Case Report

Treatment and Management of a Recurrent Cystic and Calcificated Craniopharyngioma by Percutaneous Aspiration via a Rickham Reservoir- A Case Report Over 8 Years

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Abstract

We report on a 67 years old woman presented with a suprasellar craniopharyngioma with calcification manifesting as a progressive subjective visual deficit. Magnetic resonance imaging showed a 3 x 3 centimeter solid sellar lesion with suprasellar extension and dislocation of the neurovascular structures. The preoperative tentative diagnosis was a craniopharyngioma, this could be verified in the histological examination. At first an endonasal transsphenoidal surgery (TSS) was performed. A second TSS was performed because the patient presented a bitemporal hemianopsia and a recurrent intrasellar tumor with cystic components was radiologically proved. But only a subtotal removal was possible. When new symptoms had been reported, the patient underwent a craniotomy to achieve a gross total resection. Again the total removal was impossible because of the calcified capsule of the tumor and the adhesions with the surrounding neurovascular structures with high risk of loss of function. In follow up MRIs a newly filled cyst was shown and the patient reported again progressive visual deficits. We decided to implant a catheter into the cyst, connected it with a subgaleal reservoir to drain it by percutaneous aspirations. The symptoms relieved immediately and no further surgical resection was needed. Each time the patient described visual problems an aspiration of the fluid was performed. 80-90% of this kind of tumor has cystic components and makes surgical therapy difficult. In the presented case the repeated drainage of cystic craniopharyngioma was an effective palliative treatment.

ABBREVIATIONS

TSS: Transsphenoidal Surgery; CSF: Cerebrospinal Fluid; GTR: Gross Total Resection

INTRODUCTION

Craniopharyngiomas are histologically non-malignant, partial cystic and in some cases calcified tumors of the sellar region, which accounts 1, 2-4, 6% of the intracranial tumors and have an annual incidence of 0, 5-2, 5 per million [1,2]. The presence of single or multiple cysts, which can account 80-90% of the total bulk of the tumor, is characteristic [3]. The craniopharyngioma could be primary cystic or it occurs in relapsing craniopharyngioma. This kind of tumor has a wide spectrum of presenting symptoms usually visual changes, increase of intracranial pressure or endocrine abnormalities. Motoric deficits and mental changes are reported in some cases [1].

The therapy of a craniopharyngioma is challenging. In most cases total removal is not possible. A subtotal excision will lead to a recurrence of the tumor in up to 70% of patients [3]. Alternative treatment options are chemotherapy, radiotherapy or gamma stereotactic radiation [4]. In case of a mainly cystic craniopharyngioma the drainage of the cystic lesion by a catheter which is connected to a subcutaneous reservoir or/and to the cerebral fluid interspaces could be effective and leads to a rapid
decrease of symptoms for the patients. It is a palliative treatment, if the surgical removal is impossible because of an ossified capsule or in elderly patients, who are in reduced condition. Percutaneous aspiration can be easily performed each time the patients is symptomatic. Moreover, this simple and safe method lacks the risks associated with surgery or chemotherapy [3].

However, this procedure is not a common therapy. This report is about a case of a 67 years old woman with a huge craniopharyngioma, which recurred in cystic manifestation after the primary surgical resection and was treated efficiently by drainage of the cyst.

CASE PRESENTATION

We report on a 67 years old woman, who was initially diagnosed at age of 59 with a suprasellar craniopharyngioma. At this time, in July 2007, she presented a subjective visual deficit like blurred vision. A cranial MRI demonstrated a 3x3 centimeter solid sellar lesion with suprasellar extension. The preoperative tentative diagnosis was a craniopharyngioma. Laboratory a reduction of prolactin, human growth hormone and cortisol was proved, so the tumor seemed to be inactive. A transsphenoidal approach for resection was utilized, but it was unsuccessful to remove the tumor completely. Sadly the patient showed a rhinoliquorrhoea. A lumbar drainage was implanted and the liquorrhoea suspended. The histological examination just showed some necrotic tissue.

