Original Research Article

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Paediatric Craniopharyngiomas: Our Experience of 27 Cases

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Abstract

Introduction: Craniopharyngiomas are benign neoplasms of the sellar/suprasellar region. Management of these lesions always poses a challenge to neurosurgeons.

Methods: Between 2012 and 2016, a total of 27 paediatric patients (8-16 year age) underwent surgery for craniopharyngiomas with a goal of total surgical removal without neurological risk. All patients were operated via frontopterional approach under operating microscope. Radiotherapy was offered to 7 patients in which partial decompression and excision was done.

Results: Majority of the cases (21/27) 77% were large tumours with suprasellar extension and both supradiaphragmatic and infradiaphragmatic components. Complete resection was achieved in 13/27 (48%) of the cases. Neuropsychological sequele were seen in six children. Three children had memory deficits and behavioural problems (aggressiveness and lack of attention) and 3 had change in personality.

Conclusion: We conclude that radical resection of craniopharyngiomas with perseveration of the pituitary stalk should be the goal of surgery. However; one should not be overly aggressive in approach for these tumors as radiotherapy also has a proven role in their management.

Keywords: Paedriatic Craniopharyngioma; Resection; Radiotherapy.

Introduction

Craniopharyngiomas are a type of histologically benign neoplasms which typically arise in the sellar / suprasellar region. Although they occur in adults too, they are frequently seen in paediatric age group having a bimodal peak of incidence. Management of these lesions always poses a challenge to neurosurgeons due to a variety of reasons. The ideal management should be complete tumour removal with improvement of altered visual functions, minimal deterioration of endocrinological function, and no neuropsychological impairment. We present our experience in dealing with these lesions in paediatric age group.

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Clinical Methods and Materials

Patient Demographic Data

Between 2012 and 2016, a total of 27 paediatric patients underwent surgery for craniopharyngiomas. Most of the cases were in the 8-16 year age group with only 7 cases younger than 8 years of age. Mean age was 12. Sixteen of these cases were males and 11 females. We did not include previously operated cases (both our centres or outside) in our study.

Signs and Symptoms

Headache was the commonest symptom noted in our patient population .It was reported in almost 80 percent of the cases. The severity of headache varied with 3 cases having severe debilitating headache. Signs of raised intracranial tension were noted in 7 cases and 4 of them were drowsy at admission. Our assessment of neuropsychiatric status of the patient population included work up for developmental milestones, a simple mental function exam (age adjusted) and assessment of memory. Four cases that

were drowsy at admission could not be assessed. Out of the 23 cases, 13(57%) were found to be having normal mental development. Six cases had memory deficit with slow response times. Four children had complex psychiatric symptoms. Motor symptoms in the form of difficulty in walking and ataxia were found in 3 children. Cranial nerve involvement was assessed in all cases and found to be positive in 14 cases with optic nerve being the most common followed by 7th and 5th nerves. One case had multiple lower cranial nerve involvement.

Visual defects were seen in about 74 percent of the children. The defects varied from decreased visual acquity in 20 cases (74%) and visual filed defects in 16 cases (60%). Perimetery was done preoperatively in all cases. Severe b/l visual acquity loss (<1/20) was noted in 7 cases (25%)with two children being completely blind at presentation. Fundoscopy was done in all cases. Papilloedema was noted in 9 cases (33%) with optic atrophy seen in 5 cases.

Endocrine abnormalities were seen in almost 60 percent of the children (16 cases). Preoperative hormone analysis was done in all cases and was normal in only 6 children. Eleven children had short stature and 3 children were severely malnourished. Pan hypopitutarism was seen in 3 cases (10%). Development of sexual characters was assessed in the teenage group and it was found that out of the 11 children in 13-16 age group only 2 had normal development of secondary sexual characteristics. Both were males.

Imaging Characteristics

All cases underwent CT as well as MRI imaging. Imaging revealed calcifications in almost 80 percent of the cases. Classification of tumours was grossly done into predominantly cystic, predominantly solid and mixed tumours. Out of the 27 cases, 21 (77%) were found to be mixed density tumours while 4 were predominantly cystic and two were solid. Nine patients had giant tumours with multiple lobe and large suprasellar extensions. Hydrocepahlus was noted in 9 cases (33%). The preoperative diagnosis was craniopharyngioma in all but three cases in which the differentiation from hypothalamic glioma /pituitary macroadenoma could not be done preoperatively

Treatment

Surgery

All patients in our study group were taken up for surgery with the goal of total tumour removal without neurological risk. The decision on extent of tumour resection was taken intra operatively by the surgeon based on the anatomical characteristics of the tumour. Preoperative shunting was done in 13 cases and in two cases postoperatively. All patients were operated via frontopterional approach under operating microscope.

