

Intracranial hemangiopericytoma with multiple extracranial metastases: a case report

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Meningeal hemangiopericytoma (HPC) is a rare tumor in the central nervous system. It is often reported as “Meningioma”. Unlike meningiomas, HPC has a high rate of local recurrence and distant metastasis, which may occur several years after initial treatment [1]. The purpose of this article is to report a case of parasagittal hemangiopericytoma with delayed multiple extracranial metastases and local recurrence. **Chiang Mai Medical Journal 2013;52(3-4):81-86.**

Keywords: intracranial hemangiopericytoma, multiple extracranial metastasis

Introduction

Intracranial hemangiopericytomas represent a very rare tumor of mesenchymal origin, which constitute approximately 1% of all intracranial tumors [2]. It is widely accepted that these tumors originate from Zimmerman’s pericytes. The first description of an extracranial hemangiopericytoma was made by Staut and Murray in 1942, with Begg and Garret being the first to report a primary intracranial meningeal hemangiopericytoma in 1954 [3, 4]. There was a long standing controversy regarding the histological classification, origin, and nomenclature of these tumors. Historically, they have been grouped with meningiomas, and the terminology of “meningeal” hemangiopericytoma has been confused with “vascular meningioma” and an-

gioblastic meningioma because of its radiologically and microscopically clinical resemblance to a meningioma. Although the intracranial hemangiopericytomas share macroscopic and imaging characteristics with meningiomas, their more aggressive behavior, tendency for early recurrence and extracranial metastasis as well as worst prognosis have been recognized. In 1993, the WHO separated hemangiopericytomas from meningiomas and classified them as separate histopathological entities. Hemangiopericytoma has a high rate of local recurrence and distant metastases, which may occur several years after the initial treatment. Mena *et al*, reported a 27% metastasis rate, and recurrence rate of 70% in their 94 patients with hemangiopericytoma, and the most common sites of metastasis were reported to be the bone, lung and liver [1].

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The rarity of these tumors, together with their controversial histologic classification makes their misdiagnosis quite frequent and treatment rather challenging.

Case report

A 48 years old man presented with focal seizure of his left extremities in July 2001. He had a history of a craniotomy to remove a left parietal brain tumor ten years previously. Brain computed tomography showed bilateral parasagittal meningeal tumors. He received tumor excision (Simons gr II) and his pathology was interpreted as meningioma. Two years later, he presented with recurrent right side focal seizures. Brain computed tomography showed a recurrent left parasagittal meningioma. Therefore, he received tumor excision again, and the pathologic report showed meningioma. Four months later, he presented with pathologic fracture of his right proximal humerus and he received a hemiarthroplasty of the right shoulder. The tissue pathology from the right humerus was reported as malignant hemangiopericytoma. He felt right hip pain for two months after the operation, and a plain roentgen film demonstrated a lytic lesion at the right iliac bone. He received bone curettage and the defect was filled with bone cement followed by chemotherapy with Zometa. In March 2005, right side weakness occurred and a computed

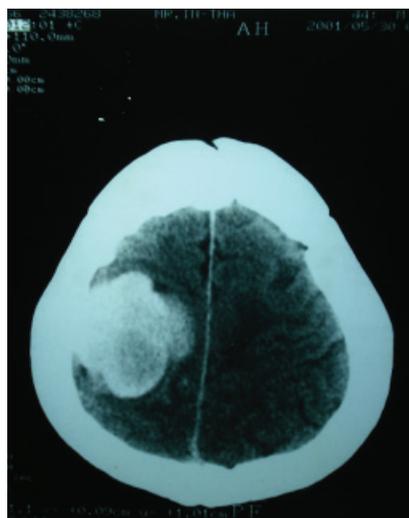


Figure 1. CT scan showing a homogeneous hyperdense enhanced lesion in the right parietal area.

tomography (CT) scan revealed a recurrent tumor in the parasagittal area. A surgical excision was performed and the pathology report showed hemangiopericytoma. Irradiation of this lesion was started. Motor weakness improved, but two months later the patient felt abdominal pain in the right upper quadrant and ultrasonography of the abdomen revealed liver and spleen metastases. He received chemotherapy with ifosfamide, mesna and adriamycin. After chemotherapy the tumor size in his abdomen remained stable.