The first year follow up MRI presented a recurrence of the lesion intrasellar with a new fluid component and compression of the overlying optic chiasma. In the clinical examination a progressive bitemporal hemianopsia was identified. Decision for a second transsphenoidal surgery to relieve the fluid of the cyst was done. Unfortunately only a small part of the solid components could be removed and the wall of the cyst did not collapse. Postoperative the visual disturbance was regressive. In the histological examination a regressive modified craniopharyngioma was proved.

Six months later the patient suffered a recurrence of the visual deficits. A recurrence of the cystic lesion was detected in the MRI. Because of the two unsuccessful TSS, a right sided craniotomy was performed to achieve a gross total resection (GTR). Opening the cyst using an ultrasonic aspirator to remove piecemeal the extremely hard calcified components of the capsule resulted in the outflow of typical craniopharyngioma fluid, dark brown in color. After this it seemed like a discreet subsidence of the capsule and a decompression of the optical nerve was reached. However, it was impossible to remove the whole tumor, because of the adhesions to the surrounding neurovascular structures. The result of the operation is a subjective improvement of the visual deficit.

In September 2009, five months after the craniotomy and the decompression of the cyst, the patient reported again symptoms like a visual lost especially of the left eye, again a bitemporal hemianopsia and progressive headaches. The cerebral MRI showed a new filled cyst and a displacement of neurovascular structures, especially of the left sided structures. We decided to proceed a palliative treatment of the cyst, because a gross total resection includes a high risk of injuring or destroying the optic chiasma, optic nerves or of the carotid artery. The patient underwent another right sided craniotomy. Intraoperative we could see a compressed left optical nerve and optic chiasma due to the cyst. After opening the calcified capsule pressured outflow of xanthochrome fluid was seen. No solid tumor components were detectable by using the endoscope. We implanted a catheter in the cavity of the cyst and the calcified capsule (red arrow) becomes apparent.
a subcutaneous Rickham reservoir. Postoperatively the patient recovered well and the drainage of the cyst produced regression of the neurological signs and symptoms. Now it was possible to aspirate the cystic fluid percutaneously. After every performed operation the patient was substituted with hydrocortisone and thyroxine and she was in ambulant endocrinological therapy and control.

In follow up MRI and clinical examination 6 months later, there was still a cystic craniopharyngioma, but the patient was without any symptoms. So there was no need for a percutaneous aspiration. In 2011 the imaging shows a reduction of the cyst.

4 years later after the last operation, in 2013, a new MRI demonstrated an extension of the cyst. Simultaneously the patient reported about headaches and subjective pressure behind the left eye. Based on these symptoms which correlate with the images, we indicated first time the aspiration via percutaneous puncture of the Rickham reservoir. It was possible to aspirate easily a few milliliters of cystic fluid. There was an immediate improvement of her symptoms.

After the first aspiration we repeated this procedure 4 times with an interval between 4-9 months, every time, when she becomes symptomatic with headaches or visual deficit. No side effects or complications could be reported during the treatment period.

**DISCUSSION**

Because of the characteristic of the craniopharyngioma like the existence of a capsule, its extra cerebral location or the risk of loss of function makes a gross total resection impossible in many cases [7]. Another limiting characteristic of a craniopharyngioma for a GTR is a cystic component and the present of a calcified capsule. The reduction of the residual cyst often becomes the main problem. Mostly tumor calcification and cystic components occur in adamantinomatous tumors predominantly occur in children but also in adults [1]. Typically the cysts are filled with thick oily fluid high in protein, blood products, and/or cholesterol, it is called “machinery oil” like we could see in the aspirated fluid.

Some reports found out that tumors with lacking calcification had a significantly better 5 year survival rate. An increased incidence of calcification in recurrent tumors has also been detected [1]. In dependence on these factors it requires an individual approach to each patient.

Palliative measures have been developed to deal with symptoms that recur during the chronic course of the disease [6]. The main object of the treatment of patients with craniopharyngioma is to maximize local control, minimizing acute and long-term complications and an optimal quality of life [7,8].

