

# Recurrent Chemical Meningitis in Craniopharyngioma without Reduction in Size of Cyst: Case Report of Two Cases and Review of the Literature

## *Kist Boyutlarında Küçülme Olmaksızın Tekrarlayıcı Kimyasal Menenjitte Yol Açan Kraniofarengioma: İki Olgu Sunumu ve Literatürün Gözden Geçirilmesi*

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### ABSTRACT

Chemical meningitis is a rare phenomenon due to rupture of a craniopharyngioma cyst; it develops because of presence of cholesterol crystals in cyst fluid secreted by the squamous epithelium lining of the cyst. Spontaneous rupture of the cyst may present with or without meningitis, depending upon the cholesterol contents of cystic fluid of tumor. A decrease in the size of cyst may or may not be noted. Here we report two cases of craniopharyngioma, one which had a recurrent episode of chemical meningitis after surgery due to leak from residual component of the craniopharyngioma without evidence of recurrence of lesion, or decrease in the size of the cyst. Another case presented with initial symptoms and signs of meningitis, with no change in the size of the lesion after the second episode of chemical meningitis.

**KEYWORDS:** Chemical meningitis, Craniopharyngioma, Rupture of craniopharyngioma cyst

### ÖZ

Kraniofarengioma kistinin rüptürü sonucu oluşan kimyasal menenjit nadir görülen bir fenomendir. Kraniofarengioma kistini döşeyen skuamöz epitelin sekresyonu içinde yer alan kolesterol kristallerine karşı gelişen reaksiyon sonucu kimyasal menenjit oluşur. Kraniofarengioma kisti rüptürü sonucu her zaman kimyasal menenjit gelişmez, kimyasal menenjit gelişimi için kist içeriğinde kolesterol kristallerinin olması gereklidir. Kist boyutlarında küçülme ile ilgili bir bilgiye rastlamamıştır. İki ayrı kraniofarengioma olgumuzda kimyasal menenjit görüldü. İlk olgumuzda cerrahi sonrası tekrarlayan menenjit atakları görüldü. Bu görülen menenjitin tümörde rekürens olmadan direkt olarak rezidüel dokuya bağlı olduğu düşünüldü. Diğer olguda ise ilk semptom olarak menenjit ortaya çıktı, ikinci menenjit atağı sırasında yapılan kontrollerde kist çapında bir değişiklik görülmedi.

**ANAHTAR SÖZCÜKLER:** Kimyasal menenjit, Kraniofarengioma, Kraniofarengioma kist rüptürü

### INTRODUCTION

Chemical meningitis following rupture of suprasellar cysts is an uncommon phenomenon. The rupture is usually associated with shrinkage of the cyst. Chemical meningitis may or may not occur; this usually depends upon the cholesterol contents of cystic fluid. Here we report two cases of craniopharyngioma where there was no change in the size of the cyst, although the patients had chemical meningitis.

### CASE REPORT

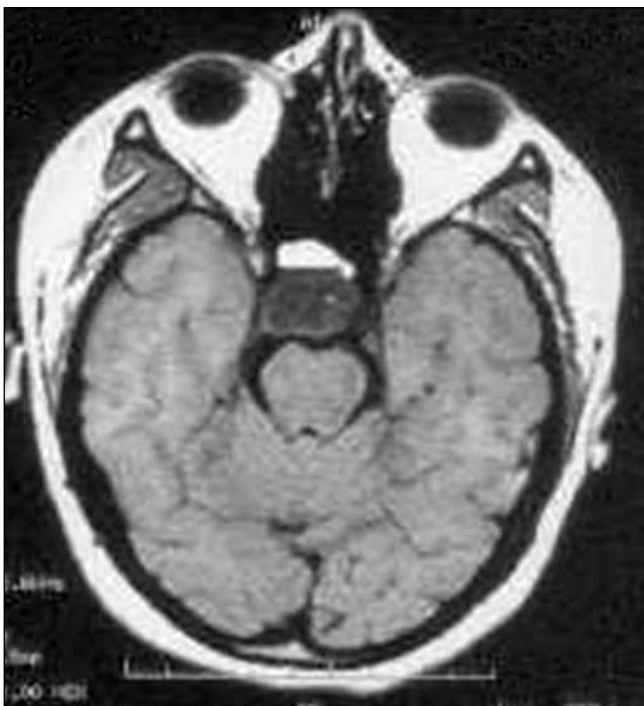
#### Case 1

A student of 17 years presented in 2002 with complaints of headache, vomiting and diminution of vision for two months. She was initially diagnosed at another center as a case of

