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Sandeep Sangale,
Sharad Pandey,
Ashok Pathak, Monica
Narayanswamy

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Evaluation of visual and oculomotor outcome in patients with posterior fossa tumours

Sandeep Sangale, Sharad Pandey, Ashok Pathak, Monica Narayanswamy

A.B.V.I.M.S and Dr Ram Manohar Lohia Hospital, New Delhi, INDIA

ABSTRACT

Objective: To evaluate and compare visual and oculomotor outcomes in patients with posterior fossa tumours.

Methods: Prospective Observational Study including 42 patients with posterior fossa tumours who underwent either CSF diversion or definitive surgery at RML Hospital, Delhi, between July 2021 and January 2023. Preoperatively and postoperatively, a neurological and visual assessment was done. The visual evaluation consisted of Visual acuity, Fundus, Diplopia charting, Perimetry, and forced duction test. Preoperative and Postoperative visual outcome was compared and statistically analysed. Visual outcomes are also compared according to the final histological diagnosis.

Results: 30 cases underwent CSF diversion, and 12 cases received definitive surgery. The visual acuity was divided into three groups i.e. Good, Fair, and Poor. The visual acuity was good and fair in 2.5% and 92.8% of the cases pre-operatively, and postoperatively, it was good in 26.2% and fair in 69% cases, respectively. The difference was found to be statistically significant ($P < 0.01$). Papilledema decreased from grade 2 to grade 1 in 50% of cases postoperatively, while it was absent in 87.5% of the cases at three months of follow-up. Diplopia was absent in all of the cases at follow-up at 3 months.

Conclusions: If treated timely, posterior fossa tumours are typically associated with a positive prognosis. The majority of patients with posterior fossa tumours had good visual outcomes with the use of CSF diversion, proper microsurgical procedures, and follow-up.

Keywords
visual,
oculomotor,
posterior fossa,
tumours

Corresponding author:
Sharad Pandey

A.B.V.I.M.S and Dr Ram Manohar Lohia Hospital,
New Delhi, India

drsharad23@yahoo.com

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INTRODUCTION

Posterior fossa tumors commonly present with ocular signs and symptoms that can compromise the ocular afferent and efferent system. Direct compression of the cerebrospinal fluid pathways can lead to obstructive hydrocephalus and papilledema.

Papilledema is a compressive optic neuropathy due to elevated intracranial pressure. The degree of optic nerve head edema is associated with ocular afferent system dysfunction, which left undetected may progress to ganglion cell axon loss, optic nerve atrophy, and permanent vision loss, even despite treatment of hydrocephalus. Posterior fossa tumors may also disrupt the ocular efferent system, which includes the extraocular cranial nerves and gaze-

holding centers. Abducens nerve palsy can occur due to injury of the abducens nerve within the subarachnoid space by hydrocephalus and downward displacement of the brain stem, as well as by direct mass effect on the pons. Abducens palsies present clinically with inward eye deviation and horizontal diplopia. Trochlear nerve resulting from mid-brain damage leads to vertical or oblique diplopia, with resultant hypertropia of the involved eye and anomalous head tilt. In third cranial nerve palsy (oculomotor nerve palsy), the involved eye usually deviates "down and out" and there may be partial or complete ptosis. Tumors that affect the midbrain, cerebellum, and brainstem may disrupt visual fixation and vestibular and gaze stabilization mechanisms which can lead to acquired nystagmus and complex gaze palsy.

There are three types of optic neuropathies caused by brain tumors: compressive, papilledematous, and infiltrative. All can lead to visual loss. The visual loss in compressive lesions is attributed to the interruption of the blood supply to the optic nerve with secondary ischemic optic atrophy. While papilledema may cause blockage of the axonal flow. Brain tumors in children can be associated with visual loss which may depend on the location, type of tumor, and duration of the disease. Although papilledema typically resolves following treatment, either by surgical resection of the tumor and/or neurosurgical shunting for hydrocephalus, severe or prolonged swelling of the optic nerve can result in optic atrophy and permanent vision loss. Tumors involving the cerebellum and brainstem may also lead to ocular motor dysfunction, resulting in nystagmus and strabismus.