In our case, the patient was treated twice via endoscopic transsphenoidal surgery. This minimal invasive approach is used for smaller and intrasellar tumors and allows a good infrachiasmatic exposure without manipulation of the surrounding neurovascular structures, avoids brain retraction and there are no visible scars [5]. But in case of our patient the tumor recurred with cystic components again, because of the incomplete removal of the cystic wall. So the next step was an open transcranial technique. However, a completely resection of the tumor was impossible, because of the extremely hard and calcified capsule and the adhesions to the optic apparatus and the carotid artery. We only could relieve the fluid of the cyst.

For the treatment of cystic craniopharyngioma, which presents as much as 60% of the cases and almost 100% of recurrences, special therapeutic options have been considered [8]. Based on the fact that the tumor is benign in theory it should be enough to reduce the production of the fluid in the cyst and to prevent the accumulation and to support the delivery into the CSF or outside. To reduce the production of the fluid you have to reduce the proliferation in the cystic wall. This could be obtained by radiation therapy [8]. Until now our patient did not undergo a radiation therapy. Instead we began a palliative treatment by drainage of the cyst. The drainage is a minor surgery and rapidly to archive and could be obtained in several ways like the classical way during a craniotomy or by a less invasive and less traumatic stereotactic percutaneous cyst tapping [4,7,8]. The implantation of a catheter is an easy way to aspirate the cystic fluid and to manage the rapid increase of the intracranial pressure caused by the cystic tumor, even if its effect is transient [4].

The fourth operation was the implantation of catheter into the cavity of the cyst with a possibility to drain in the basal cistern and connected it with a Rickham reservoir implanted subgaleal, where it is easily accessible for a percutaneous puncture. In this way a communication between the cyst cavity and the cerebrospinal fluid (CSF) for a spontaneous dilution into the CSF was done. It was efficient over 4 years, so that aspiration of the cyst through the reservoir was not necessary.
In the follow up MRI of our patient a decrease of the cyst has been demonstrated. Deductive, the mechanism with the dilution within the CSF seems to work. In other case reports with reporting about emission of cyst fluid into the CSF, no side effects like chemical meningitis or meningeal irritation nor alterations on the liquor flow or resorption were observed when this method was used [4,7]. In principle there is always the chance, like Spaziante et al., or Al Abyad and El-Sheikh reported, that there is no need to repeat the puncture because of the dilution of the cystic fluid in the CSF after some time [3,4,8]. Fahlbusch et al., reported that once the cyst collapsed and the craniopharyngioma was retracted from the brain and a removal of the solid portion was facilitated [7]. In our case there is no possibility for an event like this, because of the calcified capsule.

After 4 years with a stable condition, the patient reported to suffer from headaches and visual deficit again. In a new cerebral imaging we could see an increase of the craniopharyngioma cyst. So we performed recurrent aspirations of the cyst, which improved the symptoms of our patient. Other cases refilling of the cyst is observed as well [7].

The interval of the aspirations was getting shorter in our case and in the future we should discuss with the patient if it is reasonable to start an alternative therapy. Moussa et al., reported about patients with a cystic craniopharyngioma, treated by insertion of Ommaya reservoir systems and aspiration of the cyst, but had a recollection of the cyst in the first 3 month from surgery. After conforming that there was no leakage from the cyst, they implanted bleomycin and the patients showed an improvement and needed an aspiration only every 6 month [3].

Possible complications of this therapy include infections, dislocation of the catheter and obstruction of the catheter with debris or viscous fluid or shifting of the reservoir with problems of finding it [4].

In cases with a mixed craniopharyngioma with growing solid components the treatment with drainage of the cystic fluid will be problematic. The aspiration of the cyst does not affect the solid tumor pieces. In this content a combined radiation therapy was reported [8].

There are many ways to treat a craniopharyngioma. A radical surgery is recommended as the initial therapy. But the goal of treatment should be a selective removal of the craniopharyngioma with preservation of the functionally important structures, directly or indirectly [7]. This includes the draining of the cyst in its several ways. The drainage operation should be regarded as an easier way for the treatment of this tumor and should gain a greater use [4]. It is also a good option of treatment of elderly patients with risk factors, who are not in a good condition and are not able to undergo an operation [7].

REFERENCES