E-ISSN: 2616-3470
P-ISSN: 2616-3462

www.surgeryscience.com
2022; 6(1): 209-213
Received: 26-10-2021
Accepted: 06-12-2021

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Clinicopathological study of pineal gland lesions

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DOI: https://doi.org/10.33545/surgery.2022.v6.i1d.852

Abstract
Background: Pineal parenchymal tumors are the second most common form of pineal tumor.
Aims: To study the incidence, age distribution, gender distribution of pineal region tumors and post-operative outcome.
Materials and Methods: The present study is a prospective study comprising 20 cases of Pineal Region tumors, studied over a two and half year period. Data related to all cases were retrieved from archives. Available clinical data including, patient age, sex, clinical features, Imaging and surgical findings, tumor histology, outcome were also received from the Department of Neurosurgery Medical record Department. All pineal region tumors received at Neurosurgery department with adequate pre-operative; intra-operative and postoperative information were included in the study. Pineal region tumors comprising the Pineal parenchymal cell tumors, Germ cell tumors, Glial cell tumors, and other miscellaneous tumors and cysts.
Results: Most common age group for the pineal region tumors is second and third decade and the Male to Female ratio is 1:1. Malignant nature of pineal region tumors is 17 cases incidence being 85%. Predominant grade of the pineal region tumors are grade I tumors commonest presenting symptoms being headache and vomiting. Commonest clinical sign is papilledema of features suggestive raised intracranial pressure. Most of patients presented at delayed stage with large tumor and morbid state with hydrocephalus, majority of the patients underwent shunt surgery before definitive surgery, some patients underwent external ventricular drainage simultaneously after surgery. No surgical mortality, one patient detoriated after third post operative day did not recovered and died. Inpatient had developed meningitis, sepsis didn't recover and died. Mortality 2 cases. Medium follow up period is one year. Three cases followed recurrent symptoms advised radiotherapy. Two cases came with shunt malfunction.
Conclusion: Many of the approaches are interchangeable and they are depending on anatomical location and surgeon s choice and experience.

Keywords: Pineocytoma, Infra tentorial supra cerebellar approach, shunt malfunction

Introduction

Pineal tumors account for 0.5% of all central nervous system (CNS) tumors in adults, 1% in young adults (aged 20-34 years), and 2.7% in children (aged 1-12 years) [1]. Because these cancers are so rare, it has always been difficult to collect a large number of cases to study and compare. Pineal tumors can be classified as germ cell tumors (GCTs), pineal parenchymal tumors (PPTs), gliomas, atypical rhabdoid / tetartoid tumors, other tumors such as the most recently described entity, papillary tumors of the pineal region. Germ cell tumors are the most common subtype of pineal gland tumor. In the literature, the incidence of Germ cell tumors varies from 50% to 75% of tumors in the pineal region. These tumors arise from pluripotential germ cells, which normally do not inhabit the pineal gland. Theoretically, these germ cells mistakenly migrate to the pineal gland during embryogenesis. Per the most recent World Health Organization (WHO) CNS tumor classification system, Germ cell tumors are further classified into germinomas, which is the most common subtype, and a group of non germinomatous germ cell tumors Germ cell tumors can grow as pure forms (i.e., comprising only one cell type) or as mixed forms. Pineal parenchymal tumors are the second most common form of pineal tumor. They represent 14% to 27% of tumors in the pineal gland [2].

In the WHO classification of CNS tumors, pineal parenchymal tumors are further classified as Pineocytoma, pineal parenchymal tumors of intermediate differentiation, including mixed pineocytoma-pineoblastoma tumors and pineoblastoma. In the literature, the incidence of subtypes varies greatly, i.e., the incidence of pineocytoma ranges from 14% to 60%; that of pineoblastoma is 45%; and that of pineal parenchymal tumors with intermediate differentiation is 10%. Other CNS tumors can arise from the supporting stroma of the pineal gland.

These tumors include gliomas, fibrillary astrocytoma, anaplastic astrocytoma, glioblastoma, and pilocytic astrocytoma. we studied incidence of all pineal gland tumour of the brain and postoperative outcome [3].

Materials And Method

The present study is a prospective study comprising 20 cases of Pineal Region tumors, studied over a two and half year period from August 2017 to December 2019. Data related to all cases were retrieved from archives. Available clinical data including, patient age, sex, clinical features, Imaging and surgical findings, tumor histology, outcome were also received from the Department of Neurosurgery, Department of Pathology and the Medical record Department, Osmania General Hospital, Hyderabad.

