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Original Research Article

A Study of Pineal Region Tumors in the Northeast India

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ABSTRACT

Background: Management of Pineal region tumors is a formidable challenge to neurosurgeons. Infratentorial supracerebellar and occipital transtentorial microsurgical approaches are accepted as the main standard accesses to the pineal region. However, the treatment options for the different pineal region tumors vary according to their histological nature. The present study aims to evaluate the pineal region tumors, and document the clinical presentation, histopathological findings and the various treatment modalities of pineal tumor in North East region of India.
Materials and methods: A retrospective analysis of 2126 brain tumor cases admitted in neurosurgery ward of Gauhati Medical College and Hospital from January 2010 to June 2017 was conducted. Clinical presentation, CT scan and MRI findings were recorded. Tumor markers serum AFP and serum βHCG were evaluated.
Results: Of 2126 cases of brain tumor, 0.70% cases were pineal region tumors. 80% were male and the 20% were female patients. 46.67% germinoma, 33.33% pinealocytoma and 6.67% each of glioma, tuberculoma and benign cyst cases.
Conclusion: The treatment and prognosis differ between particular tumor categories. Noninvasive multimodality approach of radiotherapy and chemotherapy can be effective treatment preventing microsurgical resection to a small subset of benign lesions.

Keywords: Pineal tumors, benign lesions, microsurgery, treatment

INTRODUCTION

Pineal tumors are very rare and 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)] A variety of pathologies varying from benign pineal cysts to malignant tumors like pinealoblastomas and germinomas occur, in the region of the posterior third ventricle. Pineal tumors can be classified as germ cell tumors, pineal parenchymal tumors, gliomas, atypical rhabdoid/teratoid tumors, or other tumors papillary tumors of the pineal region. [(2)] Germ cell tumors (GCTs) being the most common type of tumors, incidence of which varies from 50% to 75% of tumors in the pineal region. [(3–5)] These tumors arise from pluripotential germ cells, which normally do not inhabit the pineal gland. As per the most recent World Health Organization (WHO) CNS tumor classification system, [(6)] GCTs are further classified into germinomas, which is the most common subtype, and a group of nongerminomatous germ cell tumors (NGGCTs). Pineal parenchymal tumors (PPTs) are the second most common form of pineal tumor representing 14% to 27% of tumors in the pineal gland. In the WHO classification of CNS tumors, PPTs are further classified as pineocytoma, PPT of intermediate differentiation, including mixed pineocytoma pineoblastoma tumors

and pineoblastoma. Out of all these subtypes pineocytoma is the commonest though their incidence varies greatly .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. [6,7]

Patients with primary tumors arising in the pineal region tend to have a much shorter prodrome (i.e., weeks to several months). These patients usually present with signs and symptoms of raised intracranial pressure, such as headache, diplopia, and lethargy due to aqueductal obstruction. Tectal compression can cause Parinaud's syndrome (vertical gaze paresis, impaired pupillary light reflex, and convergence nystagmus)

The complexity in accessing this anatomical location makes the management of these lesions a considerable challenge even for the skilled neurosurgeon. Some of the lesions, e.g., germinomas, are extremely radiosensitive making radiotherapy as the primary management option. [8] On the other hand, the benign lesions require surgical excision, as radiation therapy is not an effective alternative. [9] The role of surgery in the management of pineal region tumors is influenced by the histological diagnosis and tumor growth characteristics. Most of the posterior third ventricular tumors obstruct the aqueduct and are associated with hydrocephalus requiring a CSF diversion procedure. Conventionally, ventriculoperitoneal shunts have been performed for the accompanying hydrocephalus although in recent years endoscopic third ventriculostomy has emerged as a favored option for CSF diversion. The second aim of surgery is to establish a tissue diagnosis in cases where tumor markers are not elevated. Tissue samples can be acquired by stereotactic or endoscopic techniques. However, because of the increased risk of hemorrhage in these procedures, open biopsy is also preferred by some which also provides the opportunity of cytoredution which may be therapeutically beneficial. [10] For those patients who are appropriate candidates for tumor resection, the options for surgical approach are influenced by the location and extent of the tumor, and the preferences and experience of the surgeon. Supracerebellar infratentorial, suboccipital transtentorial, and interhemispheric approaches may each be appropriate, depending upon whether the tumor extends mainly above or below the vein of Galen and the degree to which the lesion grows lateral to the midline and anteriorly into the third ventricle