Radiotherapy

We did not offer radiotherapy to patients who had complete excision / incomplete excision primarily. It was only offered to 7 patients in which partial decompression and excision was done. However, it was advised to one patient who had a recurrence after complete resection and three patients who had a recurrence after incomplete resection in follow up period.

Results and Analysis

Surgical Nuances

Precise knowledge of the anatomy of the region is a prerequisite in successfully operating these tumours due to close proximity of critical neuro vascular structures like the optic chiasm, pituitary stalk and the third ventricle. It is very important that the relationship of the tumour with the diaphragm sella is judged preoperatively and confirmed intraoperatively. This is required for the preservation of the pituitary stalk. In our series only one case had a tumour small enough to be completely infradiaphragmatic intrasellar. Majority of the cases (21/27) 77% were large tumours with suprasellar extension and both supradiaphragmatic and infradiaphramatic components. However, the diaphragm sellae itself was seldom involved in these cases.

The optic chiasm is also a very important landmark for guiding surgery in these cases. These tumours can grow around in either direction. In our series, most of the lesions (70%) had expanded to the retro chiasmatic space and were not properly accessible from the interoptic corridor. We found that opening of the lamina terminalis is really helpful in these cases, even when there is no intraventricular extension. The extra relaxation provided help in delineating the anatomy better.

Ventricular extension depends on the site of origin of these tumors and the stage of presentation. Intraventricular tumors can attain large size before presentation. In our series 19/27 (70%) of the cases were extraventricular. Six cases had an

intraventricular extension with all of them involving the floor of the third ventricle and pushing it upwards. Only two cases were purely intraventricular within the third ventricle.

Tumour invasiveness is also best judged intraoperatively. We found that the ease of separation of the tumour from the brain parenchyma varied considerably. In majority of the cases the solid enhancing part was adherent to critical structures like the ventricular walls and floor, optic chiasm or the brainstem.

In our series, the cases in which we were able to achieve good separation plane between the tumor and brain parenchyma allowing complete macroscopic resection of the lesion, were considered as complete resection. This was achieved in 13/27 (48%) of the cases. We attempted to preserve the pituitary stalk in every case. The resection was considered incomplete when after gross macroscopic resection, we were unable to separate the tumour from crictical neurostructures like the stalk and a clear plane cud not be established. This was done in 7/27(26%) of the cases. The invasiveness of these cases prevented their removal without resection of the parenchyma. In seven cases, decompression and partial removal was done with macroscopic fragments being left. We found that calcified part in these lesions was the most difficult to remove. Excessive traction applied during removal can lead to devastating consequences in these cases. Despite our attempt to preserve the stalk in all our cases, it was resected in two cases.

Post Operative Neuroimaging

All patients underwent contrast enhanced CT scanning with 24 hrs of surgery. We found no residual tumor in 16 cases (60%) of the cases. Two patients had postoperative hydrocepahalus. One patient had a post operative hematoma.

Post Operative Course

Post operative stay was uncomplicated with average in hospital stay of 14 days in 17 cases (62%). Complications, both medical and surgical were experienced in 7 (26%) cases with 3 mortalities. Second surgery was required in four cases. Two were taken up for shunt surgery on 2nd post operative day, one had to operated for epidural hematoma removal on first post op day and one case had to be undergo bone removal due to infection. Three cases had prolonged ICU stay with stormy post operative course due to electrolyte disturbances. However, all cases were discharged within one month of surgery.

Surgical mortality was 3 out of 27 cases. One was a severely malnourished child with large calcified tumour who died on 5 th postoperative day due to hypothalamic failure. Second was 13 year child with large calcified tumor with intraventicular extension who died on 17 post op day again due to hypothalamic failure. We lost one patient to infection. This was an eleven year old child who developed meningitis on 7th post operative day and eventually septicaemia from which he never recovered.