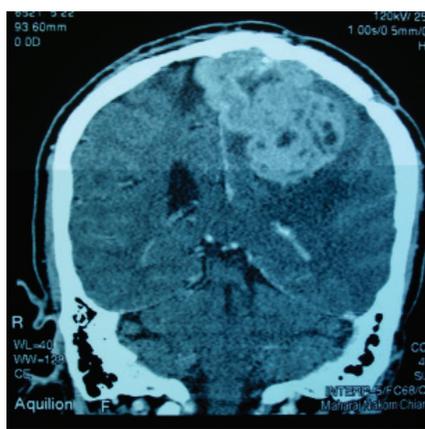
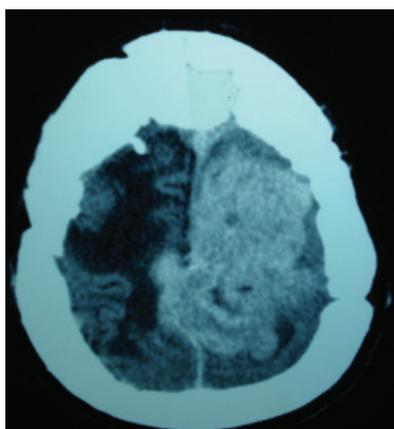


Figure 2. CT scan showing a recurrent homogeneously enhanced lesion in the left parasagittal area four years after the patient's last operation.

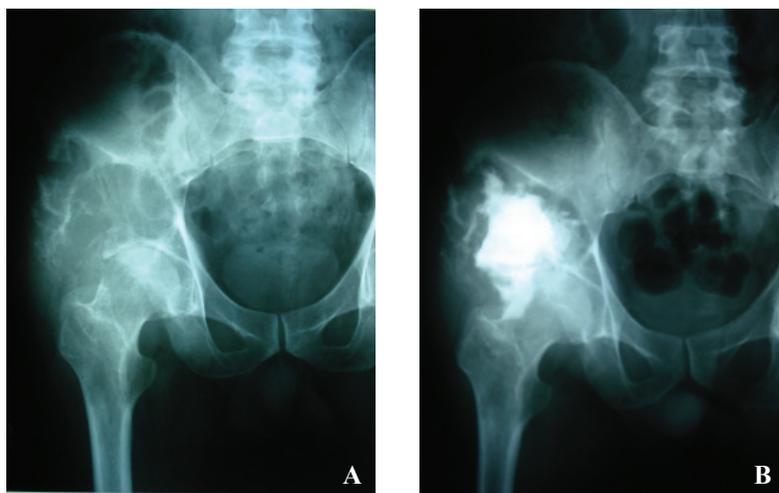


Figure 3. A, Plain pelvic film showing a lytic lesion at the right iliac with thin cortical bone; B, Plain film showing bone cement filling the lesion after curettage.

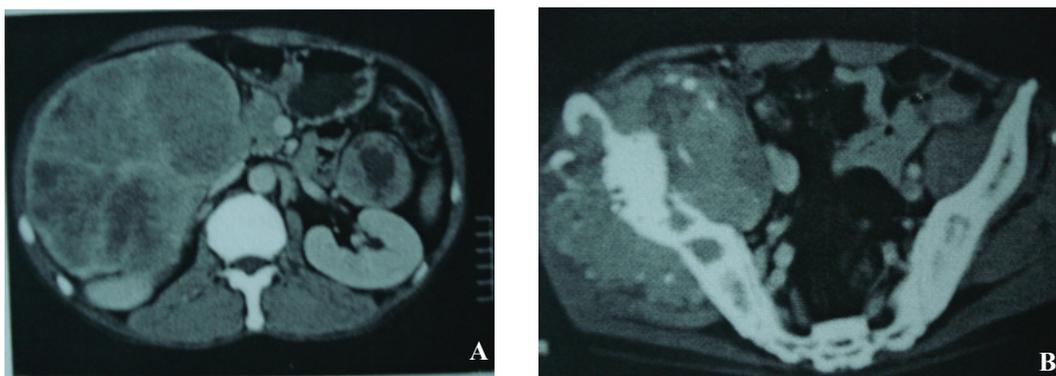


Figure 4. A, CT scan of the abdomen showing liver and spleen metastases; B, CT scan of the pelvis showing recurrent tumor in the right pelvic cavity.

