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Case Report

Should Adjuvant Radiotherapy Be Recommended for Pediatric Craniopharyngiomas?

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Intracranial tumors secondary to radiotherapy are rare. In this group gliomas are the rarest. Only 6 cases of glioblastoma multiforme (GBM) have been reported in patients undergoing radiotherapy (RT) for craniopharyngiomas of which only 4 have been in children less than 18 years of age. In recent years RT has become a mainstay of adjuvant therapy for recurrent or partially excised craniopharyngiomas. We report a child of 12 years who had previously undergone RT for a suprasellar craniopharyngioma and presented 10 years later with a GBM. This is the 5th pediatric case in literature demonstrating a GBM after RT for a craniopharyngioma. The implications of subjecting the pediatric population to RT for a benign lesion versus the outcome of gross total removal and management of RT induced tumors is discussed and the need to avail of safer alternatives such as stereotactic radiosurgery is stressed.

Key Words : Craniopharyngioma · Radiotherapy · Glioblastoma · Pediatric.

INTRODUCTION

Management of pediatric craniopharyngiomas has become increasingly conservative in recent years in recognition of the unfavorable outcome after extensive surgery[2]. Adjuvant radiotherapy (RT) after subtotal resection has become the mainstay of management. Modern RT continues to have significant adverse effects especially in the pediatric population of craniopharyngiomas, where it is mainly palliative for this benign lesion in a population with an otherwise long life expectancy[1]. Radiation induced tumors (RIT) are rare and high grade gliomas are even more uncommon[1]. In children who undergo RT for craniopharyngiomas secondary gliomas have been reported although extremely rarely[1]. We report a child who was diagnosed as a craniopharyngioma at the age of 2 years and later underwent radiotherapy. She presented with a glioblastoma multiforme (GBM) 10 years later.

CASE REPORT

A 12-year-old child was brought to our casualty services with features of raised intracranial pressure and left hemiparesis of grade 4. Magnetic resonance imaging (MRI) of the brain revealed an enhancing lesion in the right temporal region with central necrosis and post-contrast enhancement. The lesion measured 6.9×5.0×5.7 cm with mass effect and midline shift (Fig. 1E, F). There was a small calcified remnant in the sellar region. A working diagnosis of a high grade glioma was made and the child underwent an emergency craniotomy and decompression of the lesion. The post-operative period was unremarkable. She had previously undergone a ventriculo-peritoneal shunt and decompression of a recurrent suprasellar craniopharyngioma at our institute 4 years prior (Fig. 1C, D). She had an earlier diagnosis of suprasellar craniopharyngioma and had been operated at another institute 10 years prior to presentation. She had undergone a second surgery, within two years of diagnosis, for a residual/recurrent lesion. Fig. 1A and B demonstrate the lesion prior to the second surgery. She was then subjected to adjuvant conventional fractionated RT of 55 Gy with bilateral opposed ports with an anterior field, following which she was asymptomatic for 6 years.

Post operative MRI revealed complete decompression of the lesion (Fig. 1G, H). The histopathology revealed a GBM (Fig. 2). She was treated with temazolamide chemotherapy by an on-

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cologist and succumbed to the tumor a year after surgery.

DISCUSSION

Cahan et al.[1] described four criteria for the diagnosis of RIT : 1) the tumor must originate within the field of previous irradiation; 2) the latency between the RT and RIT should be sufficiently long; 3) the histological characteristics must differ from the primary lesion which was irradiated; and 4) the patient must not harbor any pathological conditions favoring the development of tumors such as neurofibromatosis. The case in discussion did conform to these criteria.

The commonest RIT tumors are sarcomas and meningiomas but gliomas have been rarely reported[1]. Fifteen cases have been earlier reported in literature[1]. The average age of patient at the time of irradiation was 12.5 years[1]. The present patient was much younger at the time of RT. An average of 55 Gy has been recommended which was the same dose received by this child[1]. Temporal lobe is the commonest area to be localized by the secondary glioma as it does come in the field of irradiation[1]. Of these 15 patients only 6 were GBMs and only 4 occurred in children[1]. This patient is the 5th case of RT induced GBM for a craniopharyngioma.

No correlation is documented between the dosage of RT and the latency or grade of the secondary tumors[1]. The histology has varied from low grade gliomas (usually adult patients) to GBM[1]. The pediatric population appears more prone for the higher grade of RT induced malignancies[1]. There have been no demonstrable differences in the histopathology or molecular markers between RT induced GBM and the spontaneous types[1]. High grade gliomas, post RT, occur more often in the pediatric population as compared to adults[1]. The latency period for the secondary pediatric GBMs is about 9 years[1]. Our patient presented after 10 years.

Although ionizing radiation inducing gliomas in primates has been proven conclusively[5], such an association appears cir-