Follow up Data

Out of the 24 surviving cases, 23 have been followed up for a minimum of three months. Six cases have been followed up for more than 3 years and 11 cases for more than a year. Five year follow up is available for two cases.

Overall tumour related mortality was 18.5 %(5/ 27). Three cases were lost in the post operative period. Two children died in follow up period within the first year. Both of them had tumour recurrences and rapid growth and these were cases in which only partial removal and decompression had been done. Complete removal was adjudged to be almost impossible in these cases.

Recurrence Data

Recurrence most strongly depends on extent of resection and tumour characteristics.

Out of the thirteen cases in which complete resection was achieved, 7 cases are recurrence free at one year and 2 cases at three year follow up. Four cases out of this group have shown recurrences. However, clinically these patients are stable at present and none of the recurrences warrant any intervention at this stage. They are being monitored and one of them has been referred for radiotherapy.

Out of the group in which incomplete resection was done, we are dealing with recurrences in 2 of the 7 cases (29%). However, most of these cases are at one or three year follow up stage at present. These group of patients were not referred for radiotherapy after primary surgery. The three cases with recurrences in this group have been offered the option of radiotherapy.

The third group of patients in which partial removal was done were sent for radiotherapy after primary surgery. Residual tumour was identified in all cases in the post op period. Out of 7 patients in this group, 2 died in the follow up period within the first year. Four cases have undergone radiotherapy and are clinically stable at two to three year follow

up. One of them had to be re operated 9 months after primary surgery for debulking.

Functional Outcome

This is the most important aspect of management of paediatric craniopharyngiomas.

Neuropsychological Outcome

Out of the 27 operated cases, 20 were in the 8-16 age group and 7 were below 8 years of age. 57 % of the cases(13/23) had normal mental development at admission. Post operatively, 3 children had memory deficits and behavioural problems in the form of aggressiveness and lack of attention and 3 had change in personality. They were able to attend school but were lacking in social interaction. However, these changes subsided after a year and at two year follow up all but one child had stabilized and achieved preoperative status.

Radiotherapy was done in 3 cases with normal mental status at admission and it was not found to have any adverse effect psychologically in the patient. However, one child did have anorexia which was managed by psychotherapy.

Six children presented to us with memory deficits and slow mentation .The symptoms improved in 3 of these cases post surgery.

In the 13 children with normal mental development, we were able to achieve complete resection in 7 cases, incomplete resection in 4 and partial decompression in 3 cases.

Neurological Outcome

Motor weakness improved in all cases with preoperative deficit. Degree of improvement varied. One patient had new post operative motor deficit which resolved in two months. Cranial nerve paresis was observed in three cases which improved over a period of six months. Patients who had preoperative cranial nerve involvement showed marked improvement in symptoms at six months follow up.

Endocrinological Status

We tried to preserve the pituitary stalk in all cases. However, it was resected unintentionally in two cases, resulting in panhypopituitarism. We lost one of these children on 5th post op day. Panhypopituitarism was seen in 6 (22%) cases, partial deficiency in 9 cases (33%) and no deficiency was seen in 8 cases in postoperative period. Out of the 8 cases with no post op defieciency, 6 had undergone complete resection and two had undergone partial removal. Children who had preoperative hypoptituitarism showed post operative improvement in hormonal levels in all but one case. At one year follow up, almost 50% of the cases showed improvement with stabilization of hormonal levels without external therapy.

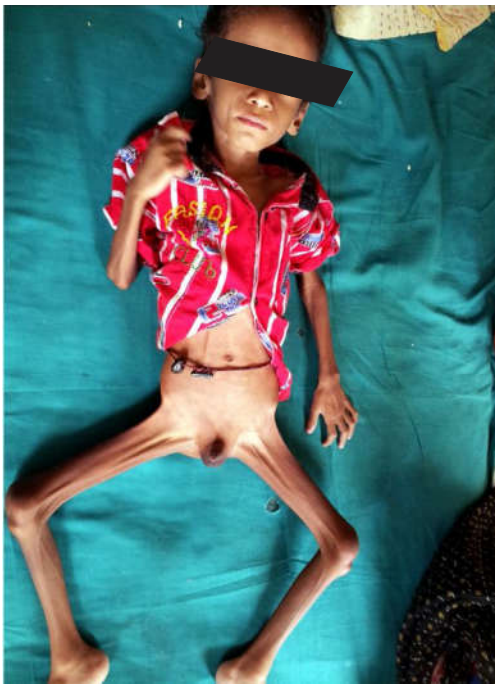

Image 1(a): Malnourished child having craniophayngioma with hypothalamic involvement