Discussion

The rarity of intracranial hemangiopericytomas and the imaging similarities of these tumors with meningiomas, make their prompt diagnosis quite challenging. The differentiation is significant in the overall management of these patients, because of the aggressive biological behavior, high rate of local recurrence, distant metastasis and the poor prognosis of these tumors [5-7].

Despite the advances made in neuroimaging, the differentiation of hemangiopericytoma from meningioma is difficult. CT scan and magnetic resonance imaging (MRI) studies were not able to differentiate between these tumors [8]. The

diagnosis of hemangiopericytoma continues to be histologic [7, 9].

Surgical management is the primary treatment of these tumors. The operative blood loss is significant, due to the high vascularity of the tumors [10]. Fonton KN, *et al* recommended preoperative embolization to reduce operative blood loss. The complete resection rate for these tumors ranges from 50% to 67% in the literature [11].

The role of postoperative adjunct radiotherapy has been established. The dose of radiation needs to be at least 50-55 Gy [12, 13]. Although Dufour et al concluded that external radiothera-

py reduced the risk of local recurrence, it did not protect against peripheral metastasis [14].

Hemangiopericytomas have a tendency to recur locally. Interestingly, the extent of tumor removal has been correlated less clearly with more recurrence of hemangiopericytomas than meningiomas. Alen, *et al* reported a 33.4% recurrence rate and Fountas, *et al* 27.3% of local recurrence at a mean period of five years [5, 11].

Intracranial hemangiopericytomas can metastasize to extracranial organs. They can give delayed metastasis, which means that “disease free” patients cannot be considered cured. Metastasis appears at a mean period of eight years after the initial therapy. The most common extracranial metastasis sites: bones, liver, lungs, abdominal cavity, lymph nodes, skeletal muscle and kidneys are in order of their decreased frequency [15-23]. Guthrie *et al*. calculated that the probability of metastasis at 5, 10 and 15 years was 13%, 33% and 64%, respectively. Frequent clinical examinations, such as chest x-ray, liver enzymes, and bone scans have been used for the follow up of these patients. Fountas *et al* reported 4/11 (36.4%) cases of extracranial metastasis [11, 12].

Many modalities can be considered as treatment candidates for extracranial metastasis. In this report, curative surgical excision was selected for only the initial, solitary metastasis. Radiation therapy has been reserved for unresectable recurrent bone metastasis. The role of chemotherapy in the treatment of metastasis hemangiopericytomas has remained very controversial, as results with some partial response to doxorubicin, were very disappointing [24]. Bastin *et al* reported some benefit from an adriamycin-based chemotherapy [25].

The five year survival rate ranged from 60% to 80%. Schroder, *et al* calculated that the 5, 10 and 15 year survival rate was at 64%, 45% and 15%, respectively [26].

Conclusion

In conclusion, intracranial hemangiopericytomas are rare tumors that are characterized by repeated local recurrence and delayed extracranial metastases. The diagnosis of hemangiopericytoma requires a histological examination, in which its distinct features can be differentiated easily from a meningioma. The reported series supported aggressive surgical excision together with post surgical radiation therapy and continuous surveillance for the early detection of delayed extracranial metastases.

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**เนื้องอกเยื่อหุ้มสมองชนิด Hemangiopericytoma ที่กระจายไปยังอวัยวะอื่นๆ นอกกระโหลกศีรษะ
(รายงานผู้ป่วย)**

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Meningeal hemangiopericytoma เป็นเนื้องอกสมองที่พบได้น้อย มักได้รับการวินิจฉัยว่าเป็นเนื้องอกของเยื่อหุ้มสมอง ชนิด meningioma hemangiopericytoma มีชื่อแตกต่างจาก meningioma คือ มีโอกาสที่จะเกิดซ้ำหลังการรักษาได้มากกว่าและสามารถกระจายไปยังอวัยวะอื่นของร่างกายได้ รายงานนี้นำเสนอผู้ป่วย emangiopericytoma ที่มีการกระจายไปยัง กระดูก ตับ และม้าม **เชียงใหม่เวชสาร 2556;52(3-4):81-86.**

คำสำคัญ: เนื้องอกเยื่อหุ้มสมอง การกระจายไปกระดูก ตับ ม้าม