MICROSURGICAL EXCISION OF GIANT CRANIOPHARYNGIOMA

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Introduction

Craniopharyngioma is a benign developmental tumor arising from embryonic squamous cell rests of an incompletely involuted hypophyseal-pharyngeal duct. Management of these tumours remains controversial whether they should be treated by radical excision alone, radiotherapy or combination of both [1-4]. Though total excision of these tumours is accepted ideal treatment, it becomes more difficult as the size of tumour increases as this increases operative mortality and morbidity due to its close relation with major blood vessels, visual apparatus and hypothalamus [5].

Giant craniopharyngiomas have been defined by Samii and Samii [5] as a tumour of more than 5 cms in its greatest dimension and any tumour more than 6 cms in one dimension by Yasargil et al [6]. We had one case of giant craniopharyngioma with dimension of 6.0 x 5.5 x 3.5 cms, total microsurgical excision of

which was carried out with good result.

Case Report

25-years-old married lady presented with history of amenorrhea of one year, headache of 6 months, visual obscuration of one month and vomiting of 2 weeks duration. The onset was insidious and course progressive. General physical examination and vitals normal. There were no features of hypopituitarism. Central nervous system examination revealed normal higher mental function. Cranial nerve examination showed visual acuity of 6/18 in both eyes and bilateral papilloedema. Other cranial nerves, motor and sensory systems were normal. Investigations revealed normal blood glucose, hemogram and routine urine examination. X-ray skull showed speckled sellar calcification. CT scan brain showed a large (6.0 x 5.5 x 3.5 cms) partly calcified well enhancing mass in the sellar and suprasellar region reaching above foramen of Monro in vertical axis leading to obstructive hydrocephalus and extending from subfrontal to interpeduncular fossa in anteroposterior axis (Figs 1, 2). Endocrinal work-up revealed hypocortisolemia and mildly elevated prolactin levels (Table 1).

She was taken up for surgery through a right frontal osteoplastic craniotomy and subfrontal approach. Right lateral ventricle was cannulated and cerebrospinal fluid drained and brain became lax. A large well encapsulated intra optic greyish white, partly calcified tumour was found. After initial intratumoural decompression, the

Fig. 1a : Axial contrast enhanced CT scan shows large enhancing mass in the area of third ventricle and length and width of mass

Fig. 1b : CT scan shows mass extending above foramen of Monro into septum pellucidum

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TABLE 1
ENDOCRINAL EVALUATION

Hormone levels	Pre-op	Post-op	Normal
T3	0.94	1.13	0.8-2.1 ng/ml
T4	9.50	8.26	4.5-13.5 µg/dl
TSH	1.19	2.36	0.5-6.5 mIU/L
LH	12.41	< 1.0	5-20 mIU/L
FSH	12.08	5.05	5-20 mIU/L
Basal cortisol	1.46	8.26	5-25 µg/dl
Prolactin	26.0	15.0	0-20 ng/ml

Fig. 2 : CT scan shows vertical extent of mass from sella to septum pellucidum

capsule was dissected off from optic nerves, internal carotid arteries and finally from hypothalamus. The attachment from pituitary stalk was coagulated and incised microsurgically. Homeostasis achieved, dura closed and wound closed. Postoperatively she was fully conscious with normal vision and no focal deficit. She developed diabetes insipidus with urinary volume of 8-10 litres per day with urinary specific gravity of 1004-1006 for 2 days. She was treated with fluid replacement with 5% glucose on first postoperative day and by oral plain water and tablet carbamazepine 200 mg thrice a day and her diabetes insipidus settled completely with urinary output of 1.5-2.0 litres and urinary specific gravity of 1014. She also received perioperatively antibiotics for 24 hours and steroids which was brought down to maintenance dose of 10 mg per day before discharge. In addition she was put on thyroxine replacement therapy with tab eltroxin 0.1 mg per day at the time of discharge. She was relieved of her headache, her visual acuity improved to 6/6 and wound healed primarily. Postoperative scan showed total excision of the tumour (Fig 3 & 4). Histopathology confirmed it to be craniopharyngioma (Fig 5). She was not subjected to radiotherapy and followed up in OPD. She remained asymptomatic with visual acuity of 6/6 in both eyes. At one year after surgery she is relieved of hypocortisolemia and was without corticosteroid and thyroxine replacement but remained amenorrheic. Her postoperative endocrinal evaluation revealed prolactin 15.0 ng/ml, cortisol 8.26 µg/dl, follicular stimulating hormone 5.05 mIU/L, luteinising hormone <0.1 mIU/L, T3 1.13 ng/ml, T4 8.26 µg/dl and TSH 2.3 mIU/L (Table-1).

Figs. 3 & 4 : Post operative CT scan shows total excision of mass in axial and vertical axis

Fig. 5 : Solid epithelial nests with peripheral palisading, alternating with cystic areas. Foci of calcification and shadow cells (haematoxylin & Eosin x 100)

Discussion

Craniopharyngioma is benign tumour of suprasellar region usually arising from pituitary stalk. Sellar region is cross road of major blood vessels of brain, cranial nerves that govern the vision, ocular movements and hypothalamus, the seat of endocrinological and autonomic nervous system. As the tumour grows these structures can be affected, thus making the surgical excision of these tumours a challenge, that requires skills and familiarity with this highly vulnerable area [5].

Radical surgical excision of craniopharyngioma is still associated with significant morbidity and mortality which has lead to alternative treatment opinions such as non radical surgery and radiation therapy [6], stereotactic aspiration [7], stereotactic aspiration and intracavitary brachytherapy with Yttrium 90 [8] and intracavitary chemotherapy with bleomycin [9]. These modalities are associated with higher recurrence rate and radiation induced delayed damage. The ideal treatment for craniopharyngioma still remains total microsurgical excision [4,5,10] as long term results are better and recurrence rates are low. However it may not be feasible to do total excision in all cases. The determinants of total excision are size of tumour, irremovable calcifications, marked adhesions to vital structures, and above all skill and experience of the surgeon [5,10-12]. The good results (totally independent and improved) are 93% in patients with tumour of <2 cms, which falls to 12.5% in giant craniopharyngioma with postoperative mortality rising to 87.5% [10]. In 10 cases of giant craniopharyngioma (with maximum diameter >5 cms) in whom total microsurgical excision was carried 8 were on DDAVP (Desmopressin) for diabetes insipidus and all were on corticosteroid and thyroxine replacement.

We had a case of giant craniopharyngioma (6.0 x 5.5 x 3.5 cms) reaching above the foramen of Monro. We could achieve total microsurgical excision of this giant tumour and our patient was spared of radiotherapy. She is not on replacement therapy with thyroxine or steroids. She is not having diabetes insipidus but continues to be amenorrhoeic due to hypogonadotropic hypogonadism.

For total microsurgical excision a thorough understanding of extent of tumour with the help of imaging

studies and preoperative planning is required. But most importantly the feasibility of total excision will be dictated on the operating table by operative findings after intratumoural decompression, if the capsule can be dissected off the vital structure i.e. carotids, optic nerve and chiasma and hypothalamus, it will be feasible to excise craniopharyngioma completely without significant mortality or morbidity as was possible in our case.

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