CRANIOPHARYNGIOMA AND RARE TUMORS

RARE-01. MANAGEMENT AND OUTCOMES OF PAEDIATRIC CRANIOPHARYNGIOMA: A 15-YEAR EXPERIENCE IN SINGAPORE

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BACKGROUND: Craniopharyngiomas are rare embryonic malformations of the sellar region with high survival rates but high morbidity due to long-term sequelae caused by the location of the tumour. We summarise our institution's experience on the management and outcomes of paediatric craniopharyngiomas in Singapore. METHODS: This was a retrospective review of all paediatric patients (18 years and below) with histologically diagnosed craniopharyngioma managed by the National University Hospital, Singapore from January 2002 to June 2017. Data on clinical presentation, imaging, treatments, and outcomes were extracted from the electronic medical records using a standardized data collection form. Data analysis was conducted using RStudio (Version 1.2.5033). Institutional ethics approval was obtained for the study. RESULTS: We identified 12 cases of paediatric craniopharyngiomas. The majority of cases were male (8, 66.7%) and the median age at presentation was 6.0 (IQR 3.8 – 9.5). Initial surgical management was tumour excision (11, 91.7%) or insertion of a reservoir into the cyst cavity (1, 8.3%). All cases had diabetes insipidus, 10 (83.3%) had endocrine dysfunction, and 8 (66.7%) had visual impairment on long term follow up. 7 (58.3%) cases had recurrence, and 3 (25.0%) had demised. Cox-regression showed that females (HR=33.9, p=0.049), and Chinese race (HR=13.3, p=0.034) were at higher risk for recurrence, but age at diagnosis and residual tumor on post-operative MRI was not significant. CONCLUSION: The management of craniopharyngioma is complex as it is complicated by high recurrence rates and significant long-term morbidity. Further research on treatment strategies focusing on maintaining quality of life is important.

RARE-02.RE-IRRADIATION FOR RECURRENT CRANIOPHARYNGIOMA

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PURPOSE: Patients with recurrent craniopharyngioma after radiotherapy (RT) have few treatment options. At our institution, re-irradiation has been offered to selected individuals with recurrent craniopharyngioma not suitable for further surgery, intracystic therapy or targeted agents. METHODS: A retrospective study was performed of patients with craniopharyngioma treated with two courses of fractionated RT. First RT (RT1) prescriptions ranged from 50–54 Gy in 25–30 fractions; re-irradiation (RT2) prescriptions were 54 Gy in 30 fractions with full, in-field overlap of dose. The maximum dose to organs-at-risk (brainstem, optic structures) were maintained at or below the prescription dose. There was no cumulative dose limit to any structure. RESULTS: We identified four patients. Median RT1-to-RT2 interval was 5.8 years (range, 4.7–20.4). Cumulative maximum doses to optic chiasma and nerves were >100 Gy in all four patients. With a median follow-up of 33 months after RT2, three patients had disease control and are alive at 9, 23 and 42 months from RT2; one patient developed progressive disease and died 33 months after RT2. In three evaluable patients, vision remained stable or improved after RT2; the remaining one patient had no light perception prior to re-irradiation. Two patients had neuropsychological testing before and after RT2; neurocognitive domains were generally stable in one patient but working memory declined in the second patient. CONCLUSIONS: Despite exceeding usual tolerances for optic chiasm and nerves, visual outcomes were stable in all living patients. Re-irradiation should be discussed as a treatment option for patients with recurrent craniopharyngioma but without other therapeutic options.

