

CASE REPORTS

ANAPLASTIC MENINGIOMA

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INTRODUCTION

Intracranial meningiomas continue to challenge our best clinical efforts to eliminate them once discovered and deemed appropriate for treatment. Malignant meningiomas constitute 2% to 10% of all meningiomas and limited information exists regarding adjuvant treatment [1]. The external whole brain irradiation is recommended. Traditional chemotherapy has proven ineffective; thus, new chemotherapeutic agents and new methods of delivery should be developed. Immunotherapy may be considered for patients with malignant meningiomas when all other previous treatments have failed. We report a case of anaplastic papillary meningioma. A 12-year-old boy presented with partial complex seizures, headache and aggressiveness. A computerized tomography and magnetic resonance image demonstrated a large left sylvian cystic mass with peripheral contrast enhancement and minimal surrounding edema. A left temporo-frontal flap was performed. The lesion was excised in toto. Postoperatively, the patient remained neurologically intact. The treatment was complemented by external whole brain radiation.

CASE REPORT

This 12-year-old boy presented with severe headache, abnormal behavior, vomiting and two episodes of partial complex seizures during last three months. The neurological examination revealed a mild mental confusion and bilateral papilledema. There were no motor or sensory signs or symptoms. A Computerized Tomography

(CT) scan of head with contrast demonstrated a large left temporo-frontal mass with rim enhancement on post contrast study. A magnetic resonance image (MRI) obtained with and without contrast showed an enhancing lesion with central lucency in the left temporo-frontal region. Patient was put on I/V antiepileptics (sodium valproate 500mg I/V bd). Surgery was planned and a left temporo-frontal flap was raised. The tumor was highly vascular and had interface with brain. The periphery of the tumor was coagulated in order to reduce the vascularity and then central debulking was done and then again the same process was repeated till whole tumor removal. Postoperatively, the patient remained neurologically intact. There were no complications and the patient was discharged at one week, with instruction to continue antiepileptic for 2 years. The histopathologic examination of the excised specimen showed anaplastic meningioma with vascular channels surrounded by neoplastic meningotheial cells with a papilliferous aspect and highly cellular areas with mitoses. The patient was referred to radiotherapy unit to receive external whole brain radiation.

DISCUSSION

Malignant meningiomas represent 2% to 10% of all meningioma [1]. The peak incidence of atypical and malignant meningioma is in the seventh and sixth decades, respectively [2]. These malignant meningiomas are defined by several criteria including: 1) invasion of adjacent brain parenchyma or skull; 2) numerous mitoses (>5/high-powered field); 3) elevated proliferative index (>3%) as assessed by either 5-bromodeoxyuridine or KI-67 staining; 4)

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necrosis; 5) increased cellularity; 6) nuclear pleomorphism; and 7) metastasis [3].

The cell origin of the meningiomas is the arachnoid cap cell, which has a slow rate of cell division. Although tumors originating from the meninges are typically benign, they occasionally behave in an aggressive fashion and carry a much poorer prognosis than do benign meningiomas [4]. Tumorigenesis must be the result of exogenous or endogenous factors acting alone or together. Exogenous factors include trauma, viral infection, and prior brain irradiation. Endogenous stimulation can occur through the action of hormones or growth factors.

There was a clear tendency toward a progressively higher frequency of malignant meningiomas among recurrent tumor. Malignant meningiomas were tumors that had undergone reoperation and had originally been either benign or aggressive meningioma. This suggests likelihood that any benign tumor that recurs will be malignant. Whereas benign meningiomas tend to show preponderance in females, atypical and malignant meningiomas have a male preponderance [5].

Pathology:

Cushing coined the term "meningioma" to describe this tumor attached to the meninges. Since that time, a number of histopathologic schemes have been presented. Currently the WHO-II classifications of meningiomas are the most used. This histological classification system divides tumors of meningotheelial cells into four groups: classic, atypical, papillary and anaplastic. These classifications can be complemented with the Helsinki grading system. This grading system assigns points from 0-3 for six pathological features: loss of architecture, increased cellularity, nuclear pleomorphism, mitotic figures, focal necrosis, and brain infiltration. The sum of these points is then used to describe the benign, atypical, anaplastic and sarcomatous forms of

meningiomas. The proliferative potential of tumors can be quantitated, using bromodeoxyuridine, KI-67, MIB-1 and proliferating cell nuclear antigen (PCNA) labeling index, and this information helps in predicting the clinical behavior of tumors and the need for treatment [6].

Surgery:

Surgery remains an important part of treatment of malignant meningiomas. Over the past several years, advances in surgical technique and a revision of surgical anatomy have prompted more aggressive approaches to brain tumors. Despite the gross-total tumor resection, the survival of malignant meningiomas without adjuvant therapy is less than 2 years [7]. In patients with malignant meningiomas treated with surgery and adjuvant therapy (either radiation alone or radiation plus chemotherapy), median survival time was 5 years and the degree of tumor resection did not predict recurrence [8].

Chen & Liu reported that a recurrence rate with a median follow-up of 3 years after surgery was 44% for atypical or anaplastic meningioma and 6% for benign meningiomas [1]. Younis et al [9] reported that recurrence and survival time was shorter in patients with malignant meningiomas who had received partial resection on first presentation of tumor than in those who underwent total resection; in their series the patient's prognosis did not improve as a result of either chemotherapy or radiotherapy.

Dziuk et al [10] reported a disease free/progression free survival of 5 years was 39% following total resection versus 0% after subtotal resection ($p=0.001$) in patient with malignant meningioma; they stated that complete surgical resection is crucial for long-term control.

Malignant meningiomas located at the parasellar region and the posterior fossa, a conservative removal of tumor followed by irradiation is advocated in preference to a

radical operation that may cause neurological injury without being curative [11].

Radiation Therapy:

The value of fractionated external beam radiation therapy or stereotactic radio surgery in improving tumor control and survival for patients with sub totally resected, recurrent and malignant meningiomas are confirmed [12].

According to Dziuk et al [10] adjuvant irradiation following initial resection increased the 5-years disease free survival rates from 15% to 80% ($p=0.002$); when administered for recurrent lesions, adjuvant radiotherapy improved the 2-years disease free survival from 50% to 89% ($p=0.015$), but had no impact on 5-years disease free survival; the authors concluded that extent of resection, adjuvant radiotherapy, and recurrence status are independent prognostic factors.

The role of cytotoxic chemotherapy in the management of recurrent or malignant meningiomas has not been fully investigated. The value of adjuvant chemotherapy is not clear giving the confounding and probably more beneficial effects of adjuvant radiotherapy also administered to these patients [13].

Immunotherapy:

Immunotherapy may be considered for malignant meningiomas when all other previous treatments have failed. The most effective immunotherapy appears to be administration of interferon-alpha, which is relatively non-toxic and easily tolerated [14]. More studies are needed to better define the roles of these agents in the treatment of malignant meningiomas.

CONCLUSION

An aggressive treatment approach with radical surgery (more than gross total resection) and postoperative radiotherapy is

warranted in patients with these tumors. Traditional chemotherapy has proved ineffective and the role of adjuvant immunotherapy, brachytherapy or radiosurgery is unknown. Because malignant meningiomas are uncommon tumors, a cooperative group study would be required to assess co variants.

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