Inclusion Criteria: All pineal region tumors received at Neurosurgery department from August 2017 to December 2019, with adequate pre-operative; intra-operative and postoperative information were included in the study. Pineal region tumors comprising the Pineal parenchymal cell tumors, Germ cell tumors, Glial cell tumors, and other miscellaneous tumors and cysts.

The clinical records and material for histologic review were available in all cases.

Results

The present study is a prospective study comprising 20 cases of pineal gland region studied over a two and half year period from August 2017 to December 2019. A total of 644 intra cranial tumors ware operated during this period in which pineal gland tumors comprise about 3.1%.

Table 1: Demographic details in study

Age-Years	Incidence	Percentage
0-10	3	15%
11-20	4	20%
21-30	6	30%
31-40	2	10%
41-50	2	10%
51-60	1	5%
61-70	2	10%
Gender		
Male	10	50%
Female	10	50%

This study revealed that the most common age group for the pineal region tumors is second and third decade. 10 cases out of 20 cases are found in between 11-30 years incidence being 50%. The sex distribution of the pineal region tumors in this study showed of equal incidence male 10 cases (50%), female 10 cases (50%) and the Male to Female ratio is 1:1.

Table 2: Incidence of type of tumor in study

Type of Tumour	Incidence	Percentage
Pineal Parenchymal Tumour	10	50%
Germ Cell Tumour	4	20%
Astrocytoma	2	10%
Meningioma	2	10%
Others Lipoma	2	10%
Sub Type		
Pineocytoma	7	70%
Pineal parenchymal Tumor of Intermediate differentiation	2	20%
Pineoblastoma	1	10%

This study shows the most common pineal region tumors were pineal parenchymal tumors (PPTS) constituting about 10 cases out of 20 cases with overall incidence of 50%, Germ cell tumors about 4 cases out of 20 cases with overall incidence 20%, astrocytoma about 2 cases with overall incidence 10%. Most common pineal parenchymal tumors (PPTS) being pineocytoma 7 out of 10 cases of pineal parenchymal tumors (PPTS), incidence being 70%. Remaining cases are pineal parenchymal tumor of intermediate differentiation 2 cases (20%), pineob1astoma 1 (10%).

Table 3: Nature and grading of tumor

Nature	Incidence	Percentage
Benign	3	15%
Malignant	17	85%
Grading of tumor		
Grade- I	11	55%
Grade – II	2	10%
Grade – III	2	10%
Grade -IV	1	5%

In our study malignant nature of pineal region tumors is 17 cases incidence being 85%, Benign cases are 3 in number 2 cases are meningioma, one case is lipoma. Remaining cases are pineal parenchymal tumors 10 cases, germ cell tumors 4 cases, astrocytoma 2 cases are malignant nature.

This study revealed the predominant grade of the pineal region tumors are grade I tumors constituting 11 cases out of 20 cases, incidence being 55% and the least predominant grade of tumors reported in this study are grade 4 tumors 1 cases out of 20 cases, incidence is 5%, grading is considered according to WHO classification 2007 system.

Table 4: Symptoms and signs of patients in study

Symptom	Incidence	Percentage
Headache	17	85%
Vomiting	13	65%
Blurring of Vision	9	45%
Giddiness	6	30%
Seizures	2	10%
Altered Sensorium	2	10%
Double Vision	2	10%
Weakness	2	10%
Memory Disturbance	1	5%
Sign		
Papilloedema	12	60%
Ataxia	5	25%
Upward Gaze Paralysis	4	20%
Weakness	3	15%
Altered Sensorium	2	10%
Memory Disturbance	1	5%

In our study commonest presenting symptom being headache. Out of 20 cases 17 cases presented with headache, incidence being 85%. Second most common symptom being vomiting with incidence of 65% (13 cases).

Commonest clinical sign is papilloedema, 12 out of 20 cases, incidence being 60%, other signs ataxia (25%), upward gaze paresis (20%), weakness (15%), altered sensorium (10%), memory disturbance (5%).

Hydrocephalus management Patients are presented features of raised intra cranial features of headache, vomiting, papilloedema two patients presented with altered sensorium. Patients are

improved symptomatically improved after shunt procedure.
Shunt requirement in 12 cases of 20 cases,
Preoperatively shunt placement in10 cases,
Post operatively shunt placement in 1 case.

Surgical Procedure

Definitive surgical procedure done in 18 cases remaining two cases only shunt procedure done in our institution prone position preferred, sitting position one case done.

