Aggressive Angioblastic Meningioma with Multiple Sites in the Neural Axis

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Summary: This case report illustrates a patient who was found to have multiple sites of angioblastic meningiomas in the spinal and intracranial subarachnoid space 15 to 18 years after resection of an angioblastic meningioma from the upper cervical canal. The presence of the multiple sites of tumor could represent either multicentric origin or subarachnoid seeding.

Index terms: Meninges, neoplasms; Subarachnoid space, neoplasms; Spine, neoplasms

Central nervous system angioblastic meningioma (hemangiopericytic variant) is a rare vascular neoplasm that is locally aggressive and has the potential for extraneural metastases. In rare instances subarachnoid seeding of meningiomas has been reported. This case describes a 42-year-old woman who developed multiple local recurrences of an upper cervical canal angioblastic meningioma as well as multiple additional angioblastic meningiomas in the spinal and intracranial subarachnoid space.

Case Report

A 61-year-old woman with shoulder stiffness was diagnosed with an upper cervical canal (C-1 to C-3) angioblastic meningioma in 1974 at 42 years of age. She underwent total resection of the lesion and did well until June 1989 when recurrent symptoms developed. MR exam (0.5 T) demonstrated a large enhancing recurrent mass at C3-C5 (Fig 1A) and two smaller enhancing tumor nodules on the left side at C-1 to C-2 (not shown). A small enhancing tumor nodule within the vallecular space of the posterior fossa was also identified (Fig 1A). The patient underwent gross total resection of the mass at C-3 to C-5, which was diagnosed as an angioblastic meningioma (hemangiopericytic variant) on histologic exam.

The patient’s symptoms recurred in May 1991 and repeat MR exam (0.5 T) revealed enlargement of the tumor nodules on the left side of the cervical canal at C-1 to C-2 (Fig 1B). The patient underwent resection of the recurrent upper cervical canal mass, which on histologic exam was an angioblastic meningioma (hemangiopericytic variant). After surgery the patient received radiation therapy.

On September 18, 1991 the patient presented with a paraparesis and a thoracic sensory level. MR (.5-T) revealed two large enhancing intradural, extramedullary masses with cord compression at T-8 and T-11 (Fig 1C). Surgical resection and histologic exam of the lesions at T-8 and T-11 confirmed that both were angioblastic meningiomas (hemangiopericytic variant). Follow-up MR exam (0.5 T) on October 27, 1992 revealed that two additional tumor nodules were present on the left side of the spinal canal at T-5 and at T6-7 (Fig 1D). The patient underwent radiation therapy from C-6 to T-7.

A repeat MR image (0.5 T) of the brain on December 3, 1992 (not shown) revealed two enhancing tumor nodules in the posterior fossa. The patient underwent stereotactic radiosurgery for these lesions, which were felt to represent angioblastic meningiomas.

Discussion

Central nervous system angioblastic meningioma (hemangiopericytic variant) is a rare vascular neoplasm whose precise origin has been controversial. According to Russell and Rubinstein, the central nervous system hemangiopericytoma should be classified as a subtype of angioblastic meningioma (1). This neoplasm exhibits a more aggressive behavior than the more typical forms of meningioma, with a higher likelihood of local recurrence and potential for extraneural metastases (2). Although rare, spread of meningiomas through the neural axis via cerebral spinal fluid pathways has been described in the neurologic/neurosurgical (3–6), pathologic (1), and oncologic (7) literature.

The question of interest in this case is whether the multiple meningiomas represented multicentric tumor or seeding of the subarachnoid space. No definite answer is possible because of the lack of imaging studies before resection of the patient’s initial upper cervical canal meningioma in 1974.
Multiple meningiomas do occur in approximately 1% to 2% of patients with meningiomas, and the incidence of multiple tumors is greater in patients with neurofibromatosis type 2 (5). As noted above, there are scattered reports in the literature of patients who were felt to have spread of meningioma to multiple sites in the neural axis via seeding of the cerebrospinal fluid pathways (1, 3–7). In most of these cases the meningiomas were histologically either angioblastic or malignant. There was no evidence of neurofibromatosis type 2 in this patient. The local aggressiveness with multiple recurrences in conjunction with the aggressive angioblastic histology and the long symptom-free interval in this patient would in my opinion favor the theory of cerebrospinal fluid seeding. Further support of this contention is provided by Guthrie et al (2) who were unable to identify any cases of multiple primary angioblastic meningioma in their review of 44 cases, nor did they encounter any reports of this in their search of the literature.

As a result of the more ominous biological behavior of this tumor, aggressive therapy consisting of complete surgical resection and radiation therapy is advocated (2). The radiosensitivity of angioblastic meningioma to external-beam radiation has been questioned and Kumar et al (7) indicates that more effective doses can be delivered by endocurietherapy techniques.

Angioblastic meningioma should be included in the differential diagnosis of multiple, intradural, extramedullary spinal lesions. More commonly accepted lesions in this compartment would be neurofibromas, seeding of the subarachnoid space from other more common primary intracranial or spinal neoplasms, and metastatic lesions from a distant extraneural malignant tumor. Clinical vigilance and appropriate follow-up imaging studies including gadolinium-enhanced MR imaging (8) is recommended in patients with angioblastic meningiomas because their biological behavior is aggressive.

References
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