

SHORT COMMUNICATIONS AND CASE REPORTS

Optimal treatment of a recurrent giant craniopharyngioma: lessons from a case

K. Kyrgiannis¹, S. Mourgela¹, P.N. Karamanakos², K. Liaropoulos³, N. Papadakis³

¹Department of Neurosurgery, "Agios Savvas" Hospital, Athens, Greece; ²Department of Neurosurgery, University Hospital of Kuopio, Kuopio, Finland; ³Department of Neurosurgery, "Henry Dynan" Hospital, Athens, Greece

Summary

A case of a 33-year-old man presented with symptoms of dramatic deterioration of the level of consciousness because of a recurrence of a previously aspirated and irradiated craniopharyngioma is described. The tumor had grown enormously in dimensions and was extending in the region of hypothalamus, third ventricle and brain stem, with signs of local compression and obstructive hydrocephalus. Radi-

cal surgical excision, despite the size and the location of the lesion, was the therapy of choice. The surgical technique is described. We conclude that in expert hands, microsurgery aiming at total removal should be the therapeutic option for the treatment of recurrent as well as primary craniopharyngiomas.

Key words: craniopharyngioma, microsurgery, optimal treatment, recurrent

Introduction

Craniopharyngiomas are tumors of nonglial origin, histologically benign, that arise from remnants of the Rathke's pouch, originating usually around the infundibular stalk and extending through the infundibulum to the anterior pituitary gland. They constitute 2.5-4% of all brain tumors with peak incidence at 5-15 years of age, being relatively rare in adults [1]. Despite their benign histological character and slow growth, these tumors are often associated with neurological deficits, cognitive disorders, and endocrinologic impairment. Microsurgical resection should be the first procedure considered for patients with an expanding cystic or solid craniopharyngioma [1]. With modern neurosurgical techniques, many tumors can be totally resected. However, sometimes the tumor adheres to critical nerves and blood vessels at the hypothalamus, making complete surgical resection difficult. Thus subtotal resection and/or radiotherapy have to be chosen which usually leads to a higher recurrence rate than total resection [2]. Recurrent craniopharyngiomas usually adhere strongly to the surrounding structures of the hypothalamus, enhancing the technical difficulty of repeat surgery and

increasing the surgical mortality and morbidity [3,4]. Consequently, even though microsurgery is generally accepted as the mainstay of management for recurrent craniopharyngiomas, the optimal treatment of these tumors is still controversial. In the present study we report a case of a large solid recurrent craniopharyngioma, which was treated successfully by microsurgery.

Case presentation

A 33-year-old man was admitted in our outpatient department with signs of increased intracranial pressure and frontal lobe symptoms with intense psychomotor disorders. From his previous history, at the age of 25 he suffered a mild head injury after a car accident, resulting in vision disturbances from the right eye and quick-tempered behavior. Brain magnetic resonance imaging (MRI) at that time revealed a suprasellar cystic craniopharyngioma, 4×5.5×3.5 cm in size. The patient underwent stereotactic evacuation of the cystic part of the craniopharyngioma and placement of an Omay reservoir in the tumor cavity for drainage of the cystic elements. Afterwards, whole brain radiotherapy of

Correspondence to: Petros N. Karamanakos, MD, PhD. Department of Neurosurgery, University Hospital of Kuopio, Puijonlaaksontie 2, SF-70211, Kuopio, Finland. Tel: +358 44 0246835, Fax: +358 17 173916, E-mail: petros.karamanakos@kuh.fi and me00188@cc.uoi.gr

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complete dose (28 sessions-60 Gy) was delivered and the patient improved slightly. Five years later, he presented with tumor recurrence and underwent stereotactic radiosurgery (gamma-knife), without any particular results. Regular hormonal control revealed complete insufficiency of the hypophysis-thyroid axis and received substitution treatment.

During his stay in our clinic, brain MRI revealed a large suprasellar tumor ($6 \times 4 \times 3$ cm) which was compressing the brain stem, hypothalamus, third ventricle and was elevating the optic chiasm with accompanying hydrocephalus (Figure 1). The clinical picture of the patient showed marked and rapid deterioration because of worsened hydrocephalus, and he was operated urgently

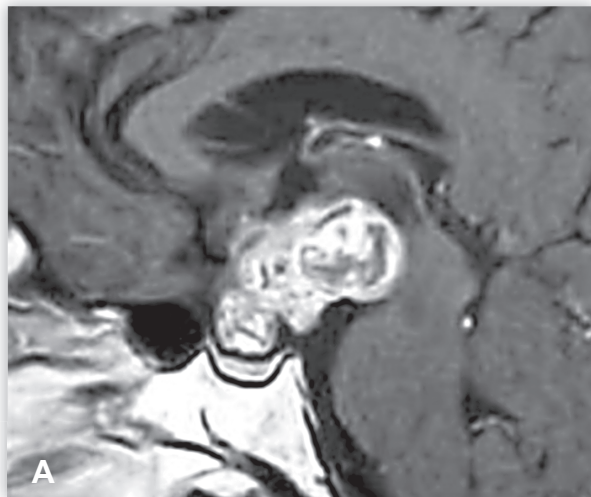


Figure 1. Preoperative post-contrast sagittal (A) and axial (B) T1-weighted MRI scans showing a suprasellar craniopharyngioma compressing the brain stem, hypothalamus, third ventricle and elevating the optic chiasm.