Posterior fossa tumors may require ophthalmological assessment and intervention. The prevalence of unrecognized visual field deficits in children with brain tumors can be surprisingly high. Serial neuro-ophthalmologic evaluation of brain tumors is often required to diagnose a visual field deficit since patient or caregiver reporting may be limited. Posterior fossa tumors are generally associated with a favorable outcome if they are managed appropriately. The total removal of Posterior fossa tumors without neurological deficit is possible with appropriate microsurgical techniques.

Method

This study was a prospective study of "Evaluation of Visual and Oculomotor outcome in patients with posterior fossa tumors" performed at the Neurosurgery Department, ABVIMS& Dr RML Hospital from 1st July 2021 to 1st January 2023. Preoperatively and postoperatively visual assessment was done. Demographic profile of patients, History, Neurological examination, and neuroimaging were recorded. Preoperative and postoperative ophthalmology examinations consist of Visual acuity (uncorrected visual acuity and Best Corrected Visual Acuity), Fundus examination, diplopia charting, perimetry, and forced duction test.

Follow-up after 1 month, 2 months, and then after 3 months was done including neurological and ophthalmological examinations were done. Appropriate descriptive statistics were used.

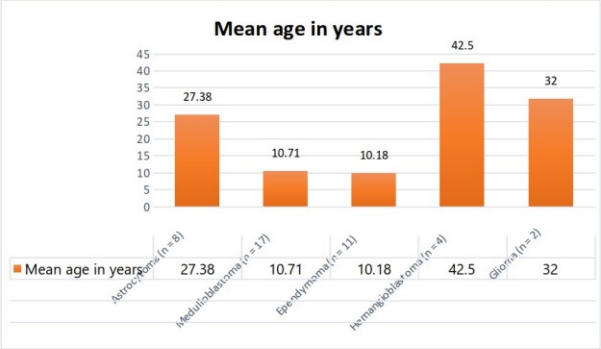

Figure 1. Distribution of cases by age and diagnosis.

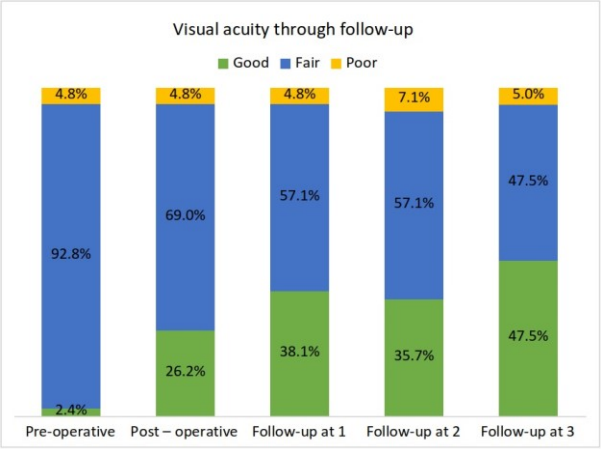

Figure 2. Distribution of cases by visual acuity in pre-operative, post-operative, and follow-up at one month, two months and three months.

Results

We have admitted 68 patients with posterior fossa tumors. 26 were excluded who did not meet the

study criteria (Recurrent posterior fossa tumor, pre-existing visual and oculomotor dysfunction). 42 patients remained who were included in the study. The majority 66.7% of the cases were children followed by 31 % of adults and 2.3% of old aged respectively. 57.1% of the cases were males and 42.9% were females. Headache was the most common symptom seen in 85.7%, followed by Ataxia in 21 (50%) and multiple vomiting in 18 (42.8%).

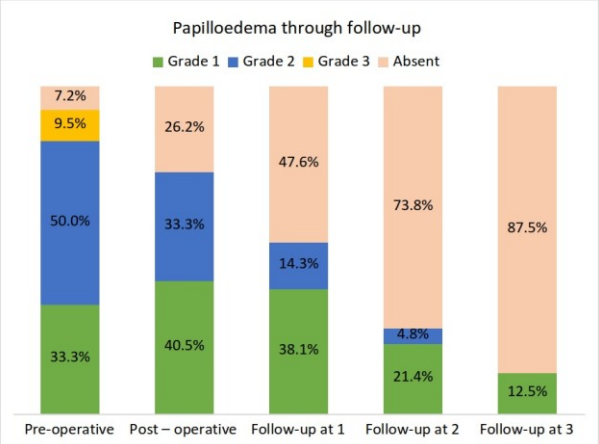

Figure 3. Distribution of cases by Papilloedema in pre-operative, post-operative, and follow-up at one month, two months and three months.