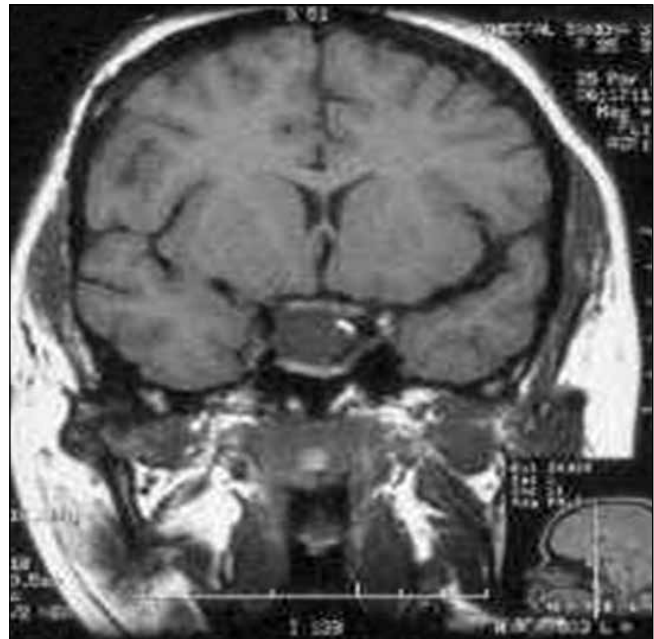
tubercular meningitis and antitubercular treatment was started. The patient did not responded hence was referred to us. She had severe holocranial headache and vomiting with bilateral papillaedema. There was no neurological deficit other than the sign of meningismus. CECT scan head showed a small well defined hypodense, homogenous, nonenhancing space occupying lesion in the sellar suprasellar region sized 1.53 x 1.96 x 1.5 cm without calcification. The ventricular size was normal. MRI Head showed a similar sized mass on sellar suprasellar cistern with T1 hypointense and T2 hyperintense appearance, with a solid component present in the prepontine and hypothalamic area. It was displacing the brain stem posteriorly and extending to the anterior third ventricle superiorly.

Right pterional craniotomy and excision of the tumor were performed. The capsule was punctured initially and the cyst fluid was aspirated. The fluid was examined and had no malignant cells. A small solid portion of tumor present in the posterior part of third ventricle and partly attached to the pituitary stalk was left behind as it was attached firmly to these structures. Histology showed that the tissue was composed of cystic spaces lined by stratified squamous epithelium resting on a fibroconnective stroma infiltrated by a mixed inflammatory cell infiltrate composed of groups of foamy macrophages and lymphoplasmacytic cells. The biopsy was suggestive of craniopharyngioma (papillary type). Postoperatively she improved clinically and was discharged on the ninth postoperative day. She was advised radiotherapy for residual solid tumor but declined. She remained asymptomatic for two years. She again developed symptoms of meningismus, but had no evidence of any foci of infection. CSF study revealed cell count 650, neutrophils 51%, lymphocytes 49%, CSF glucose 62 mg % with blood sugar 100 mg %, and CSF cholesterol was 11 mg %. MR imaging showed no change in the size of the tumor, and a diagnosis of chemical meningitis was made. She was treated with steroids and recovered within one week. The third episode of meningismus occurred after six months but with less intensity. MR head (Figure 1-3) showed no change in the size of the tumor. She improved again on steroids. CSF study was done and was again suggestive of chemical meningitis. She was again counseled for radiotherapy but she declined. The fourth episode of similar complaints occurred after 6 months. CSF study showed cell count 40/mm all lymphocytes,

CSF sugar 46mg % blood sugar 76mg % proteins 29.5 mg %, cholesterol was 8 mg %. MR imaging again had no change in the size of the tumor. She was managed again on steroids and the symptoms improved gradually with decreasing CSF cholesterol levels. She is being admitted again with similar complaints for the 5<sup>th</sup> time and surgery was done for the residual tumor, to take out residual tumor with cyst decompression. She has been on regular follow-up for 6 months and has not had any complaints.



