recurrent Intracranial Meningeal Hemangiopericytoma with an Isolated Bone Metastasis: A Case Report and Literature Review

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SUMMARY

Intracranial meningeal hemangiopericytoma is a rare vascular tumor with a high rate of both local recurrence and the development of distant metastases. The aim of this report is to present the first case of recurrent meningeal hemangiopericytoma with an isolated femur metastasis and discuss the clinical features and treatment of this rare tumor. A 46-year-old woman was admitted with the complaint of sudden loss of consciousness and subsequently underwent an urgent craniotomy with total excision of the lesion. The diagnosis was meningeal hemangiopericytoma. The patient presented with a painful left hip 5½ years after the initial surgery for the primary tumor. A lytic lesion, which was 4x4.5 cm in diameter, was seen on an X-ray of the proximal left femur. The patient underwent lesion removal surgery and pathological diagnosis was compatible with meningeal hemangiopericytoma. At 69th month, no progression has been observed in follow-up. Long-term follow-up is mandatory for meningeal hemangiopericytoma due to high recurrence rate many years after diagnosis. Further multi-institutional studies are strongly needed to understand the clinical behavior and treatment modalities of this rare disease.

Keywords: Meningeal hemangiopericytoma; isolated metastasis; radiotherapy.

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Introduction

Hemangiopericytomas are mesenchymal neoplasms originating from pericytes. Primary meningeal hemangiopericytomas are aggressive vascular tumors with a high rate of both local recurrence and the development of distant metastases.[1] They constitute approximately 2% of meningiomas and less than 1% of central nervous system (CNS) tumors.[2]

Most hemangiopericytomas are located in the

musculoskeletal system and the skin. They are more common in men than in women. Intracranial hemangiopericytomas are rare and are often located supratentorially.[3] They are clinically and radiologically similar to meningiomas, but they have a stronger tendency for both local recurrence and the development of extracranial metastases than meningiomas.

The rates of metastatic disease vary from 23.4%[1] to 64%. Metastases can be seen many years after removal of the primary tumor. The most common sites

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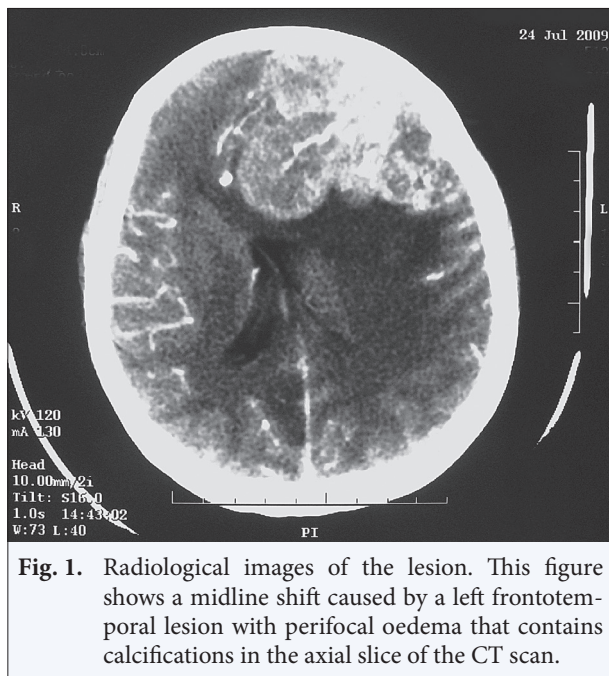
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of metastatic disease are the bone, liver, and lungs.[1] Most patients who develop extracranial metastases have multiple metastases in different sites.[4]

In this report, we present a rare case of an isolated metastatic recurrence of an intracranial meningioma 5.5 years after the first diagnosis. In the medical literature, there are six reported cases of intracranial hemangiopericytomas with an isolated bone metastasis, and our case is the seventh. However, our patient is the first case with an isolated femur metastasis. The aim of this report is to present a very rare case and discuss the clinical features and treatment of this tumor.

Case Report

A 46-year-old woman was admitted to the emergency room in July 2009 with a complaint of sudden loss of consciousness. Computed tomography scan (CT) of the brain revealed a midline shift caused by a left frontotemporal lesion with a perifocal oedema that contained calcifications (Figure 1). On the basis of this imaging, a diagnosis of a meningioma was suspected. The patient underwent an urgent craniotomy with a total excision of the lesion.