the same day. The Omayya reservoir was removed, and a ventriculoperitoneal shunt (magnetic Codman valve with opening pressure at 130 mm Hg) was placed left frontally. Two days later he was reoperated and the tumor was totally excised. Because of the tumor extension in the interpeduncular cistern and the cerebral crura, bicoronal skin incision and bifrontal craniotomy with removal of supraorbital arcs and crista galli was selected. Ligation of the superior sagittal sinus in front of the coronary suture was indicated, aiming at the larger possible exposure of the tumor.

The postoperative brain CT scan with contrast material confirmed the radical excision of the tumor without any other pathological findings (Figure 2). Critical for the postoperative course was the management of the hormonal postoperative disorders (hypocortisolemia and diabetes insipidus). After fluid substitution and administration of hydrocortisone and desmopressin the patient's condition improved. The neurological picture, although in the first 10 postoperative days was that of frontal lobe symptoms with drowsiness, negativism, aboulia, inert kinetic reactions, oculomotor and sphincter disturbances, it progressively improved. The mobility and superior intellectual functions such as memory and perception were acquired and only soft drowsiness remained. Five years after the operation for the recurrent tumor the patient is free of disease and with a normal brain MRI (Figure 3).

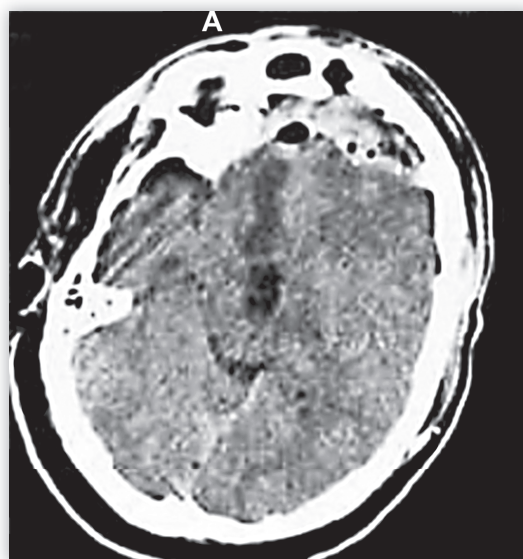


Figure 2. Axial contrast-enhanced CT scan in the immediate post-operative period showing the radical excision of the tumor without any other pathological findings.

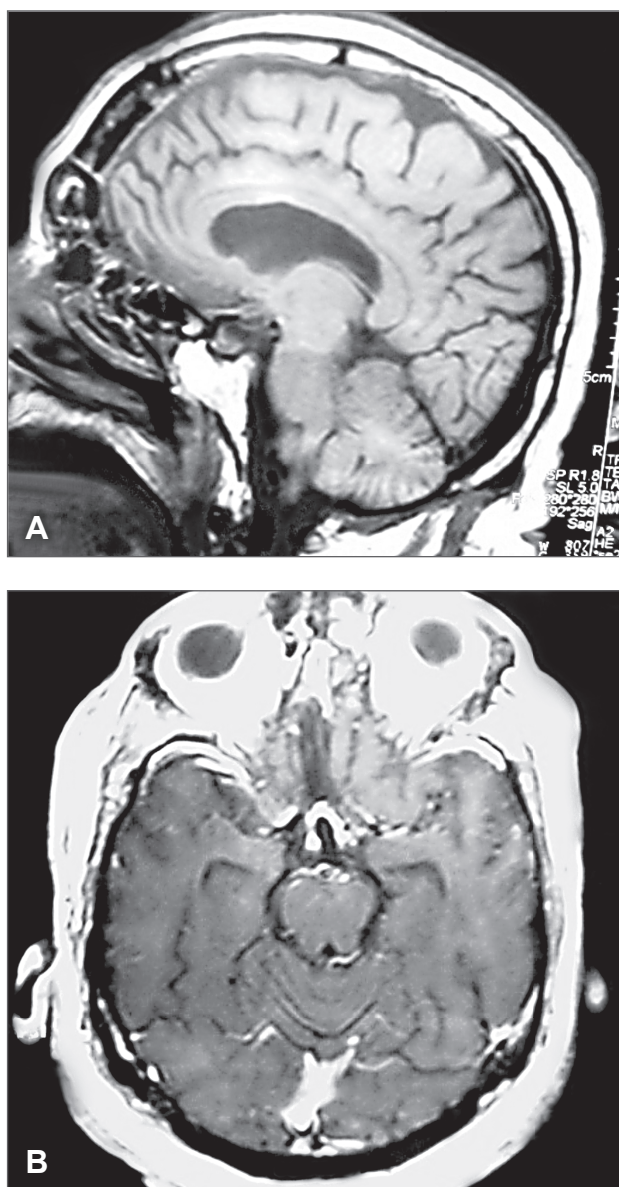


Figure 3. Five years after the operation, sagittal (A) and axial (B) T1-weighted MRI demonstrating complete removal of the tumor with preservation of the pituitary stalk.