With histologic confirmation, 40.5% of the cases were diagnosed as Medulloblastoma followed by 26.2%, 19%, 9.5%, and 4.8% of ependymoma, astrocytoma, hemangioblastoma, and glioma respectively. Regarding grade, 100% of the Astrocytomas were grade 1 tumors, and 100% of the Medulloblastomas were grade 4 tumors. Among ependymoma tumors, 55.5% were grade 2 and 45.5% were grade 3 tumors and Among hemangioblastoma tumors, 50% were grade 1 and 50% were grade 2 tumors. The mean age of the cases with astrocytoma, medulloblastoma, ependymoma, hemangioblastoma, and glioma was 27.38 years, 10.71 years, 10.18 years, 42.50 years, 32 years, and 17.79 years respectively and the difference was found to be statistically significant. The majority of the cases in children were medulloblastoma (53.6%) and ependymoma (50%). In adults, the majority of the cases were Astrocytoma (42.9%) and hemangioblastoma (42.9%), in middle age, the majority of the cases were Astrocytoma (50%) and hemangioblastoma (50%) and in old age the case was glioma (100%).

The visual acuity was divided into three groups i.e. Good, fair, and poor. The visual acuity was good and fair in 2.5% and 92.8% of the cases pre-operatively, in postoperatively it was good in 26.2% and fair in 69% cases respectively. The visual acuity increased from good in 2.4% of cases in the pre-operative period to good in 47.5% of cases at three months of follow-up and the difference was found to be statistically significant. The presence of papilloedema decreased from grade 2 in 50% of cases in the pre-operative period to grade 1 and absent in 12.5% and 87.5% of the cases respectively at three months of follow-up and the difference was found to be statistically significant.

In the present study, among the cases of Astrocytoma, visual acuity was fair in 100% of the cases pre-operatively and good in 85.7% of the cases at follow-up at 3 months. Papilloedema was seen in 87.5% of the cases of Astrocytoma and absent in 100% of the cases at follow-up of 3 months. Diplopia was seen in 12.5% of the cases of Astrocytoma pre-operatively and absent in 100% of the cases at follow-up at 3 months.

Among the cases of Medulloblastoma, pre-operative visual acuity was fair and good in 88.2% and 5.9% of the cases respectively, and good and fair in 64.7% and 29.4% of the cases respectively at follow-up at 3 months. Papilloedema was seen in 84.1% of the cases of Medulloblastoma and absent in 88.2% of the cases at a follow-up of 3 months. Diplopia was seen in 17.6% of the cases of Medulloblastoma pre-operatively and absent in 100% of the cases at follow-up at 3 months.

Among the cases of Ependymoma, pre-operative visual acuity was fair and poor in 90.9% and 9.1% of the cases respectively, and good and fair in 18.2% and 72.7% of the cases respectively at follow-up at 3 months. Papilloedema was seen in 100% of the cases of Ependymoma and absent in 81.8% of the cases at a follow-up of 3 months. Diplopia was absent in 100% of the cases of Ependymoma both pre-operatively and at follow-up at 3 months.

Discussion

The posterior fossa tumors are considered critical as they cause brainstem compression, herniation, and death. However posterior fossa tumors are generally associated with a favorable outcome if they are managed appropriately. The total removal of Posterior fossa tumors without neurological deficit is

possible with appropriate microsurgical techniques. 1

Age

In our study, the majority of cases were children followed by adults and old age. It was similar to a study by Rehman et al., 2009 where they concluded Posterior fossa tumors are more common in children than in adults. Between 54% and 70% of all childhood brain tumors originate in the posterior fossa. About 15-20% of brain tumors in adults occur in the posterior fossa. 2

Sex

In our study, 57.1% of the cases were males and 42.9% were females. Similarly in a study by Peeler et al., 2017, Meenakshisundaram et al., 2018, Out of 108 cases, the gender distribution was 60 (56%) male and 48 (44%) female patients with male preponderance.3,4

Symptoms

The cerebellum is involved in many complex aspects of human behavior and function, and when it is disrupted or insulted, this can lead to significant sequelae in children with posterior fossa tumors. A constellation of impairing and distressing symptoms, including mutism, ataxia/hypotonia, and emotional lability, develops in approximately 25% of children after the surgical resection of posterior fossa tumors. These symptoms may impede treatment and frequently require intervention in order for children to be able to participate in their care. The eventual recovery of speech occurs for most, but with slowly improving dysarthria over many months. Behavioral changes and emotional lability also occur.5 In our study, headache was the most common symptom seen in 85.7%, followed by Ataxia in 21 (50%) and multiple vomiting in 18 (42.8%).