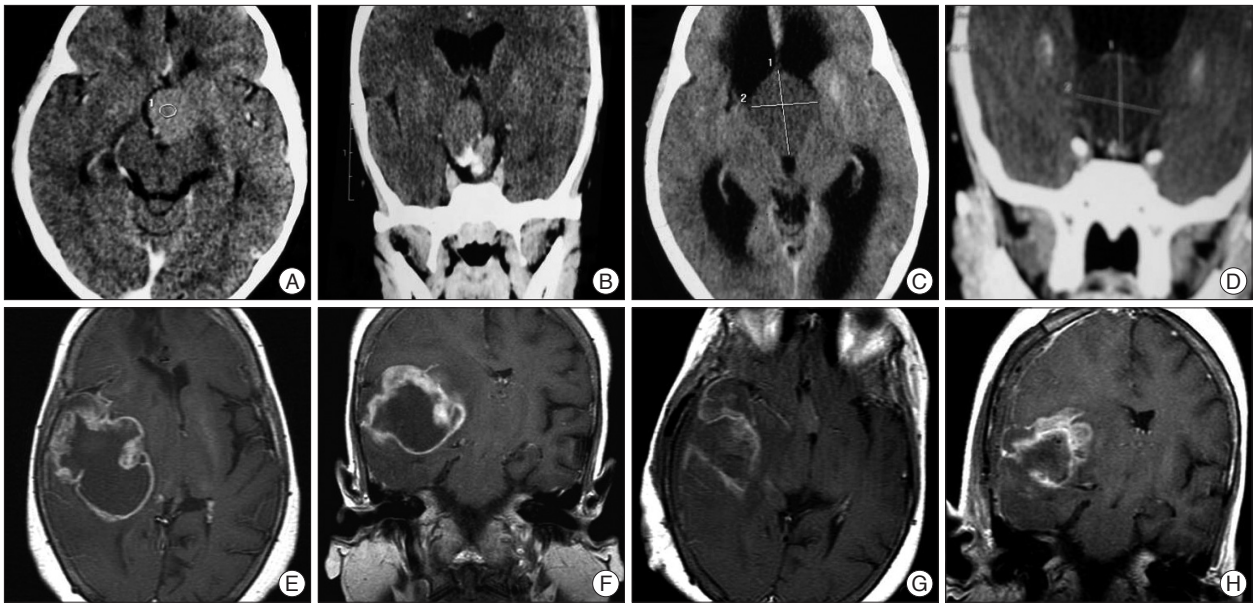

Fig. 1. A and B : Images demonstrating the craniopharyngioma at recurrence. These were the earliest CT images available. C and D : Images demonstrate the lesion with hydrocephalus when the patient had presented to our institute for the first time. E and F : Images are T1 weighted MRI images demonstrating the right temporal GBM. There is peripheral contrast enhancement with central necrosis. Mass effect and midline shift is demonstrated. G and H : Images are post operative MRI images delineating the tumor cavity with near complete excision.

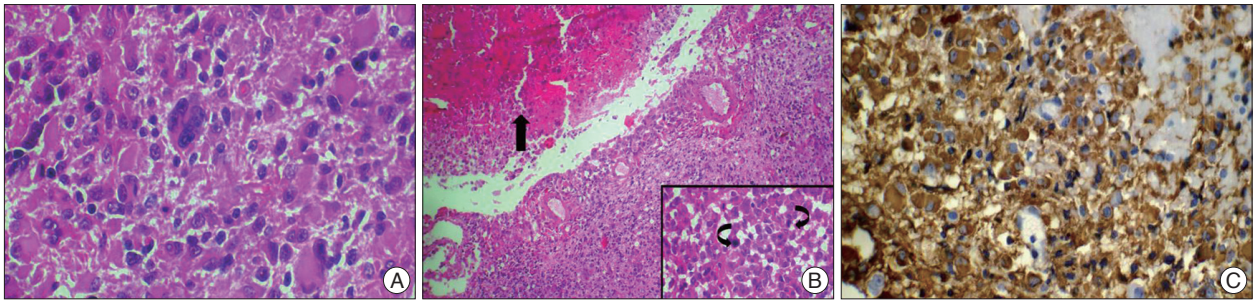

Fig. 2. Paraffin section of glioblastoma multiforme showing. Cellular and nuclear pleomorphism with tumor giant cells (A), hemorrhagic necrosis and (inset) frequent mitotic activity (curved arrows) (B). Immunopositivity for glial fibrillary acidic protein in tumor cells (C). Hematoxylin & Eosin ×400 (A); ×100 (B); ×400 (B, Inset). Avidin Biotin Complex Immunoperoxidase ×400 (C).

cumstantial based on the criteria outlined above. The actual etiological association between the RT and the occurrence of gliomas may never be substantiated at a molecular level and thus "radiation associated tumors" has been suggested as an alternative to RIT[1]. The incidence of the secondary gliomas is so small in comparison to the actual number of irradiated patients that the etiological proof is at best speculative[3]. Further research into radiation induced neuronal damage may shed light on the molecular pathogenesis of this entity.

The "do-no-harm" management for pediatric craniopharyngiomas has become popular[2,4] and reports have highlighted the individualization of therapy for craniopharyngiomas to avoid RT induced adverse effects such as neurocognitive and neuroendocrine disturbance, optic neuropathy, radiation induced vasculopathy and RIT[2,4].

Rare instances such as this case report should not deter RT being used as an adjuvant therapy especially since the benefits of RT in halting the progression of pediatric craniopharyngiomas is well documented[3,4].

Stereotactic radiosurgery (SRS) has become a popular alternative to conventional fractionated RT in recent times[7]. Although a recent report of c-knife 60Co radiosurgery-induced glioblastoma has been reported[8], SRS still appears to be a safer alternative smaller irradiated fields and the less radiation dose to the surrounding normal tissue[1]. Other modern modifications such as 3D conformal RT, intensity modulated RT and stereotactic RT (SRT) appear to be promising[3].

Recent reports have also indicated a possible role of alkylating chemotherapeutic agents in the management of radiation induced gliomas[6]. This patient did receive a course of temazolamide.

CONCLUSION

This case does stir up the debate whether adjuvant RT is warranted in benign lesions such as craniopharyngiomas, which although does appear to arrest the disease in many cases, but may be associated with the deadly complication of radiation induced malignancy. Newer modalities such as SRS or SRT may prove to be a better alternative to conventional RT.

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