Case Report

Recurrent Intracranial Hemangiopericytoma with Multiple Bone Metastases: Case Report

Carlos Antonio da Silva Franca;[1] Sérgio Lannes Vieira;[1,2] Miguel Fernando Guizzardi;[1,3] Mônica Rodrigues Klemz;[4] Antonio Carlos Pires Carvalho;[5] Antonio Belmiro Rodrigues Campbell Penna;[1]

1 Inga Radiotherapy Clinic, Brazilian Institute of Oncology, Brazil

2 Pontificia Universide Catolica, Rio de Janeiro, Brazil

3 Brazilian National Cancer Institute

4 Niteroi´s Oncology Center

5 Rio de Janeiro Federal University

Abstract

Meningeal hemangiopericytoma is a rare tumor with an uncommon location in the central nervous system. We report a case with multiple brain recurrences and bone metastases. A better tumor control was obtained with the combination of surgery, radiotherapy, radiosurgery and chemotherapy. Despite the tumor native tendency to recur several times, the treatments were effective, offering a long and comfortable survival.

Keywords: Hemangiopericytoma. Radiotherapy. Brain Neoplasms. Surgery. Chemotherapy. Central Nervous System.

Introduction

Meningeal hemangiopericytoma (HPC) is a rare tumor, corresponding to 2-4% of meningeal tumors and to less than 1% of all intracranial tumors. HPC is more frequently located in the musculoskeletal system and the skin, with rare intracranial location.[1]

In the last years there has been a discussion about its true origin, and it is very often classified as a meningeal tumor due to a 1979 WHO classification which included it in the group of meningiomas, with the specific name of hemangiopericytic meningioma (grade II).[1] WHO 1993 classification distinguished hemangiopericytoma an isolated entity, putting it in the non-meningotelial "mesenchimal" group of tumors.[2] WHO current 2000 classification distinguishes it as an entity of its own.[3]

HPC is a fast growing mesenchimal neoplasia with an elevated tendency to have local recurrence and high risk of metastases.[4]

Preoperative clinical differential diagnosis is possible, but of difficult execution. Computerized tomography shows the image of a fast growing and highly vascularized meningioma, suggesting a malign tumor.[5]

HPC biological characteristic is its malignity, and local recurrence is common even after many years. In tumors of intracranial location, late extra-cranial metastases may appear. Some HPC are accompanied by paraneoplastic syndromes, specially hypoglycemia.[1]

The ideal treatment is radical surgery. Adjuvant radiotherapy increases local control rates, tumor-free

Correspondence
Carlos Antonio da Silva Franca
Clínica de Radioterapia Ingá
Rua Presidente Pedreira, 27
24210470 Niteroi, Brazil
Tel/Fax 55 21 26208401
E-mail: csfranca@ig.com.br

survival and global survival.[6]

Case Report

A male patient, 43 years, with a history of brain tumor diagnosed in 1998, submitted to surgical resection, presenting a histopathological finding of atypical meningioma. In March 2002 a brain recurrence was detected, and chemoemobilzation followed by external radiotherapy, with a 54Gy in 30 fractions. Control magnetic nuclear resonance (MNR) in January 2003 did not show evidences of residual disease (Figure 1).

In July 2003 the patient presented a lythic injury in lumbar column (L5). In November 2004 a brain recurrence was detected, and he was submitted to surgical resection followed by stereotaxic radiosurgery in March 2005, with a 7Gy dose. In August 2005 lythic injuries appeared in right femur, right iliac and right collar bone, and femoral injury biopsy confirmed the diagnosis of metastatic hemangiopericytoma. A review of

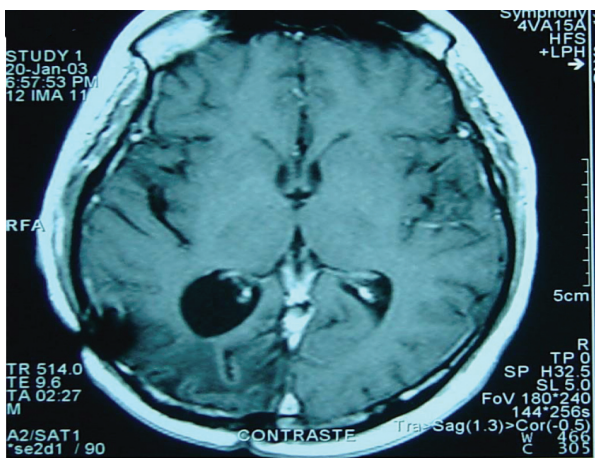

Figure 1 – Magnetic Resonance from January 2003.