Upon histopathological examination, microscopically, it was discovered that the whole specimen was composed of a tumor with extensively branched, thin-walled vessels. The tumor cells, which had enlarged

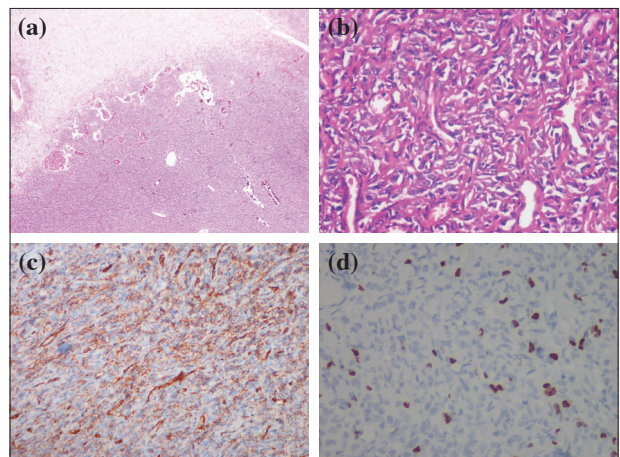


Fig. 2. Histopathological and immunohistochemical findings of the lesion. Panel a (HEX40) shows a panoramic view of the tumor with hemangiopericytomatous vasculature. Panel b (HEX400) shows moderate nuclear pleomorphism in the tumor cells around tiny branching vessels. Panel c (IHCx100) shows CD34 positivity in the tumor cells. Panel d (IHCx200) shows a high proliferation rate with Ki-67.

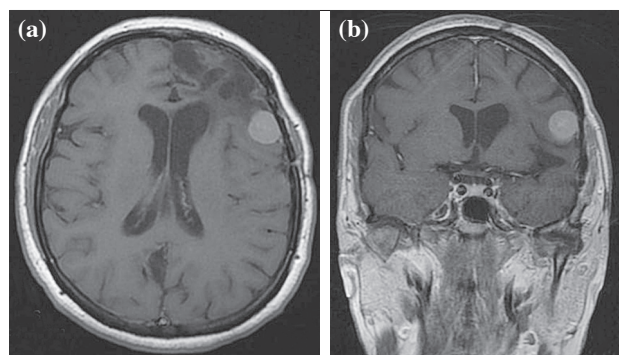


Fig. 3. MRI findings of the recurred lesion, which is 20 mm in diameter, next to the previous tumor site. Panel a shows an axial view of the initial tumor bed and the recurred lesion in the left frontal lobe. Panel b shows a coronal view of the same lesion.

nuclei with moderate pleomorphism and scanty cytoplasm, were arranged as nested solid sheets around the vessels. Immunohistochemically, the tumor cells were positive for vimentin, CD34, and Bcl-2, but the cells were negative for CD31, desmin, pancytokeratin, S100 protein, Factor VIII, smooth muscle actin, and alpha epithelial membrane antigen. Ki-67 showed a proliferation in around 10% of tumor cells. The diagnosis of WHO Grade II hemangiopericytoma was accepted in accordance with these findings (Figure 2).