Discussion

A clear consensus on the best therapeutic approach of primary or recurrent craniopharyngiomas has not been established yet. Craniopharyngiomas remain challenging and often enigmatic tumors. Despite their benign histological appearance, they are often associated with unfavorable and occasionally disastrous sequelae. The lack of prospective randomized studies (which practically may not be feasible and seem unlikely to be undertaken) makes their optimal therapy controversial. Total removal is undoubtedly the best treatment for craniopharyngioma if the tumor can be removed safely.

Incomplete removal at the first operation will result in recurrence, which is difficult to remove [3,5] and carries a higher morbidity and mortality compared to the primary operation [6,7]. Previous manipulation of a tumor can produce such strong adhesion that the tumor capsule becomes firmly adherent to the contiguous structures so that curative surgery is not possible for all cases. In such patients, more conservative alternative approaches are required. These include radiotherapy, which has a very high salvage rate after local recurrence following surgery [8,9], as well as gamma knife surgery, which is also reported to be effective for achieving long-term control of recurrent tumors, without compromising the patients' quality of life [10]. If the recurrent tumor is purely cystic and the surgeon is unsure whether the thin adherent wall of the cyst would be amenable to re-resection, intracavitary ^{90}Y (yttrium) may be useful as a salvage procedure, compatible with relatively long progression-free survival [11]. In situations where focal radiation is undesirable due to proximity of the visual apparatus, intracavitary bleomycin has been used with some success [12]. Finally, progressive craniopharyngiomas that had failed all conventional therapies have been treated with interferon- α -2 α with variable responses in 50% of patients [13].

Regarding the type of surgical approach that should be followed, many different opinions have been reported that divide these approaches in extra-axial (subfrontal, pterional, translamina terminalis) and transaxial (transsphenoid, transcallosal and transgyral), each of which with certain advantages but also disadvantages [5]. In our case, the tumor was enormous, of solid consistency, encapsulated, and located between sella turcica, chiasm and optic nerves. Reactive gliosis and haematoma existed in its superior part. Its exposure was facilitated by retracting the right frontal lobe and by opening the arachnoid layer across the tumor and above the optic nerves. The maintenance of the plain of the subarachnoid space was critical for total tumor resection. After the excision of the larger part of the tumor, the technique followed was internal debulking with the use of ultrasound suction (CUSA). Great attention was paid in the meticulous coagulation of the feeding vessels of the tumor, and also the maintenance of arterial anastomoses, because these arteries supply the chiasm and the optic tracts. In the regions that were adjacent to the basilar and posterior cerebral arteries, smooth manipulations should be undertaken, because of the existence of dense adhesions. Careful excision of tumor capsule in the hypothalamic area was necessary, because of the risk of penetrating the posterior anastomotic arteries that supply this region. The calcified parts of the tumor needed careful drilling before

the excision for the protection of the optic chiasm and hypothalamus. We maintained the pituitary stalk by using the microscope, despite the extent of the intervention. Even if the pituitary stalk would have been damaged, the existence of stalk remnants could lead to rebirth. The pituitary stalk was dispositional, and could be recognized by its long portal veins with the parallel course, which could be located as they penetrated the diaphragm of the pituitary gland to reach it.

In the case we present the patient underwent stereotactic evacuation of the cystic part of his primary craniopharyngioma, which was followed by whole brain irradiation. Five years later he suffered a recurrence treated unsuccessfully with stereotactic radiosurgery (gamma knife). After that point the tumor continued to grow until the patient presented to our department with symptoms of obstructive hydrocephalus. Treatment of the hydrocephalus and total resection of the recurrent tumor constituted the only and final treatment. Even though the management of recurrent craniopharyngiomas in general remains difficult due to scarring/adhesions from previous operations or radiation, which decrease the possibility of successful excision, in the case of our patient total resection of the tumor was accomplished. Accordingly, it is reasonable to conclude that if microsurgery was chosen at the first place for the treatment of the primary craniopharyngioma, most possibly total resection of the tumor would have been achieved and the operation would be probably safer and less demanding. Moreover, the possibility of a recurrence would have been significantly lower.

In conclusion, we believe that two specific "lessons" could be obtained from the presented case. First, we should properly evaluate every patient with a primary craniopharyngioma, keeping in mind that total removal of the tumor is the treatment of choice and should be attempted if possible. The recent introduction of microsurgical techniques, new devices such as the ultrasonic aspirator, modern neuroimaging methods, and advanced postoperative care allow surgery of previously inoperable craniopharyngiomas. Aggressive surgery and complete tumor resection at the first operation will ensure a low recurrence rate and an acceptable

surgical mortality. Second, even in the cases of recurrent craniopharyngiomas, microsurgery aiming at radical excision of the tumor should also be the major therapeutic option, even though previous treatment leading to extensive scar tissue formation makes surgical experience and skilled technique necessary. However, in expert hands recurrence of a craniopharyngioma can be safely and successfully managed by meticulous contemporary microsurgery without additional radiation therapy.

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