CASE REPORT

Craniopharyngioma and epidermoid tumour in same child: a rare association

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neha.singh.dr@gmail.com**SUMMARY**

Simultaneous occurrence of histologically different primary brain tumours is rare, and its preoperative diagnosis is still challenging. The explanations for the simultaneous occurrence of different primary intracranial tumours in the absence of phacomatoses or prior radiation exposure are at present hypothetical, and these tumours could be simply coincidental. Herein, we report a case of a boy presenting with features of raised intracranial pressure and right-sided sensorineural hearing loss. Brain MRI revealed two different neoplastic pathologies at different sites: an intrasellar and suprasellar craniopharyngioma and a right cerebello-pontine angle epidermoid. To the best of our knowledge, this is the first report in literature of a

craniopharyngioma coexisting with an epidermoid, in the same individual.

BACKGROUND

Most primary intracranial neoplasms occur as solitary lesions. Presence of a single tumour at multiple sites in same individual or even collision tumours have been described only in a few patients. With the advancement in neuroimaging techniques, more such synchronous tumours are being identified. The most common combination identified till date is meningioma associated with glioma. Our understanding of multiple, dissimilar intracranial

Figure 1 MRI brain axial scans show a well-circumscribed lesion in the right cerebello-pontine angle displaying heterogeneously hypointense signals on T2 fluid attenuated inversion recovery, (A) hyperintense on T2WI (B), hypointense on T1WI (C) and restriction on diffusion weighted imaging (D) consistent with epidermoid.

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tumours will gradually improve as more cases are identified, reported and studied.

CASE PRESENTATION

A 12-year-old boy presented with progressively worsening headache, episodic vomiting for 3 months, diplopia and blurring of vision for last 1 month. Neurological examination revealed hypothalamic dysfunction with significant cognitive impairment. No evidence of motor or any other sensory deficit was identified. Routine laboratory investigations were consistent with hypopituitarism.

INVESTIGATIONS

Brain MRI revealed a well-defined non-enhancing cystic lesion in the right cerebello-pontine (CP) angle. The lesion was hyperintense on T2WI, hypointense on T1WI, heterogeneously hypointense on T2 fluid attenuated inversion recovery and showing restriction on diffusion weighted imaging diagnostic of the epidermoid cyst. The lesion was causing encasement of VII–VIII nerve complex and basilar artery with mild distortion of fourth ventricle (figure 1).

A well-defined solid cystic lesion was also noted in intrasellar and suprasellar region causing marked widening of sella with erosion of clinoid processes and thinning of the roof of sphenoid sinus. The cystic areas were hyperintense on both T2 and T1WI and solid areas were showing heterogeneous postcontrast enhancement (figures 2 and 3). Foci blooming on gradient

recoiled echo were noted s/o calcifications (figure 3D). The lesion was causing splaying of optic chiasma and compression over third ventricle leading to obstructive hydrocephalus with transependymal cerebrospinal fluid ooze. These MRI findings were consistent with the diagnosis of craniopharyngioma.

Based on MRI findings a preoperative diagnosis of right CP angle epidermoid with intrasellar and suprasellar craniopharyngioma was made and the patient was referred to the neurosurgery department for definitive management.

TREATMENT

The patient was operated in two sittings under general anaesthesia. In first sitting, a right suboccipital craniotomy was completed and a pearly white tumour was excised from the right CP cistern. Part of a tumour capsule adherent to the brainstem was left intentionally. The patient was re-operated after 2 weeks for intrasellar and suprasellar tumour through the right trans-sylvian approach. On exploration a cystic-solid tumour with areas of putty calcification was identified occupying the sella and suprasellar cistern. Gross total excision of tumour was achieved using microneurosurgical techniques. Parts of the tumour adherent to perforating vessels were left. Histopathology of the lesions was consistent with epidermoid and adenomatous variant of craniopharyngioma, respectively (figure 4).

The patient was discharged 3 weeks after the second surgery with significant improvement in headache, vomiting and diplopia.

Figure 2 MRI brain axial scans show a well-defined solid cystic lesion in the intrasellar and suprasellar region. The cystic part of the lesion is hyperintense on T2 weighted images (WI) (A), T2 fluid attenuated inversion recovery (B) and T1WI (C). Solid portion of the lesion is showing enhancement on contrast enhanced T1WI (D). Right cerebello-pontine angle epidermoid is also visible showing no enhancement.

Figure 3 MRI brain sagittal T2 weighted images (WI) (A), contrast enhanced T1WI (B) and coronal contrast enhanced T1WI (C) shows craniopharyngioma (arrow head) and epidermoid (empty arrow) in same sections. Axial gradient recoiled echo image (D) shows calcifications (solid arrow) within the craniopharyngioma.

DISCUSSION

Multifocal primary lesions of a single histological type and multiple metastatic lesions of the brain have been published in the literature. However, the simultaneous occurrence of multiple primary brain tumours with different cell types is rare with the reported incidence of 0.3% and their occurrence is not well understood.¹ Even rarer is the occurrence of histologically different multiple primary brain neoplasms in paediatric population. The most common tumour associated with the simultaneous presence of other histologically different tumour is meningioma. The most frequently described combination is meningioma associated with glioma followed by schwannoma and by pituitary adenoma.²⁻³

Gironimi *et al*⁴ reported the case of a primary central nervous system lymphoma in a child 6 months after extirpation of an astrocytoma, emphasising the concept of collision tumours and their differential diagnosis. Coexistence of oligodendroglioma with anaplastic astrocytoma,⁵ juvenile pilocytic astrocytoma,⁶ pleomorphic xanthoastrocytoma⁷ and metastatic breast carcinoma⁸ has been described in published literature.

Poeta *et al*⁹ reported three different cases describing simultaneous occurrence of meningioma with glioblastoma, pituitary adenoma and acoustic schwannoma.

Simultaneous occurrence of multiple brain tumours in the same individual is usually associated with phacomatosis or previous irradiation. A number of possible explanations are given for

Figure 4 (A) Histopathological examination of the cerebello-pontine tumour shows the cysts filled with packed, concentric lamellae of stratified squamous epithelium containing keratohyalin granules (black arrow heads). The white matter adjacent to the cyst was hypercellular and contained reactive astrocytes (white arrow heads). (H&E, $\times 100$). (B) On histopathological examination the intrasellar and suprasellar tumours appear as islands of epithelial cells with palisading of basal nuclei containing a calcified keratin core (black arrow heads). (H&E, $\times 20$).

these rare associations. Russel and Rubinstein¹⁰ made the hypothesis that the first tumour provides a stimulus for the surrounding brain parenchyma or meninges to induce a new tumour in a different tissue. Alexander¹¹ proposed that residual embryonic structures form the basis for the development of subsequent multiple brain tumours. Some authors held carcinogenic stimuli responsible for the simultaneous development of different brain tumours¹² and other attributed it as entirely coincidental.¹³

However, based on our case and published literature, we assume that increased research work could provide significant understanding in the development of multiple primary brain tumours.

Learning points

- Syndromic association should be ruled out in patients of histologically different multiple brain tumours.
- Most common primary brain tumour associated with other histologically different brain tumours is meningioma.
- Thorough clinical and imaging studies are mandatory in these cases to provide optimal treatment.

Contributors I myself have prepared the manuscript under guidance of AP and RS. DKS is the clinician involved in management.

Competing interests None.

Patient consent Obtained.

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