**Figure 1:** Axial section T1-weighted contrast study of brain, showing residual solid part attached to pituitary stalk.



**Figure 2:** Coronal section T1-weighted contrast study of brain, showing residual solid part attached to pituitary stalk.



**Figure 3:** Sagittal sequence T1-weighted contrast study of brain, showing residual solid part attached to pituitary stalk.

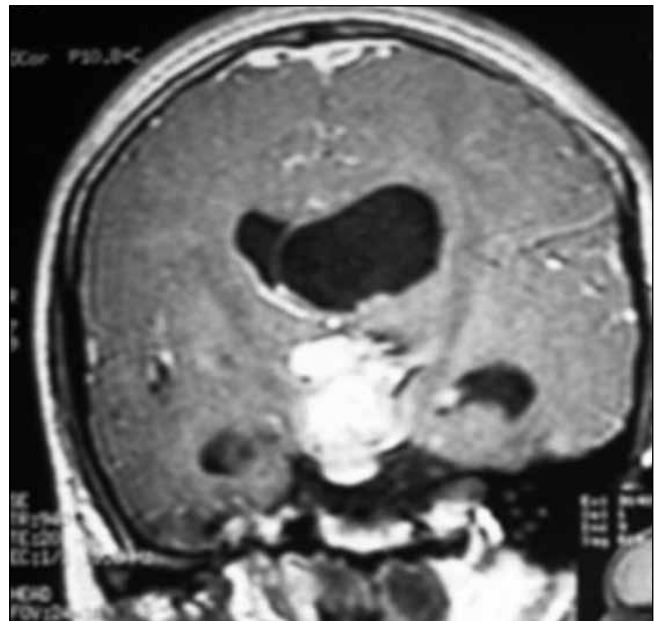
## Case 2

A male student of 16 years presented with complaints of high grade fever, moderate to severe holocranial headache with recurrent episodes of vomiting, photophobia and restriction of neck movements. On examination he had no neurological deficits except neck rigidity. He was treated on broad spectrum antibiotics for two weeks in a private hospital; details of treatment were not available. He was referred when he did not respond to treatment. NCCT head showed large sellar suprasellar lesion of size, 2.8 cm x 2.0cm with hydrocephalus, MR head showed 3.1 cm x 2.9 cm x 2.7 cm sized lesion in the sellar suprasellar region indenting and obstructing the third ventricle, hyperintense on T1- and T2-weighted imaging with contrast enhancement and hydrocephalus with significant periventricular oozing suggestive of craniopharyngioma with hydrocephalus (Figure 4).

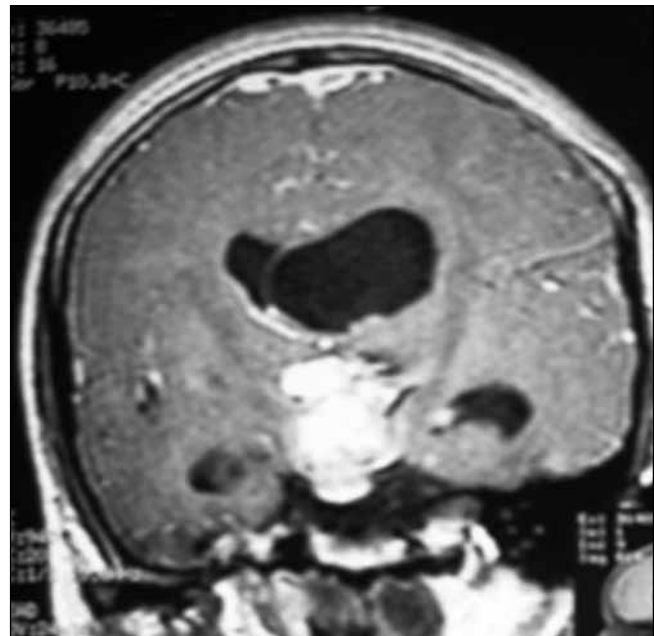
In view of the meningismus, a CSF study was done. Cell count was five, all lymphocytes, CSF glucose 40 mg % blood glucose 65 mg % , protein was 17 mg % CSF cholesterol 8 mg %. The CSF culture was sterile. A diagnosis of chemical meningitis was made and he was put on steroid therapy. He responded well to steroids. MRI brain was repeated (Figure 5) which showed same size of tumor with nearly similar characteristics. The patient again had similar complaints 1 week after the resolution of the first episode of meningitis. Again steroids were given with increased dose, to provide an improvement. He was operated by right pterional craniotomy with subtotal excision of tumor. The tumor was solid cystic, and the cyst had a minute tear through which cystic fluid was leaking. A small solid part attached to the hypothalamus was left, and otherwise the whole cyst was removed. Biopsy result was craniopharyngioma (papillary type). Patient has had no complaints of meningismus during follow-up.