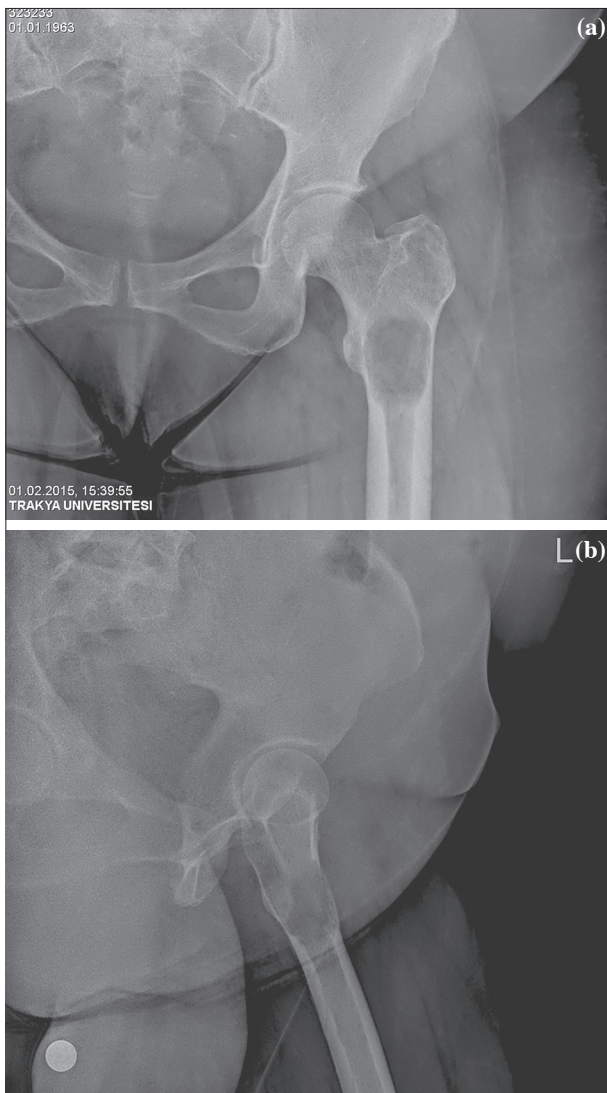


Fig. 4. X-ray images of the lytic metastasis on the femur neck on the level of the trochanter minor. The lesion destructed over 50% of bone diaphyseal cortices. Panel a shows an antero-posterior view. Panel b shows an oblique view of the left femur and the lesion.

No further treatment was offered by neurosurgeons and the patient attended follow-up visits. Radiological assessments with magnetic resonance imaging (MRI) of her brain showed no residual or recurrent tumor, and her neurological examinations were normal until 44 months after surgery.

In April 2013, the patient was asymptomatic, but a cranial MRI showed a lesion, which was 20 mm in diameter, in the left frontal lobe near the previous tumor site (Figure 3). Because of the local recurrence, the

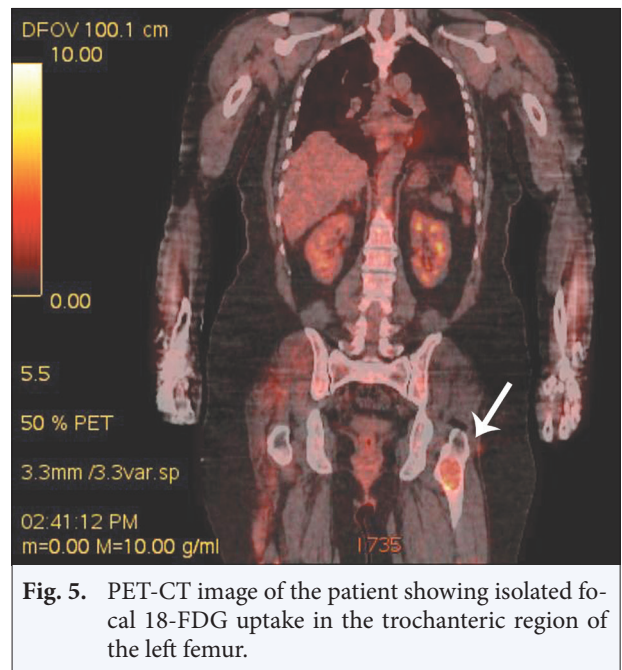


Fig. 5. PET-CT image of the patient showing isolated focal 18-FDG uptake in the trochanteric region of the left femur.

patient underwent another operation. The pathological diagnosis was compatible with the previous pathology (WHO Grade II hemangiopericytoma). The patient was referred to our clinic for adjuvant radiotherapy. There was no sign of a residual tumor on her cranial MRI, and 60 Gy (2 Gy/fraction daily and 5 fractions per week) of radiotherapy was postoperatively administered to the tumor bed.

During the first year after radiotherapy, the patient had no neurological symptoms, and radiologic follow-up was normal. One year after radiotherapy, she presented with a painful left hip. A lytic lesion, which was 4x4.5 cm in diameter, was seen on an X-ray of the proximal left femur (Figure 4). A PET-CT performed for restaging and to find other possible metastatic lesions showed isolated focal 18-FDG uptake in the trochanteric region of the left femur on behalf of a malignant lesion that could have been metastatic or primary bone disease (Figure 5).