Table 5: Surgical Approaches in present study

Surgical approach	No of cases
Infra tentorial supra cerebellar approach	15
Trans cortical trans ventricular approach	1
Trans callosal inter hemispheric approach	1
Endoscopic biopsy	1

Surgical procedures preferred are Infra tentorial Supra cerebellar Approach, one ease done through trans cortical trans ventricular approach because of tumor extended into third ventricle. One case done through trans callosal inter hemispheric approach.

Table 6: Extent of removal in present study

xtent of Removal	Number of Cases	Percentage
Total	2	11%
Sub Total Removal	12	66%
Partial Removal	4	22%
Biopsy	2	11%

Complete removal of tumor done in two cases. Subtotal removal in 12 (66%) tumor that adhered to internal cerebral vein, great cerebral vein Galen at velum interpostium are spared. Partial removal of tumor done in four ease, in one patient having dural AVM noticed intraoperatively partial resection of tumor done, in two cases pineal lesion (astrocytoma) densely attached to surrounding structures so decompression of tumor done, in one case tumor which extended to third ventricle partial removal tumor done.

Table 7: Post-operative complications in present study

Post-operative complications	Number of cases	Percentage
New ocular movement disturbance	2	1.1%
Ataxia	1	0.5%
weakness	1	0.5%
Meningitis		0.5%

New ocular movement disturbance-2 cases, one case developed upward gaze paresis, one case developed diplopia subsequently improved. Patient developed swaying to right side afterward improved. Patient developed right lower limb paresis after one month improved. Patient developed fever, neck rigidity treated with antibiotics.
No surgical mortality, one patient detoriated after third post operative day did not recovered and died. Inpatient had developed meningitis, sepsis didn't recover and died. Mortality 2 cases.
Patients are followed at 1 month, 3rd month, 6th month followed by every 6 month. Medium follow up period is one year. Among 18 cases 11 cases are regularly followed up. Three cases followed recurrent symptoms advised radiotherapy. Two cases came with shunt malfunction. Shunt revision done. One patient had expired due to cardio respiratory problem. Six patients are lost followup.

Discussion

The present study is a prospective study comprising 20 cases of pineal gland region studied over a two and half year period from August 2017 to December 2019. A total of 644 intra cranial tumors were operated during this period in which pineal region tumors comprise about 3.1%. Vivek R. Deshmukh, M.D. Kris A. Smith, MD [4] *et al.* Pineal region tumors are rare and account for 0.4 to 1% of intracranial tumors in Western countries and for 2.2 to 8% of intracranial tumors in north- eastern Asian countries [5-9] Ivan Radovanovicl, Kemal Dizdarevic *et al.* pineal region tumors are rare and account for 0.4 to 1.0% of intracranial tumors in adults and 3-8% in children [10]. Maysa Al-Hussaini, Iyad Sultan *et al.* Pineal tumors account for 0.5% of all central nervous system (CNS) tumors in adults, 1% in young adults (aged 20-34 years), and 2.7% in children (aged 1-12 years) [11].
The sex distribution of the pineal region tumors in this study showed of equal incidence male 10 cases (50%), female 10 cases (50%) and the Male to Female ratio is 1:1. Maysa Al-Hussaini, Iyad Sultan 2 *et al.* among 633 Pineal tumors. The cohort consisted of 477 (75%) males and 156 (25%) females [11]. This study revealed that the most common age group for the pineal region tumors is second and third decade, 10 cases out of 20 cases (50%) in this age group. Maysa Al-Hussaini, Iyad Sultan *et al.* The median age for the cohort was 17 years (range, 0-83 years); 56% of the patients were 18 years or younger at the time of diagnosis [11].
This study shows the most common pineal region tumors were pineal parenchymal tumors (PPTS) constituting about 10 cases out of 20 cases with overall incidence of 50%, Germ cell tumors about 4 cases out of 20cases with overall incidence 20%, astrocytoma about 2 cases with overall incidence 10%. Others 2 cases (20%), one case is papillary tumor of pineal region, other is pineal region lipoma. Jefrey C. Allen, Jeffrey Bruce *et al.* Summary of Pathology in 154 Patients in 154 patients. Undergoing Surgery for Pineal Region at the New York Neurological Institute surgical pathology cases seen at the New York Neurologic Institute Germ cell 57 (37%) Pineal cell 35 (23%) Glial cell 43 (28%) [12].
A Cytopathologic Study of 20 Specimens of pineal region tumors by Anil V. Parwani, M.D., Ph.D, l Blaire L. Baisden, M.D. 2 Yener S. Erozan. M.D. Peter C. Burger, M.D. *et al.* shows pineal parenchymal tumors of 9 cases (45%of pineoblastoma (five specimens), Pineocytoma (four specimens), astrocytoma (three specimens) (15%), germ cell tumor (three specimens) (15%), meningioma (one specimen) (5%), epidermoid cyst (EC; three specimens) (15%), and pineal cyst (one specimen) (5%) [13].

