CRANIOPHARYNGIOMA DURING INFANCY

(A Case Report)

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Introduction

Craniopharyngiomas are the commonest non-glial tumours during childhood. Indian reports show a prevalence of 3.9 per cent of all intracranial tumours, forming 12-16 per cent [1] of tumours in childhood. A bimodal age distribution is seen in these tumours at presentation, with the first peak between 5-10 years and the second between 55-60 years of age. Tumours may rarely develop during foetal life, becoming clinically apparent during neonatal period or during infancy, when they appear to have uniformly poor outcome. One child with suprasellar craniopharyngioma diagnosed at the age of 4 months is reported. The relevant literature is briefly reviewed.

Case Report

A 4-month-old female child was admitted with one month history of inability to follow objects. Parents suspected visual deterioration in the child. There was no history of seizures, altered sensorium or vomiting. Clinical examination revealed a chubby child, with a large head, tense anterior fontanelle and right abducens paresis. Fundoscopy revealed bilateral optic atrophy.

Cranial CT scan revealed a large mixed density, partially cystic suprasellar craniopharyngioma with hydrocephalus (Fig 1). Right ventriculoperitoneal shunt was inserted to relieve hydrocephalus. Three weeks later, she was taken up for excision of the tumour. Right frontal craniotomy was done and the tumour approached subfrontally. The tumour was seen to occupy the suprasellar region, stretching the optic nerves and chiasma. The tumour was partially excised through the right opticocarotid corridor. Part of the tumour in the region of hypothalamus was left in place, and wound closed. Pre-, peri- and post-operative corticosteroids (dexamethasone and hydrocortisone) were administered.

Post-operative period was hectic, with high grade fever and episodes of bradycardia. Child had polyuria, and remained drowsy after surgery. Her serum electrolytes were normal. Fever and drowsiness persisted and she expired on the seventh post-operative day. Request for autopsy was refused by the parents. Histopathology of the tumour confirmed craniopharyngioma with areas of cyst formation and calcification (Fig 2).

Discussion

Craniopharyngiomas arise from embryonic

squamous cell rests on an incompletely involuted hypophyseal-pharyngeal duct, and can occur in the pituitary stalk extending from tuber cinereum to the pituitary gland. These tumours grow in size by cellular proliferation and secretion, forming cysts, and excite an intense astroglial reaction in the hypothalamus as they grow into the later [2]. As the tumour grows, it

Fig. 1 : CECT brain showing large suprasellar craniopharyngioma.

Fig. 2 : Histopathology (X 100, H & E) showing squamous epithelium, cystic spaces, pearls and calcification.

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usually impinges first on the optic system. In infancy, the resultant visual deficit is easily missed until it is marked and has grown large [3]. These tumours are slow growing and are rarely seen in children younger than 5 years of age [4]. In the pre-CT era, these tumours were rarely diagnosed in small children. Iyer [5] reported the first case after autopsy of a 3 month old child who had hydrocephalus at birth. Gass [6] reported first case who had been diagnosed ante mortem. Subsequently, craniopharyngioma during infancy was reported by Madj et al [7] and Tabaddor et al [8]. All these tumours appeared calcified on skull radiographs.

Even with the diagnostic and therapeutic explosion in the post-CT period, there are very few reports of craniopharyngioma during neonatal period and infancy, underscoring its rarity. In a series of 100 infants with intracranial tumours, Tomita and McLone [9], and in another series Timothy and Benjamine [10], there was no child with craniopharyngioma. Zuccaro et al [11] found only one craniopharyngioma in their series of 40 infants with intracranial tumours. Wakai et al [12] reviewed 200 cases of intracranial tumours within 2 months after birth and reported only 9 craniopharyngiomas. From India, Venkatramna et al [13] reported one 5 month old child with craniopharyngioma.

Although recent advances in pre-operative, peri-operative and post-operative endocrine management as well as advances in surgical techniques have improved long term survival in children with intrasellar and suprasellar neoplasms, prognosis of craniopharyngiomas detected during neonatal period and infancy remains poor [6-8,13]. It is likely that the neonatal tumours are aggressive and of fast growing type, and infiltrate the hypothalamus without exciting astroglial reaction to the extent seen in older children and in adults. Dissection in the region may lead to hypothalamic and vascular damage. In the post-operative period, these children develop electrolyte imbalance, hyperosmolality, leading to death [13]. While the microneurosurgical

techniques have been standardised, peri-operative endocrine and electrolyte monitoring is crucial in these infants for improving survival. Autopsy studies may throw more light on hypothalamic infiltration and damage due to tumour, and a lot yet remains to be learnt about craniopharyngioma during neonatal period and infancy before their prognosis and surgical results can be brought at par with those in older children and adults with craniopharyngioma.

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