Malignant Meningioma after Radiotherapy for Tinea Capitis in Childhood

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We present the case of a 70 year old female diagnosed with malignant meningioma, treated with radiotherapy for tinea capitis in her childhood. The first symptoms of the disease, personality changes and left motor deficit, were present 5 years ago (2006). The imagistic evaluation at that time revealed a right frontal lobe mass. A macroscopic resection of the tumor was performed in August 2006. In 2008 the patient presented the first local recurrence and underwent surgical intervention with macroscopic resection of the lesion. One year later Gamma Knife radiosurgery with 12 Gy was performed for a second local recurrence and for a new lesion detected in the left frontal lobe. In 2011 the patient underwent a third surgical intervention and adjuvant external radiotherapy with 50Gy/25 fractions for the third recurrence in the right frontal lobe. Three months after completion of the treatment, the patient showed no clinical or imagistic signs of disease progression. The particularity of this case is the association with radiotherapy for tinea capitis in childhood, which is a proven etiologic factor for meningioma.

Key words: malignant meningioma, tinea capitis, radiotherapy

Introduction

Meningiomas represent approximately 30% of all CNS neoplasm (1) and postmenopausal women are the age-sex group with the highest incidence of these tumors (2). Benign meningiomas (WHO grad I) represent 70-85% of all meningiomas and have a good prognosis: 80% remain progression-free at 10 years after surgical resection. Only 1-3% of all meningiomas are anaplastic (WHO grad III). These are aggressive malignant tumors with a patient median overall survival of less than 2 years (1). Treatment for these aggressive tumors is multidisciplinary and includes surgery and different types of radiotherapy (fractionated radiotherapy or radiosurgery). Atypical (WHO grad II) meningiomas are the third subtype of meningiomas and account for 15-25% of cases. These tumors have an intermediate risk of recurrence and only about 35% of patients remain disease-free at 10 years with appropriate treatment.

Meningiomas are relatively understudied with regards to etiologic risk factors. Risk factors currently under study are cranial trauma, increased body mass index (BMI) and female gender. Ionizing radiation is the only proven environmental risk factor associated with meningioma. Increased risk for meningiomas was reported after high doses of ionizing radiation (3), but also after low dose levels (4), such as doses used to treat scalp ringworm (tinea capitis).

Case report

We are presenting the case of a 70 year old female patient, who was presented at our institution in October 2011 with the diagnosis of malignant meningioma. The medical history revealed: tinea capitis in childhood (when she was 8 years old) treated with radiotherapy, increased body mass index in the last decade and cranial trauma 20 years ago.

First signs and symptoms of the disease were present in August 2006: personality changes, for which she was admitted at the Psychiatric Hospital. During the hospitalization she developed a left side motor deficit. A computer tomography (CT) of the brain was performed and showed a tumor mass with edema and mass effect in the right frontal lobe (Fig. 1).

A total macroscopic resection of the tumor was performed on 22 August 2006 at the Neurosurgery Hospital. The histological report was meningioma. After surgery the patient made a full recovery of the motor deficit and was free of any signs and symptoms of disease until November 2008, when a progressive left motor deficit reappeared associated with cephalalgia. A CT scan of the brain was performed in December 2008 and revealed: a mass (4cm/3.5cm) in the right frontal region associated with edema, midline shift (Figure 2a) and inhomogeneous enhancement (Figure 2b) and a new smaller mass with the same characteristics in the left frontal region (Figure 2c). Resection of the right frontal tumor was performed on 17 December 2008.

In October 2009 a magnetic resonance imaging (MRI)
of the brain was performed and revealed a round extra-axial mass in the left frontal lobe (Fig. 3a) with contrast enhancement (Fig. 3b) and a right frontal lobe mass with contrast enhancement (Fig. 3c). Gamma Knife radiosurgery with 12 Gy for both tumors was performed (we could not acquire further information about the technique used).

The patient had no signs or symptoms of disease until June 2010, when she presented a minor left motor deficit. The MRI revealed two right parasagittal extra-axial tumors with contrast enhancement. The left frontal lobe tumor was also present, but with no signs of progression. Observation was considered the best approach at this moment. The left motor deficit gradually worsened until July 2011, when signs and symptoms of increased intracranial pressure appeared. At this time the brain MRI revealed a right frontal lobe mass with important edema and midline shift (Fig. 4). Resection of the tumor was performed in July 2011. The histological report showed anaplastic meningioma. She was referred to our institute for external radiotherapy.