## DISCUSSION

Chemical meningitis has been reported from intracranial rupture of several types of cystic lesions such as dermoids (9), epidermoid cysts in the cerebellar pontine angle cisterns, cysts of Rathke pouch (2, 9), cholesteatoma following radical mastoidectomy and remnant after geniculate ganglion surgery. The mechanism of rupture of a craniopharyngioma cyst may depend on weakness of the cyst wall caused by cyst expansion inducing degeneration of cyst wall (5). Chemical meningitis in craniopharyngioma is supposed to be caused by the cholesterol crystals contained in the cystic fluid (3, 8). The rupture or leak result in headache, fever, nuchal rigidity, headache and photophobia (3, 8). The cyst rupture can be associated with or without resolution of pressure-related symptoms (ocular finding) caused by the cyst. Most of the time it is associated with a decrease in size of cyst unlike our case where clinical features and CSF finding were strongly suggestive of chemical meningitis but there was no decrease in the size of cyst or improvement in neurological status. In both our cases it may be on account of small and recurrent leak of cystic fluid of craniopharyngioma and may be explained on the basis of increase of intracystic pressure at a particular



**Figure 4:** T1-weighted magnetic resonance imaging showing sellar and suprasellar mass with brilliant enhancement on contrast, after first episode of meningitis.



**Figure 5:** T1 weighted magnetic resonating imaging showing sellar and suprasellar mass with brilliant enhancement on contrast, after second episode of meningitis. There is no change in size of tumor.

level after a particular duration resulting in a small leak from minute opening of cysts. In addition, a rupture of the cyst may cause cerebral infarction following vasospasm (7). However, the quantity of cholesterol crystals produced by ruptured craniopharyngioma varies greatly. Cases of low cholesterol content have also been reported (1). Therefore the possibility

of chemical meningitis is low if the cholesterol content of the craniopharyngioma is low (4,8,9). Unfortunately, the cholesterol content cannot be estimated on MR imaging, because the protein concentration contributes more to the signal intensity than the lipid concentration on the T1-weighted image (6,10). Estimation can be possible by NMR spectroscopic finding or MR mass studies, once done on solid state MR. We consider cholesterol level as significant in our cases causing chemical meningitis. An improved method of MR imaging is required to detect the cholesterol content of the cyst so that problem of chemical meningitis due to cyst rupture can be anticipated quantitatively and qualitatively in advance. Cases of residual or recurrent tumors should be followed carefully, taking the possibility of chemical meningitis into account. Chemical meningitis should be considered in differential diagnosis even in postoperative periods of craniopharyngioma. Long-term follow-up is required for recurrence of the cyst. Radiotherapy may be considered if cyst recurrence is seen. Chemical meningitis is therefore a strong possibility in operated cases of residual or recurrent craniopharyngioma though the occurrence of meningitis depends on a leak and contents of cholesterol in particular for a cyst of craniopharyngioma. It may be surprising to note chemical meningitis even without the radiological evidence of leak or decrease in tumor size. CSF cholesterol important in the diagnosis of chemical meningitis. It is almost undetectable in the CSF. The presence of cholesterol is in favour of chemical meningitis. Regarding treatment, the source must be excluded along with administration of steroids.

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