the blade of atypical meningioma diagnosis was done and immunohistochemistry showed positivity for vimentin and CD34 and negativity for keratin, CEA, S-100 protein and membrane antigen, with the conclusion of a meningeal hemangiopericytoma. Palliative external radiotherapy was applied in lythic injuries with a 30Gy dose in 10 fractions. The patient received chemotherapy with adriamicin from November 2005 to February 2006. In March 2006 L5 injury became symptomatic, and external radiotherapy with a 30Gy dose in 10 fractions was given. In June 2006 a growth was detected in brain injuries (Figure 2), and radiotherapy with a 40Gy dose in 20 fractions was given. At present the patient presents important neurological improvement and radiological tests show the disease to be stable (Figure 3).

Discussion

Surgical resection is the ideal treatment for HPC, and it must be as radical as possible. However, complete excision takes place in only 50-67% of cases due to the possibility of bleeding caused by the high tumor vascularization.[7] To reduce the risk of preoperative

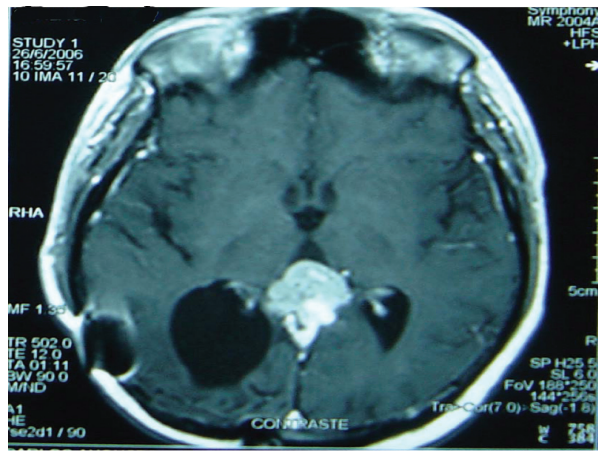

Figure 2 - Magnetic Resonance from June 2006

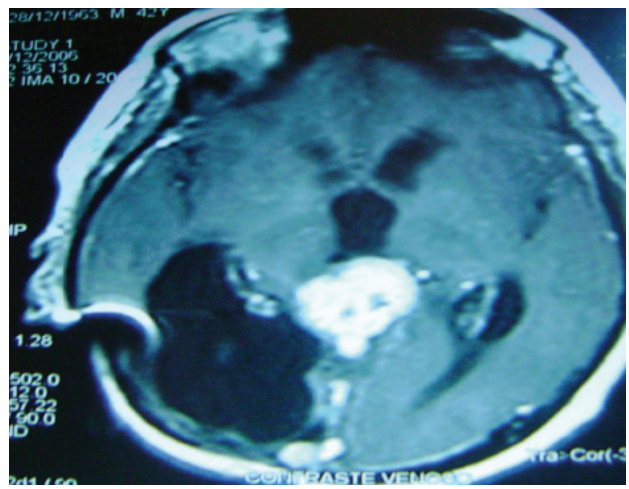

Figure 3 - Magnetic Resonance from December 2006

hemorrhage, doctors may use neoadjuvant radiotherapy, which reduces the rates of surgical complications by a nearly 80% reduction in tumor size with 20-30Gy doses.[1]

The literature shows that adjuvant radiotherapy, with a 50-60Gy dose, offers better rates of local control (57-88%) than isolated surgery (12-28%).[2,7]

In spite of this, the risk of extracranial metastases, which is around 20%, is not reduced. The most common locations are the liver and bones.[1]

Stereotaxic radiosurgery is an efficient treatment for brain recurrences with a size no higher than 40 mm of diameter, and a good local control rate is obtained (around 100% of cases).[8] Palliative radiotherapy in bone metastases is efficient, and an analgesic effect and tumor control are obtained with a 30Gy dose.[7]

Chemotherapy is indicated in HPC recurrence, but with limited results. The most efficient drug is adriamicin, which offers complete and partial remission in 50% of cases, but do not increases global survival rates. Chemotherapy is limited to very advanced stages of HPC.[4]

In this patient, recurrences appeared 4 years after the first intervention, and he is now in the ninth year of treatment. We may conclude that, in spite of tumor aggressiveness, it is possible to reach a long survival period with appropriate treatments. Due to tumor radiosensibility, skull re-irradiation may be done in patients able to enjoy clinical benefits before the appearance of late actinic complications (about 18 months).

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