Histopathological diagnosis

In the present study, 40.5% of the cases were diagnosed as Medulloblastoma followed by 26.2%, 19%, 9.5%, and 4.8% of ependymoma, astrocytoma, hemangioblastoma, and glioma respectively. Similarly in a review by Gilliard 2022, medulloblastoma: most common (30-40%) and posterior fossa astrocytoma -pilocytic astrocytoma: second most common (25-35%) Whereas in another study by Meenakshisundaram et al., 2018, where the final diagnosis was made by histopathological examination which is as follows, schwannoma 36 (33%) cases, meningioma 24 (22%) cases, medulloblastoma 14 (13%) cases, pilocytic astrocytoma 17 (16%) cases, high-grade astrocytoma 7 (7%) cases, Metastasis 8 (8%) cases, round blue cell tumor 2 (2%) cases6.

The majority of the cases in children were medulloblastoma (53.6%) and ependymoma (50%), in adults, the majority of the cases were Astrocytoma (42.9%) and hemangioblastoma (42.9%), middle age and in old age the case was glioma (100%). In the present study, the mean age of the cases with astrocytoma, medulloblastoma, ependymoma, hemangioblastoma, and glioma was 27.38 years, 10.71 years, 10.18 years, 42.50 years, 32 years and 17.79 years respectively and the difference was found to be statistically significant.

Brain metastases in adults are the most common malignancies at this localization. Ependymomas and medulloblastomas occur mostly in children. Other tumors that occur in the posterior fossa are meningiomas, schwannomas, hemangioblastomas, brain stem gliomas, and epidermoid tumors. Due to the fact that the various tumors of the posterior fossa have different treatment approaches and prognoses, an accurate and specific diagnosis is mandatory.7

Distribution of cases by pre-operative and post-operative visual acuity and papilloedema

In the present study, the visual acuity was good and fair in 2.5% and 92.8% of the cases pre-operatively and was fair and good in 69% and 26.2% of the cases respectively in the postoperative period and the difference was found to be statistically significant. The visual acuity increased from good in 2.4% of cases in the pre-operative period to good in 47.5% of cases at three months of follow-up and the difference was found to be statistically significant. This implies the importance of timely surgical intervention and follow-up.

Similarly, Kaif, 2021, A shifting trend towards normalization of visual acuity was seen post-surgery in all age groups. Overall improvement was seen in the majority of cases having pupil normal size sluggish reactive (NSSR) after surgery. Papilledema improved in the majority of patients in all age groups and patients having hydrocephalus. The visual field defect was not improved postoperatively in the

majority. Visual parameters like visual acuity, pupil size, and reactivity to light; color vision, and night vision were improved significantly after surgery whereas the cut field of vision did not improve.

The presence of papilloedema decreased from grade 2 in 50% of cases in the pre-operative period to grade 1 and absent in 12.5% and 87.5% of the cases respectively at three months of follow-up and the difference was found to be statistically significant. Similarly, in a study by Kaif, 2021, all fundoscopic findings like papilledema, retinal venous dilation, and retinal splinter hemorrhage disappeared in a significant number of patients post-operatively.8

Distribution of cases of Astrocytoma in pre-operative, post-operative, and follow-up

It is favorable for low-grade tumors, with survival times approaching 7 to 8 years after surgery. In anaplastic astrocytoma, therapy focuses on improvement in symptoms. Radiotherapy of partially resected tumors increases postoperative survival rates. Survival rates after post-surgery radiation are nearly double that of only surgical intervention .9

In the present study, among the cases of astrocytoma, visual acuity was fair in 100% of the cases pre-operatively and good in 85.7% of the cases at follow-up at 3 months. Pre-operatively, papilloedema was seen in 87.5% of the cases of Astrocytoma and absent in 100% of the cases at follow-up of 3 months. Diplopia was seen in 12.5% of the cases of Astrocytoma pre-operatively and absent in 100% of the cases at follow-up at 3 months.