In February 2015, the patient underwent a resection of the left femur head, and prosthetic replacement surgery was performed. The pathological diagnosis was compatible with metastatic meningeal hemangiopericytoma (Figure 6). She was postoperatively given 30 Gy of radiotherapy for her metastatic lesion. The patient is attending follow-up visits. She is alive and well at 69 months after the initial diagnosis, and there has been no further progression of her disease.

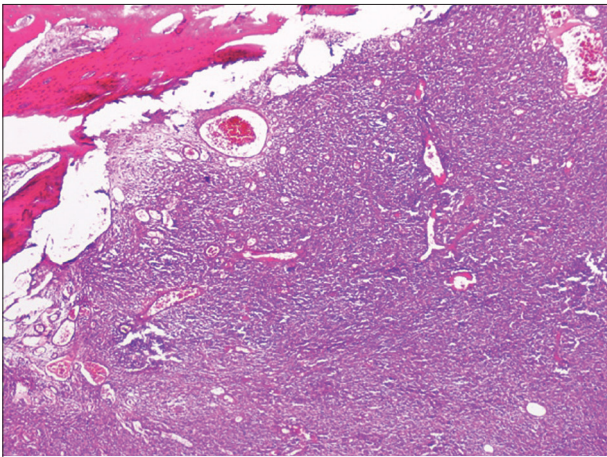


Fig. 6. (HEEx40) Bone invasion of hemangiopericytoma (upper left).

Written informed consent was obtained from the patient who participated in this study.

Discussion

Hemangiopericytomas are rare vascular tumors arising from pericytes. They were first described by Murray and Stout in 1942.[3] Hemangiopericytomas mostly occur in the skin, retroperitoneal spaces, and the soft tissue of the lower extremities. However, they can rarely occur in the larynx, bones, spleen, and CNS as a primary tumor.[5] Intracranial meningeal hemangiopericytomas are uncommon. They account for approximately 2% of meningiomas and less than 1% of CNS tumors.[2]

Surgery is the mainstay of treatment for meningeal hemangiopericytomas. For unfavorably located tumors, conservative surgery with postoperative radiotherapy has been suggested. A dose of 50–60 Gy of radiotherapy is suggested to improve local control. The presented patient was treated and followed in surgical clinic after primary surgery. Since she was referred to our clinic after recurrence surgery, she has not received any radiotherapy. They have a strong tendency to recur locally and to develop distant metastases. Mena et al. reported that 57 patients developed local recurrence (60.6%) and 22 patients developed single or multiple metastases (23.4%) in their review, which consisted of 94 cases of CNS hemangiopericytoma.[1] The most common sites for metastases were the bone, liver, and lungs.[1]

In the literature review, we used PubMed to identify cases of bone metastases originating from intracranial meningeal hemangiopericytoma. The key words

“hemangiopericytoma”, “cranial”, “meningeal”, “bone”, “metastasis”, “radiotherapy”, “surgery” were searched and cross-referenced. Articles including metastatic patients without bone metastasis were excluded. Twenty english-language articles were found, including 28 described cases of bone metastasis originating from an intracranial meningeal hemangiopericytoma. In three articles, the sites of the bone metastasis were not noted. Table 1 presents a summary of the reported cases. Of the 28 cases, 17 had bone metastasis, with the vertebra (61%) being the most frequently involved site. Bone metastasis involved the pelvic bones in nine patients (32%), the femur in seven patients (25%), the ribs in four patients (14%), the humerus in two patients (7%), and the scapula in two patients (7%). There are six reported cases with an isolated bone metastasis throughout the skeleton, but there are no cases with an isolated bone metastasis in the femur. Our patient is the first case with an isolated femur metastasis that originated from an intracranial meningeal hemangiopericytoma.

A bone metastasis can be seen as a lytic lesion with a sclerotic margin on plain roentgen film. On plain roentgen film and CT scans, the involved bones could show thinning and expansion.[6] The role of bone scans is limited for hemangiopericytomas because lesions may not be detected due to the lack of radionuclide isotope uptake of the metastases.

