Rapid and complete cyst resolution in a case of craniopharyngioma following Gamma knife radiosurgery

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Abstract:
A case of cystic craniopharyngioma is presented in which multiple micro-surgeries failed to result in cyst resolution and significant enhancement in patients symptoms. Tumor irradiation using Leksell Gamma knife resulted in complete absorption of the remained cystic parts of the tumor and dramatic improvement in patients symptoms three months after the radiosurgery .Possible mechanisms are discussed.

Introduction:
Craniopharyngiomas are the most common nonglial intracranial tumors. The negligible distance of these tumors with the visual apparatus, the hypothalamus, the vascular structures and the ventricular system frequently leads to serious endocrine dysfunction and visual impairment. Tumor recurrence is common, and it may occur despite apparent gross total resection, so the treatment of craniopharyngiomas remain controversial.(1-3)
Gamma Knife radio-surgery has become an increasingly common modality for focal therapy. More recently, the use of Gamma Knife radio-surgery has expanded to the treatment of craniopharyngiomas and several retrospective studies demonstrate good outcome (the cystic component of the tumor is always treated by combination therapy). Using multiple, convergent beams and treatment planning software, Gamma Knife radio-surgery is a highly accurate and precise technique. The combined treatment of stereotactic intra-cavitary irradiation and Gamma Knife surgery on craniopharyngiomas or gamma knife radio-surgery and stereotactic drainage of the cyst has led to tumor control in both cystic and solid components of the cyst in more than 80% of cases.(4-9)
In this paper, a case of craniopharyngioma is presented in which the cyst component is totally ablated following Gamma knife radio-surgery.

Patient and methods:
The patient was a 17 year old boy in whom a brain tumor was discovered following a CT-scan for head trauma caused by a car accident in Jan 2004. The patient underwent surgery for the brain tumor with a diagnosis of solid craniopharyngioma 3 days later, with partial tumor resection. The patient became bilaterally blind and contracted brain hydrocephalus following the craniotomy.
Pathologic examination of the resected tumor showed an invasive tumor. 27 sessions of conventional whole brain radiotherapy was performed for the patient. CT and MRI imaging two months after the first surgery and after completion of the radiotherapy course showed that the tumor has formed four small cysts, the patient was re-operated 40 days before radiosurgery for local tumor control.
The patient was then referred for radio-surgery for further treatment. At the time of admission to the Gamma knife radio-surgery unit, the patient was bilaterally blind with no light perception, had abnormal sleeping patterns, headache, nausea, paresis of the right hand and leg and symptoms of SIADH (polyuria, polydypsia).
MRI and CT scan showed well defined cystic mass with enhancing nidus in the suprasellar area (Figure 1).

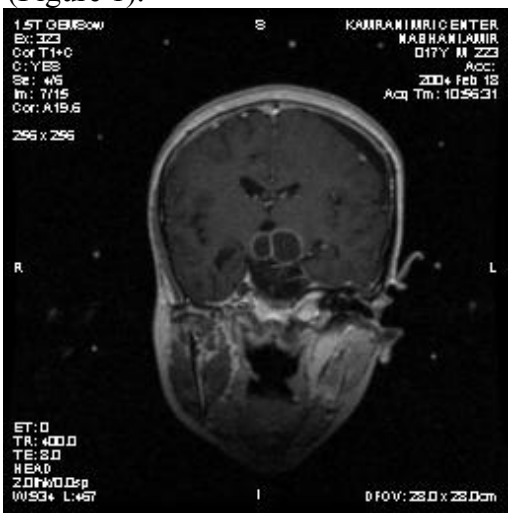

Figure 1.The MRI imaging study before radiosurgery shows a multicystic craniopharyngioma.

The patient underwent radio-surgery using Leksell Gamma Knife with the following parameters: The entire volume (9.2 ml) of the tumor was targeted with a 46% isodose of 23 Gy and with 99% coverage of the tumor and a conformity index of 1.14. The number of treatment targets were 17. The visual pathways received maximum of 10 Gy.

On repeat examination three months after the surgery, the vision was improved to the light perception status. The right-sided paresis was improved to 4/5 + power; the patient had no headache or vomiting. The endocrine abnormalities of the patient was improved with no polyuria and polydypsia.

On imaging, the cystic part of the tumor was resolved and there was no remnant present 4 months after the radio-surgery (Figure 2).

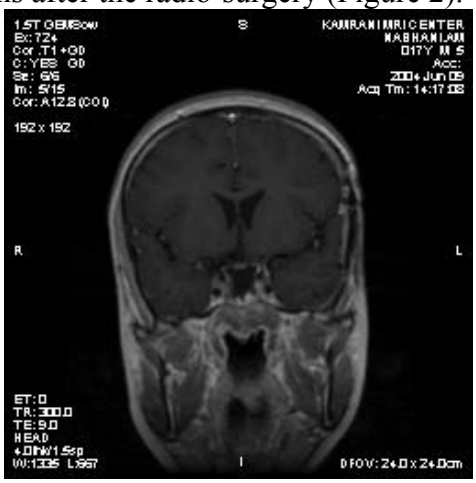

Figure 2.Control magnetic resonance imaging four months after radiosurgery indicating complete resolution of the cyst.

Discussion

Management of tumors with a cystic component, especially craniophayngioma is a major challenge in neurosurgery. Most authors recommend a multimode treatment for the cystic component of the tumor, which might be injection of radioactive compounds into the tumor cyst,[7] intracystic injection of the bleomycin[4] and insertion of Ommaya reservoir, in addition of to Gamma knife radio-surgery or craniotomy and surgical resection and drainage of the tumor. These approaches have led to variable results in the patients: while brachytherapy of the tumor leads to 90% resolution of cyst, Gamma knife radio-surgery alone is not reported elsewhere to affect the size of the cystic component of the tumor and cyst size is increased or unchanged following radio-surgery.[4-7]

The mechanism for the observed phenomenon of cyst resolution should be sought in the way they are formed. Generally, all authors suggest that the fluid forms in cystic brain tumors because of diffusion of plasma from the capillaries of the tumor,[8] Gardner et al[9] has expanded this view and suggested that tumor forms a cyst as a result of the damage to the blood-brain-barrier and accumulation of the transudated blood from the tumor capillaries inside the tumor. From a histopathologic point of view, the cavity might be formed within three locations in this tumor: **1.**Within the connective tissue containing fenestrated blood vessels; **2.**Cavity originating from the keratin-forming surface; **3.**Created by separation of desmosomal connections and fusion of numerous small honeycomb-like extracellular spaces.[10] So ablation of the intra-tumor capillaries with gamma knife radio-surgery might be a possible mechanism for cyst resolution in our case, it can also be considered that gamma knife ablation of the cyst wall, after which the cyst wall has become permeable has contributed to this phenomenon. We have used higher prescribed dose (46% isodose of 23.5 Gy) and more target (isodose) centers than the previous studies (17 in our case, median of 4 in previous studies[4]) witch allows more conformality of the prescribed dose. Both of these factors might have contributed in more effective ablation of tumor margins and cyst resolution.

In conclusion, this case might be regarded as an evidence of possible effectiveness of radiosurgery in selected cystic tumors.

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