Fisher et al. 2008 found out that low-grade astrocytomas, particularly pilocytic astrocytoma have excellent long-term overall survival. While tumor location and resection extent affect the outcome, pathologic diagnosis when carefully interpreted significantly influences long-term survival.10

Distribution of cases of Medulloblastoma in pre-operative, post-operative, and follow-up

Medulloblastoma is the most common malignant brain tumor in children constituting nearly 20% of all pediatric brain tumors. It is categorized as an embryonal neuroepithelial tumor of the cerebellum. Multimodality treatment regimens have substantially improved survival in this disease; however, the tumor is incurable in about a third of patients with medulloblastoma.

This is a high-grade tumor that has a propensity to spread via the cerebrospinal fluid. Within the first few years of diagnosis, mortality approximates 15%; however, cure rates can reach as high as 60% with current therapeutic modalities. Surgical resection followed by radiation and chemotherapy is the mainstay of therapy, with five-year survival rates of between 50% to 90%. This wide range is multifactorial, owing in part to age at diagnosis, the presence of metastases at diagnosis, and a histologic variant of medulloblastoma. Age, hemispheric location of the tumor, the extent of resection, and adjuvant therapy status were the important clinical prognostic factors for survival. 14 Complete resection should be performed if possible as several studies have correlated outcome with the extent of resection and amount of residual tumor.11

In the present study, among the cases of Medulloblastoma, pre-operative visual acuity was fair and good in 88.2% and 5.9% of the cases respectively, and good and fair in 64.7% and 29.4% of the cases respectively at follow-up at 3 months. Preoperatively, papilloedema was seen in 84.1% of the cases of Medulloblastoma and absent in 88.2% of the cases at follow-up of 3 months.15 Diplopia was seen in 17.6% of the cases of Medulloblastoma pre-operatively and absent in 100% of the cases at follow-up at 3 months.

Distribution of cases of Ependymoma by visual acuity in pre-operative, post-operative, and follow-up at one month, two months and three months

In a study by Mork and Loke, One hundred and one patients with histologically confirmed ependymomas were studied over a 22-year period. Choroid plexus papilloma and sub-ependymoma were not included. About half of the tumors were intracranial, with the majority of these infratentorial. The intraspinal tumors were equally divided between the intramedullary and the "cauda" group. The majority of the intracranial tumors occurred in children, while almost all the intraspinal tumors were in adults.12

In the present study, among the cases of Ependymoma, pre-operative visual acuity was fair and poor in 90.9% and 9.1% of the cases respectively, and good and fair in 18.2% and 72.7% of the cases

respectively at follow-up at 3 months. Papilloedema was seen in 100% of the cases of Ependymoma preoperatively and absent in 81.8% of the cases at a follow-up of 3 months. Diplopia was absent in 100% of the cases of Ependymoma both pre-operatively and at follow-up at 3 months. Simarly in a study by Lee et al. 1998, all patients were followed up postoperatively for an average of 50.6 months (range 6 months to 6 years). All patients are surviving to date. Surgical resection of these tumors led to significant alleviation of pre-operative symptoms. There has been no radiographic evidence of tumor recurrence or growth in any patient to date.13

The majority of the patients with posterior fossa tumors underwent CSF diversion (ventriculoperitoneal shunt, External Ventricular Diversion) and some underwent definitive surgery (Midline sub-occipital approach, Retro-mastoid approach)

Conclusion

If treated properly, posterior fossa tumors are typically associated with a positive prognosis. The majority of the cases in children were medulloblastoma and ependymoma, in young adults cases were Astrocytoma and hemangioblastoma, and in old age the case was glioma. The visual acuity increased from good in 2.4% of cases in the pre-operative period to good in 47.5% of cases at three months of follow-up. Pre-operatively, papilloedema was seen in all of the cases of Ependymoma and absent in 81.8% of the cases at follow-up of 3 months and diplopia was absent in 100% at follow-up at 3 months. With the use of the proper microsurgical procedures and proper follow-up, posterior fossa tumors can be completely removed without causing neurological damage.

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