Metastases typically occur 63–99 months after the initial diagnosis of hemangiopericytomas. When a single bone metastasis is determined, synchronous multiple metastases can be observed in bones or in different organs, such as the lungs, liver, pancreas, and kidneys. The median survival rates of patients with metastases are paradoxically longer than expected. One study reported that the median survival rates of patients with and without systemic metastases were 202 months and 99 months, respectively.[7] High rates of local recurrences, the development of distant metastases, the occurrence of metastases a long period after the initial surgery, and the appearance of synchronous metastases are characteristic features of this tumor.[8]

Outcomes of chemotherapy for the management of metastatic disease, have been disappointing. The most frequent symptom at the time of diagnosis of bone metastases is pain. The total surgical excision of a bone metastasis may be beneficial for patients with limited distant disease, but the long-term benefits of this excision are unclear. Radiotherapy for bone metastases is effective for palliation. The most commonly used radiotherapy regimen is 30 Gy/10 Fr. The aim of radiotherapy for bone metastases is not only pain relief but also

Table 1 Summary of hemangiopericytoma cases with bone metastases originating from intracranial meningeal hemangiopericytoma

No.	Author (year)	Age (years)/Sex	Metastatic bone lesions	Other metastatic lesions
1	Kruse F Jr. (1961) [9]	22/F	Lumbar vertebra	None
2	Scott et al. (1974) [10]	38/M	T12/L1 vertebra, cervical vertebra, temporal bone	None
3	Thomas et al. (1981) [11]	69/M	Jaw	None
4	Itoh et al. (1984) [12]	49/F	Rib, cervical vertebra, sternum	Liver
5	Iwaki et al. (1988) [5]	54/F	Skull	None
		40/M	Vertebra	Lung, liver, kidney, pancreas, lymph nodes
		63/M	Rib	Lung
		67/M	Vertebra, rib	None
		58/F	Vertebra	Subarachnoid space
		ND	Ilium	Liver, lung
6	Younis et al. (1995) [13]	55/M	Cervical vertebra	Liver, lung, pancreas
7	Koyama et al. (1997) [14]	49/F	T8 vertebra, caput femoris, rib	Lung
8	Nonaka et al. (1998) [15]	49/F	Lumbar vertebra	Liver, lung
9	Someya et al. (2001) [2]	42/F	Bone (ND)	Brain, pleura
10	Prakasha et al. (2001) [8]	37/F	L5 vertebra, femur, multiple bony	None
11	Suzuki et al. (2002) [16]	49/M	Pubic bone	Pancreas
12	Woitzik et al. (2003) [17]	65/M	C6-T2, L2 vertebra	Liver
13	Chang et al. (2004) [18]	40/F	C2-C3 vertebra	Lung, liver, kidney
14	Hoshi et al. (2005) [6]	43/F	Scapula, sacrum, humerus, femur, cervical-thoracic vertebra, pelvis	Lung
		49/F	Pelvis, femur	Lung
		32/M	Pelvis, rib, thoracic vertebra, femur	Pancreas
		45/F	Pelvis	Lung, liver, kidney
		48/M	C6-T1 vertebra	None
15	Lee et al. (2006) [19]	55/F	Scapula	None
16	Siegel et al (2012) [20]	51/M	Sacral vertebra	None
17	Satayasoontorn et al. (2014) [21]	36/M	Sacroiliac bone	None
		40/M	Femur, lumbar vertebra	None
		66/M	Humerus, femur, iliac wing	None
		44/M	Femur head	None
18	Present case	46/F		None

ND: No description.

functional preservation and maintenance of a patient's quality of life. Besides, palliative radiotherapy is given as standart procedure to eradicate the microscopic tumor cells that may be left behind after metastasectomy. Consequently, 30 Gy (3 Gy/fraction daily and 5 fractions per week) of radiotherapy was given to metastasectomy area in concordance with the literature.

To the best of our knowledge, this report is the first reported case of an isolated femur metastasis that originated from an intracranial meningeal hemangiopericytoma 5.5 years after the initial surgery. The metastatic potential of this rare tumor should be considered. Long-term follow-up is mandatory due to the high recurrence rates many years after the surgical resection of the primary tumor. Since it is a rare disease, further multi-institutional studies are strongly needed to understand the clinical behavior and treatment modalities of this disease.

Disclosure Statement

The authors declare no conflicts of interest.

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