RARE-03. AGGRESSIVE RESECTION FOR PEDIATRIC CRANIOPHARYNGIOMAS VIA ENDOSCOPIC ENDONASAL APPROACH

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OBJECTIVE: In recent years, the endoscopic endonasal approach (EEA) has been increasingly used for pediatric craniopharyngiomas. We here present our experience and the outcomes of the EEA resection of pediatric craniopharyngiomas. MATERIALS AND METHODS: Between April 2014 and December 2019, 16 cases of pediatric craniopharyngiomas were operated at the Osaka city university (OCU) hospital. Eight patients were diagnosed with primary craniopharyngiomas while 8 had a recurrent tumor. There were 5 males and 11 females, with a mean age of 10.7 years (3–17 years). EEA was selected in all patients and a case of large mutilobulated tumor was resected by combination of microscopic transcranial approach. RESULTS: Gross total resection was achieved in 14 patients and near total resection in other 2. Post-op CSF leak occurred in 3 patients, which was treated with re-exploration. Pituitary stalk was preserved intraoperatively in 4 cases, and 15 patients developed diabetes insipidus and anterior hormonal replacement therapy was required in 15 patients at last follow-up. Visual improvement was noted in 4 patients while vision remained unchanged in the rest. Neuropsychological function status was preserved in all patients, and there was no new-onset obesity postoperatively. The mean follow-up duration was 35.1 months (2 – 69 months) and 4 of 8 recurrent cases had re-recurrence during this period, however there was no recurrent in 8 primary cases. CONCLUSIONS: EEA should be the surgical modality of choice for treating pediatric craniopharyngiomas. It results in better visual and cognitive outcomes with a significantly increased extent of resection.

RARE-04. INTELLECTUAL DEVELOPMENT IN CHILDREN WITH PEDIATRIC CRANIOPHARYNGIOMA AFTER TUMOR REMOVAL

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INTRODUCTION: Intellectual assessment in children with craniopharyngioma after tumor removal is still unknown. We assessed intellectual development in children who underwent microsurgical resection in our institute over the last twelve years. MATERIALS AND METHODS: Ten children among 41 patients with craniopharyngioma treated and followed at Kagoshima University Hospital between 2007 and 2019 were reviewed. We also assessed intellectual development in 10 years or younger children with craniopharyngioma one year after tumor removal. Intelligence was assessed using the Wechsler Intelligence Scale for Children-Fourth Edition (WISC-IV). RESULTS: Ten children underwent microsurgical tumor removal. The mean age at surgery was 5.8 (range 1–10) years. Transcranial approach was performed in 8 children, transsphenoidal approach in two children. The mean follow up period was 110 months. Gamma knife surgery (GKS) was performed in 6 children less than 6 months after first surgery. Regional recurrences occurred in 5 children, and additional GKS was performed in four children, second microsurgical removal in one child. Severe obesity with a transient electrolyte imbalance occurred in one child. Eight children with GH deficiency underwent GH replacement therapy. Eight children were assessed working memory index (WMI), processing speed index (PSI), Perceptual reasoning index (PRI), and verbal comprehension index (VCI) using WISC 4. Each mean value of WMI, PSI, and PRI was lower than VCI, except for 2 children with normal full scale intelligence quotient. CONCLUSION: WMI, PSI and PRI in children with intellectual disabilities were lower tendency than VCI after surgical removal of craniopharyngiomas in the present study.

RARE-06. OPTIMIZATION OF PROTON RADIATION THERAPY FOR GIANT CRANIOPHARYNGIOMAS

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Craniopharyngiomas are benign intracranial tumors located in the sellar and suprasellar region. Their size and extent of invasion into surrounding structures vary considerably. While the majority of craniopharyngiomas on presentation are between 1–3 cm without hypothalamic invasion, a significant proportion of patients present with 'giant' craniopharyngiomas of >4cm in dimension with large cystic extension through the 3rd ventricle. These tumors pose a challenge both for surgical resection as well as for radiation therapy. Proton beam therapy (PBT) has become the preferred standard of care after subtotal resection of pediatric craniopharyngiomas. In the setting of giant craniopharyngioma, the use of proton therapy allows a reduction of dose to surrounding normal brain, but changes in cyst volume can result in either under-coverage of tumor or excess dose to surrounding brain, an effect further magnified by the sharp gradients associated with proton dose distributions. In this case report we describe the proton treatment planning and intra-treatment monitoring of two patients with giant craniopharyngiomas with largest pre-operative of dimension 6cm, and 9cm, respectively, and 6cm and 5.5cm, respectively, pre-radiation. Both patients had drains inserted to Ommaya reservoirs. We performed surveillance imaging during RT utilizing spiral computer tomography (CT) on a weekly basis and reconstructed the treatment dose on the surveillance CTs to ensure target coverage and normal tissue sparing. We compared the dosimetry in