Table 8: Comparison of incidence of tumor with our study

Incidence of type of tumor.	Present study	Jefrey C.Allen, Jeffrey Bruce *et al.* [12]	Anil V. Parwani, M.D., Burger, M.D.*et al.* [13]
Pineal parenchymal tumors (PPTS)	55%	23%	45%
Germ cell tumor	20%	37%	15%
Astrocytoma	10%	28%	15%
Others	15%	22%	25%

Most common pineal parenchymal tumors (PPTS) being pineocytoma 7 out of 10 cases of pineal parenchymal tumors (PPTS), incidence being 63%. Konovalov AN *et al.* [14] and Regis J *et al.* [15] pineocytomas represent 14- 60%. Pineocytomas occur throughout life, but most frequently affect adults (mean age: 38 years). There is no sex predilection.

In our study commonest presentating symptom being headache. Out of 20 cases 17 cases presented with headache, incidence being 85%. second most common symptom being vomiting with incidence of 65% (13 cases).

commonest clinical sign is papilledema, 12 out of 20 cases, incidence being 60%.

Most common initial symtoms is headache, which is associated with obstructive hydrocephalus secondary to compression of. the aqueduct of sylvius. Further progression of hydrocephalus can lead to nausea, vomiting obtundation, cognitive impairment, papilledema and ataxia [16].

Patients are presented features of raised intra cranial features of headache, vomiting, papilledema two patients presented with altered sensorium. Patients are improved symptomatically improved after ventriculi peritoneal shunt procedure.

Shunt requirement in 12 cases of 20 cases, Preoperatively shunt placement in 10 cases, Post operatively shunt placement in 1 case. Of these five patients required ventriculoperitoneal shunting and four were treated by third ventriculostomy. In another patient, a transient external ventricular drainage was applied.

Total removal -2 (11%), subtotal removal -12(66%), partial removal 4(22%), Biopsy-2(11%) Complete removal of tumor done in 2 cases. Subtotal removal in 12 (66%) tumor that adhered to internal cerebral vein, great cerebral vein Galen at velum interposition are spared. Partial removal of tumor done in 4 case in one patient having dural AVM noticed intraoperatively partial resection of tumor done in 2 cases pineal lesion densely attached to surrounding structures so decompression of tumor done, in one case tumor which extended to third ventricle partial removal tumor done.

Kobayashi T, Lunsford LD *et al.* shows total removal 54, near total removal 19, partial 10 cases [17].

New ocular movement disturbance -2 cases. One case developed upward gaze paresis, one case devolved diplopia subsequently improved. Patient developed swaying to right side afterward improved. Patient devolved right lower limb paresis after one month improved. Patient developed fever, neck rigidity treated with antibiotics.

No surgical mortality. One patients detoriated after third post-operative day and died. One patient had developed meningitis, sepsis didn't recovered and died. Mortality in 2 cases. Bruce and stein *et al.* shows morbidity rates ranging from 0% to 12% and mortality rates ranging from 0% to 8% [18]. Bruce and Ogden *et al.* shows the surgical major morbidity rate associated with pineal region tumor 3-6.8% and the permanent minor morbidity rate was 3-28% [19].

Conclusion

Pineal gland tumors accounts for 3.1% of all intracranial space occupying lesions peaks third and fourth decades combined of 50%. Equal male and female distribution. This study shows the most common pineal gland tumor are pineal parenchymal tumor followed by germ cell tumors. Among pineal parenchymal tumors pineocytomas being most common.

Commonest presenting symptoms being headache and vomiting. Commonest clinical sign is papilledema of features suggestive raised intracranial pressure. Most of patients presented at delayed stage with large tumor and morbid state with hydrocephalus, majority of the patients underwent shunt surgery before definitive surgery, some patients underwent external ventricular drainage simultaneously after surgery.

In most of the cases, many of the approaches are interchangeable and they are depending on anatomical location and surgeon s choice and experience. In majority of cases, morbidity and mortality are avoided with improvements in surgical techniques (like endoscopy) along with improved neuro anaesthesia.

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