Synchronous Morphologically Distinct Craniopharyngioma and Pituitary Adenoma: A Rare Collision Entity

Harjinder S Bhatoe*, Prabal Deb and Sudip Kumar Sengupta
Institute of Neuroscience, Max Super Speciality Hospital, New Delhi, India

Abstract

While pituitary tumors and craniopharyngiomas share a common lineage, their simultaneous occurrence is distinctly rare. We present one such patient, an adult male with two distinct tumors, that were excised by two different approaches. Relevant literature is briefly reviewed.

Keywords: Brain tumor; Collision tumor; Craniopharyngioma; Pituitary tumor

Introduction

Simultaneous occurrence of morphological distinct, discreet intracranial tumors sharing the same cell lineage is a rarity. Pituitary tumors and craniopharyngiomas share a common lineage. Simultaneous occurrence of these two tumors in the same patient is rare and has been reported only nine times so far (Table 1). While pituitary tumors are centred in the sella, craniopharyngiomas may occur anywhere from the pituitary gland to the third ventricle. Association of intra-third ventricular craniopharyngioma and growth hormone-secreting pituitary macroadenoma as two distinct, unconnected tumors occurring synchronously has not been reported so far.

Case Report

A 35-year-old male was admitted with six-month-history of generalized headache, gradual loss of vision and intermittent generalized tonic clonic seizures. Clinically, he had acromegaly and optic atrophy with no perception of light. There was no other neurological deficit. His growth hormone level was 18 ng/ml (normal 1-5 ng/ml).

MRI brain revealed lobulated frond-like solid discreet tumor within the third ventricle causing obstruction to ventricular system and obstructive hydrocephalus. There was no evidence of calcification or hemorrhage. He also had sellar enlargement with homogenously isointense pituitary macroadenoma (Figures 1-3).

He underwent right fronto-parietal minicraniotomy and transcortical transforaminal excision of light coloured, solid, relatively avascular third ventricular tumour. The tumour could be excised completely (Figure 4). Postoperative period was uneventful. MRI three

Table 1: Reported cases of pituitary adenoma with craniopharyngioma.

<table>
<thead>
<tr>
<th>S. No</th>
<th>Reference</th>
<th>Craniopharyngioma</th>
<th>Pituitary adenoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>Cusimano, et al. [3]</td>
<td>Suprasellar cystic</td>
<td>Microprolactinoma</td>
</tr>
<tr>
<td>5</td>
<td>Moshkin, et al. [8]</td>
<td>Calcified craniopharyngioma</td>
<td>Nonfunctioning adenoma (subtype 3)</td>
</tr>
<tr>
<td>6</td>
<td>Prabhakar, et al. [10]</td>
<td>Foci of squamous cell rests</td>
<td>Hormone profile not available; acromegalic features</td>
</tr>
<tr>
<td>8</td>
<td>Wheatley, et al. [12]</td>
<td>Cystic</td>
<td>Prolactinoma</td>
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Table 1: Reported cases of pituitary adenoma with craniopharyngioma.
days later did not reveal any residual tumour (Figures 5 and 6). Five days later, he underwent transsphenoidal excision of pituitary adenoma. The tumor was pinkish, soft and vascular.

Histopathology of the excised third ventricular revealed anastomising epithelial islands with palisading layer of epithelial cells. There were focal areas of calcification, and cystic degeneration (Figure 7). These findings confirmed it to be a craniopharyngioma. Pituitary tumor revealed typical adenoma, while immunohistochemistry confirmed it to be growth hormone-secreting adenoma (Figure 8). Patient was discharged ten days later and referred for postoperative radiotherapy. During the course of follow-up over the past three years, he has remained neurologically static. Vision had not improved.

**Discussion**

Multiple intracranial tumors are often considered to be part of neurofibromatoses mosaic, when one of the tumors is a vestibular schwannoma. Tumors occurring at the same site or adjacent anatomical sites may differ on histopathology while retaining the original cell lineage. During embryogenesis, intricate and precise neuroembryological events take place in the sellar region. Both a craniopharyngioma and the pituitary gland share the same embryonic origin, that is Rathke's pouch, which is a diverticulum of the roof of the embryonic oral cavity. The adenophyphyseal is formed from the oropharyngeal duct, while craniopharyngomas arise from the embryological cell rests of an incompletely involuted hypophyseal-pharyngeal duct [1]. These rests locate on the pituitary stalk anywhere from the tuber cinereum to the pituitary gland. Craniopharyngiomas intrinsic to the third ventricle are thought to arise from tuber cinereum and grow upward into the ventricle [2,3]. Such a tumor arising from...
the roof of third ventricle is distinctly rare. Both, pituitary adenoma and craniopharyngioma are ontogenetically related, and there is documented evidence of presence of squamous nests with metaplastic potential in the pars tuberalis. A metaplastic change in pituitary adenoma following irradiation can also explain the rare coexistence of pituitary adenoma and craniopharyngioma in the same tumor [4-6]. Only nine well documented cases of coexistence of pituitary adenoma and craniopharyngioma have been reported so far (Table 1) [1,3,4,7-13]. Only in one report, the two tumours were distinct entities [3]. In all others, the two tumor elements were mixed and could be distinctly identified by light microscopy and immunohistochemistry. Interestingly, El-Bilbeisi, et al. [5] reported occurrence of craniopharyngioma in a patient with acromegaly due to pituitary adenoma; the two tumours were separated by a decade.

The histogenesis of synchronous neoplasms demonstrating two different phenotypes is controversial. There may be coincidental meeting of two different tumors derived from independent tumor stem cells, with or without a shared tumorigenic stimuli. On the other hand there may be a composite mechanism by which a single neoplastic clonal proliferation undergoes divergent differentiation either synchronously or metachronously. There may be metaplastic change in the normal or tumorous tissue. Neoplasms showing dual phenotypes can be explained by one of these theories on the basis of morphologic, immunologic, immunohistochemical or molecular criteria. The prognostic significance of such an association will be known only after accumulation of more data and experience with these cases.

The co-occurrence of craniopharyngioma and pituitary adenoma appears to be a case of ontogenetically related tumors that manifested simultaneously.

Conclusion

Endocrinologically active pituitary adenoma with a pure intra-third-ventricular craniopharyngioma is a rare occurrence. Such an association necessitates precise planning after prioritization of the lesion to be excised. The long-term results are no different from that of the individual lesions.

References