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The clinical examination at presentation revealed Karnofsky Index 90, BMI=31.2, no neurological deficits, no signs and symptoms of increased intracranial pressure.

The patient underwent 3D conformal radiotherapy with total dose of 50 Gy/25 fractions/35 days to the right frontal tumor (Figs. 5, 6 and 7).

At a total dose of 40 Gy the patient developed grad 2 neurologic toxicity: increased intracranial pressure treated with dexamethasone and mannitol for 5 days.

At the last follow up visit in February 2012, the patient showed no neurological signs or symptoms of disease progression and the MRI performed showed no radiographic progression.

**Discussion**

The first clinical signs of disease in our patient and the early recurrence after treatment are typical for malignant meningioma. This subtype of meningioma has the tendency to recur after resection, while benign meningiomas have a good prognosis after surgical resection.

Fig. 1. CT scan showing right frontal tumor

Fig. 2. a. CT scan without contrast showing right frontal tumor; b. CT scan with contrast showing right frontal tumor; c. CT scan with contrast showing left frontal tumor.

Fig 3. a. MRI T2 image showing a left frontal lobe tumor; b. MRI T1 image with contrast showing a left frontal lobe tumor; c. MRI T1 image with contrast showing a right frontal lobe tumor.
With regards to the first clinical symptoms of our patient such as personality changes and motor deficit, these are between the most common symptoms of malignant meningiomas, occurring in 21% and 43% of patients (5). Unfortunately personality changes, when occurring without other neurologic deficits, are misdiagnosed as psychiatric disorders and patients are admitted in Psychiatric Departments until other neurological symptoms appear.

The majority of radiation-induced meningiomas occur after low-dose irradiation, less than 10 Gy, doses used in the past for treatment of tinea capitis (6). Children irradiated for scalp ringworm in Israel (1948-1960 the Tinea Capitis Cohort) were observed to have a relative risk of almost 10 for meningioma (4). Radiation-induced meningiomas have the tendency to have atypical histology and to recur after resection. The latency between radiation and the development of these tumors ranges from 19.5 to 35 years depending on the patient’s age and radiation dose (7). The latency period in our patient was 56 years.

Unfortunately, we could not acquire the dose and technique of irradiation used for the treatment of tinea capitis in our patient, but to our best knowledge, a low-dose (less than 10 Gy) superficial irradiation was used. Standard external beam radiotherapy dose for malignant meningiomas is 60 Gy. However, due to prior gamma knife radiosurgery and the need to spare critical structures, the optic nerve and chiasm, the total dose established for our patient was 50 Gy.

Our patient presented also other risk factors for meningioma such as: female gender, increased age, cranial trauma in her medical history, greater body mass index and low level of physical activity. Johnson et al evaluated associations of demographic, lifestyle, medical history and anthropometric variables with the risk of meningioma in the Iowa Women’s Health Study. The results of this prospective cohort study were published in 2011 and positive associations of meningioma risk with lower levels of physical activity, greater body mass index, greater height and a history of uterine fibroids, were revealed (8).
Recurrence is still the rule in atypical and malignant meningiomas. This can be recognized clinically, by appearance of new symptoms and signs or worsening of preexisting ones, or it can be seen on brain imaging before it produces clinical symptoms. Recurrence is local in the majority of cases, but multiple intracranial lesions have been reported (9). Benign and atypical meningiomas demonstrate progression of the histological subtype to a more aggressive disease at the time of recurrence: 14% of benign meningiomas progress into atypical or anaplastic histology upon recurrence, and 26% of atypical meningiomas progress into anaplastic meningiomas (10).

Surgical resection is still the most effective treatment for recurrent meningiomas and multiple surgical resections are not unusual in patients with atypical and malignant meningiomas. Radiosurgery is used for small meningiomas (tumors with a diameter of less the 4 cm), either residual after resection, progressive after prior resection, or primary untreated lesions. Local control rates range from 75% to 100%, at 5 to 10 years (1). Radiation has been used as a second-line therapy for managing intracranial meningiomas, and was often reserved for recurrences or otherwise inoperable lesions. However, in patients with atypical and malignant meningiomas, adjuvant radiotherapy improves overall survival.

As mentioned before, malignant meningiomas carry a poor prognosis. The 5 year recurrence rate is 78% and the median time to first recurrence is 3.5 years (10).

Conclusions

This case offers proof of the association between radiotherapy for tinea capitis in childhood and the development of meningioma in adulthood.

References