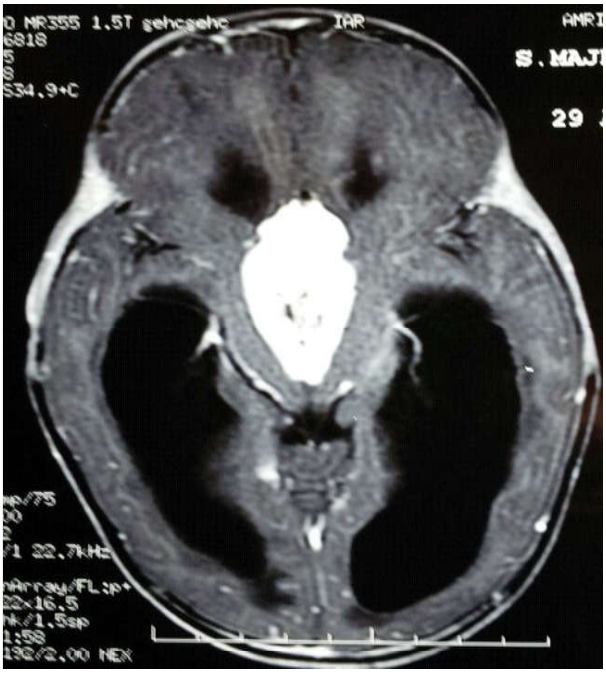

Image 1(b): Contrast MRI of the patient showing large, predominantly solid craniopharyngioma with hydrocephalus

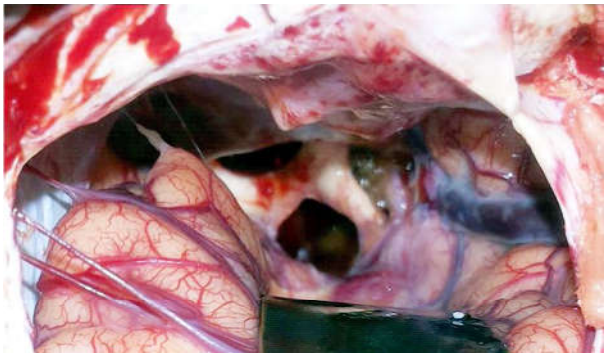

Image 2: Intraoperative photograph showing complete removal of craniopharyngioma in a patient

Visual Status

Surgery for these tumors had a positive impact on the visual status of 70% of the children with improvements noted in both acquity and visual fields. Children who had severe b/l acquity loss preoperatively showed minimal improvement.

Social Acceptance

Out of the 24 surviving cases, 4 presented to us in a drowsy state and 7 had severe b/l vision loss. All of these children were able to integrate into society and were able to adjust according to their disability.

Four children in the 12-15 age group required special caring due to persistent hormonal problems and neuropsychological impairment.

Patients who had severe psychiatric problems in the preoperative period were not helped by surgery for these symptoms.

Thirteen children (48%) were able to develop normally with normal school going status till 1.5 year follow up on average.

Discussion

Paradigm of Management

As more and more research has come forward into these tumors and neurosurgical techniques have advanced, broad consensus on the need for surgery in these has emerged (1, 2, 3, and 4).

Surgery in these lesions, especially in the paediatric age group has many benefits like confirmation of diagnosis, decompression of critical neurovascular structures which may prevent development of neuro deficit and even reverse it and delay recurrence. These tumours are many times cystic and have significant mass effect.

Preoperative shunting in these tumours has been a controversial topic especially in children. Many authors have tried to emphasize that shunt insertion should be avoided [5,6]. In our rather short experience, we believe that children who present with gross hydrocephalus, the benefits of early shunt insertion outweigh the risks. However, if the child can be taken up for early definitive surgery, it can be avoided. Unfortunately, in developing countries like India, early surgery cannot be guaranteed especially in govt. sector due to various socioeconomic factors. We did not find any significant difference intraoperatively in tumour resection surgery in shunted and non shunted children.

