Pan Hypopituriarsme and Central Diabetes Insipidus Secondary to Craniopharyngioma

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Letter to the Editor

Craniopharyngioma (CP) is a rare solid or mixed cystic epithelial tumor in the sellar and suprasellar region, Patients with craniopharyngioma frequently exhibited hypothalamic–pituitary axis dysfunction, including growth hormone deficiency (GHD), adrenocortical insufficiency, central hypothyroidism, hypogonadism, precocious puberty, hyperprolactinemia, central diabetes insipidus, and hypothalamic obesity [1]

CP accounts for 1.2-4.6% of all intracranial tumours, with a prevalence of 0.5 -2.5 new cases per 1 million people per year world wide [2]. Craniopharyngiomas can present at any age but usually exhibit a bimodal age distribution, the first peak occurring in childhood between 4–15 years and the second peak during adulthood between 40–79 years [3]. The male to female ratio is 1.1–1.4:1 [4] CPs have the highest mortality of all pituitary tumours and women are more affected than men [5].

Here we report the case of a 25-year-old man, who presented to the neurosurgical emergency room with worsening headaches, impaired vision, polydipsia and polyuria. He drank approximately 10 liters of fluids per day; every 20 minutes during the night. Further, his vision was blurred, and he was vomiting twice per week. He also had features of raised intracranial pressure for over 2 months. Initial non-contrast computed tomography (NCCT) revealed cerebral oedema with hydrocephalus due to a mass lesion around the 3rd ventricle, suggestive of a colloid cyst, which required urgent decompression via a ventriculo-peritoneal (VP) shunt. He underwent an MRI brain with pituitary protocol revealing a cystic suprasellar mass with well-defined margins measuring 2 cm in size with chiasmal compression, suggestive of a craniopharyngioma . Moreover, endocrinological examination revealed complete panhypopituitarism including deficiencies of ACTH, TSH, PRL, GH, LH, and FSH. Urine and serum osmolality were suggestive of diabetes insipidus.. Once the diagnosis of panhypopituitarism and central diabetes insipidus secondary to craniopharyngioma was established, hormone replacement therapy was commenced with oral hydrocortisone followed by thyroxine after an interval of 1 week, as well as Desmopressin for diabetes insipidus . Patient referred to neurosurgery for complete care.

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A clinical picture at the time of diagnosis often dominated by nonspecific manifestations of intracranial pressure (eg, headache and nausea). Further primary manifestations are visual impairment (62– 84%) and endocrine deficits (52– 87%). Among adult-onset craniopharyngioma patients, hormonal deficits at the time of diagnosis are much more pronounced when compared with childhood-onset craniopharyngioma patients. Endocrine deficits are frequently caused by disturbances to the hypothalamic–pituitary axes that affect GH secretion (75%), gonadotropins (40%), ACTH (25%), and TSH (25%) [6] [7]. At the time of diagnosis, 40 to 87% of patients present with at least one hormonal deficit , and other endocrine symptoms such as neurohormonal diabetes insipidus are present preoperatively in 17 to 27% of patients [4,6,7].

The imaging workup of CPs and the differential diagnosis from other sellar and suprasellar tumours is based on MRI with an adjuvant role of computerized tomography (CT) in the detection of calcifications [2,4].

Although the standard treatment for craniopharyngioma, including surgical resection and radiotherapy, can achieve local tumor control, active local treatment often declines quality of life due to permanent neuroendocrine and neuroendocrine defects [1].

The objectives of treatment in CPs are: (1) relief of raised intracranial pressure if present;

(2) reverse visual compression symptoms; (3) restoration or substitution for pituitary hormone deficits plus other supportive measures; (4) prevention of tumour regrowth/ progression, while keeping acute and long-term morbidity and mortality as low as possible[2].

CP in adults is associated with significant mortality. The overall survival rates in recent years ranges from 89% to 94% at 5 years, from 85% to 90% at the 10-year follow-up and an average of 62%–76% at 20 years [5].

Although craniopharyngiomas are generally benign their location, size, and tendency to infiltrate adjacent cerebral structures makes their management rather demanding and may lead to sometimes devastating complications [4].

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