Choice of Surgery

The frontopterional approach is the most favoured approach among most authors [1, 2, 3, 5, 7]. The best part about this approach is the versatility it provides to the surgeon in approach and resection. Splitting of sylvian fissure can be done as required which greatly expands the view of the retrochiasmatic extension of the tumour. All critical structures are in direct view of the surgeon which lowers the chances of inadvertent injury. Easy approach to the lamina terminalis is another advantage. We did a right sided approach in 90% of our cases . Preoperative study of the imaging helps in appropriate planning and choosing the best corridor of approach.

Extent of Resection

This is the point of contention among neuro-surgeons with many advocating a more conservative approach towards these lesions [8]. In our experience, it is very difficult to go in for a craniopharyngioma surgery with a fixed frame of mind. Of course, the goal is always complete removal; however, intraoperatively if the surgeon feels that the tumor is densely adherent or very invasive, we might end up doing a lot of harm in achieving that goal.

Tumor invasiveness, locations, size, presence of calcification are all factors which influence the extent of safe resection. The most important factor is tumor invasiveness and also the one which cannot be reliably judged on preoperative imaging [9]. In many series, complete resection has been defined as removal of all involved part including pituitary stalk and infundibulum However, we feel that these structures should not be sacrificed [10]. The problems arising out of resection of these structures outweigh the benefits. Complete resection with development of a good plane with normal parenchyma in less invasive tumors was possible in 48% of the cases which is quite low.

The location of the lesion also determines the extent of resection .Retrochaismatic lesions is more difficult to remove completely .Prefixed chaism also makes things more difficult. We feel that opening of lamina terminalis helps in more ways than one and should be part of the standard operating procedure in large tumours [11].

Whatever be the achieved extent of resection, it has to be classified and documented with the help of a postoperative CT scan which must be done in all cases. CT scan tend to overestimate the extent or resection [12]. Surgeon is the best judge and intra operative findings must be meticulously jotted down after very surgery .

Perioperative Mortality

We lost three patients in the postoperative period. It gives us a mortality rate of around 10% which is much higher than the accepted norm of 2.5% [7, 13, 14].

We accept that the sample size is rather small to actually debate on the high mortality rate in our series. Late presentation with preoperative malnourished status in two of the three children that we lost could be one of the factors. However, we do believe that the size of the lesion have no correlation with surgical mortality [8].

Tumour Recurrences

Recurrence is one factor which always weighs heavily on the mind of a neurosurgeon especially in paediatric cases. These are slow growing benign tumors and require a minimum of 5 and preferably ten year follow up for an accurate recurrence study [14,15]. What we are seeing in our series is probably regrowth of residual lesions than true recurrence [16,17].

Extent of resection seems to be the most important factor in predicting regrowth. We advised radiotherapy in all our cases of partial removal and have observed that harmful effects of radiotherapy are over stated [18,19]. It is a viable and efficient alternative to resurgery which has very high rates of complications [20,21]. Modern neuroimaging helps us in diagnosing recurrences at very early stages in postoperative cases and radiotherapy has a critical role in these cases [22].

Quality of Life Assessment

It is in our opinion the most important parameter that has to be evaluated satisfactorily. In reality, it is also the most difficult to judge. Duff et al [6] proposed eight criteria for revaluation: 1) still alive at the follow-up examination; 2) no major motor deficit related to treatment or tumor progression; 3) functional vision; 4) a Katz grade of A; 5) a Karnofsky Performance Scale score of at least 80; 6) school status of no more than 1 year behind the expected grade for children and young adults; 7) employability for adults of working age; and 8) absence of debilitating psychological or emotional problems.

In our series, satisfactory results according to above criteria were obtained in 17/27 cases that we operated. Visual status of the patient at presentation seems to be one of the most important factors in achieving the desire quality of life later on as surgery does not help once optic atrophy is already present.

Anterior pituitary failure is another complication common after surgery [21].

Preservation of stalk does not guarantee endocrinological stability in the post operative period but we do believe that preservation of stalk should be attempted in every case. Partial hypoptuitarism allows a much better functional outcome than pan hypopituitarism.

Conclusions

We conclude that radical resection of craniopharyngiomas with preservation of the pituitary stalk should be the goal of surgery. However; one should not be overly aggressive in approach for these tumors as radiotherapy also has a proven role in their management. Further follow up is required in our series to judge the role of radiotherapy in the recurrence patients in our study group.

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