MATERIALS AND METHODS

The present study aims at evaluating the pineal region tumors presenting in Gauhati Medical College and Hospital as this is the only institute in this part of the country to encounter such cases. In the present study, a retrospective analysis was done of all the brain tumor cases admitted in neurosurgery ward, Gauhati Medical College and Hospital from January 2010 to June 2017. The cases diagnosed with pineal region tumor were studied. The variables that were taken into account during the study were the sex, age group of the study population, clinical presentation, radiological features, management and pathology. The incidence of pineal region tumors among all the brain tumor cases was also obtained. The name, age, sex were obtained from the hospital data and clinical presentation were recorded at the time of presentation and also at the time of discharge. CT scan and MRI were done in all the patients pre-operatively and also post-operative CT was done to rule out any post operative bleed or intra-tumoral bleed. Tumor markers viz. serum AFP and serum βHCG were also tested in the patients and taken into account in this study. All the patients were undertaken for emergency VP shunt as all the patients had raised ICP features with obstructive hydrocephalus. The CSF specimen collected after VP shunting procedures were sent for physical chemical and cytological examination..The cases were undertaken for the definitive

surgery for excision and biopsy. The main approach used for the surgery was supracerebeller infratentorial approach in sitting position. Midline suboccipital approach was also used. The histopathology samples attained by the procedures were examined by two senior pathologists and tallied. Postoperative radiotherapy was used wherever necessary and follow-up was done in all the cases.

RESULTS AND OBSERVATIONS

A total of 2126 cases of brain tumor presented to our institute in which 15 cases were diagnosed to have pineal region tumor in the above mentioned time period. This implies that among all brain tumor cases, 0.70% cases were pineal region tumors. Out of this 15 cases, 12 (80%) were male and the rest 3 (20%) were female patients. The age group varied from 2 years to 35 years with a mean age of 13.13 years.

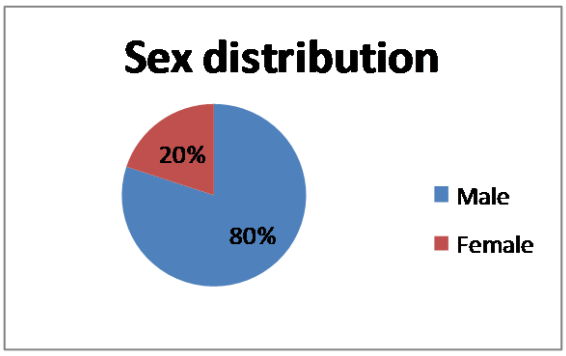

Chart 1: Sex distribution

All the patients presented with features of raised ICP. 5 patients showed features of Perinaud's Syndrome and 2 patients had altered sensorium.

Table 1: Clinical features

Features	Number of cases	percentage
Headache	15	100%
Vomiting	10	66.67%
Visual disturbances	6	40%
Diplopia	4	26.67%
Quadriparesis	3	20%
Perinaud's syndrome	5	33.33%
Altered sensorium	2	13.33%

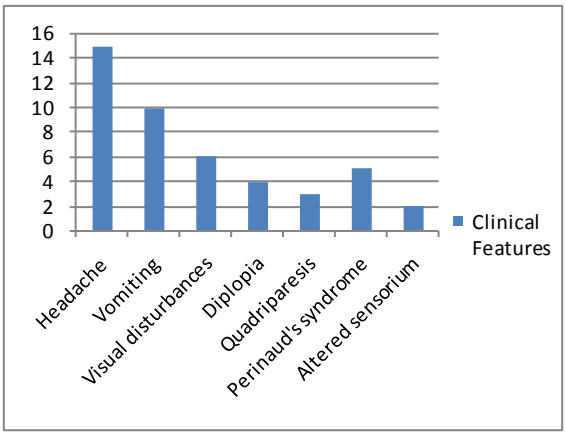

Chart 2: Clinical features

In plain CT scan, all the patients had obstructive hydrocephalus. On contrast 8 patients had isodense enhancing lesions in posterior third ventricle, 5 patients had hyperdense enhancing lesions in pineal region, and 1 patient had isodense non-enhancing lesions whereas the other patient had hypo-dense non-enhancing lesion in the posterior third ventricle. Calcification in the lesions was seen in 3 patients.

MRI was also done in all the patients and there were hypointense to hyper intense lesions in the pineal region as seen in MRI. Germinoma patients have hydrocephalus in all the cases with isointense lesions in T1 weighted MRI and slightly hyper to hyperintense in T2 weighted MRI. Contrast enhancement was seen in all the cases of germinoma. One case of germinoma had single cystic area in the lesion and one had multiple cystic areas intralesionally. The pinealocytoma cases had isointense lesions in T1 images and iso to hypointense lesions in T2 images. The tuberculoma and benign cyst show hypointense lesions in both T1 and T2 images. The glioma patient had iso intensity lesion in T1 image and hyper intense image in T2 where as contrast enhancement was also present.

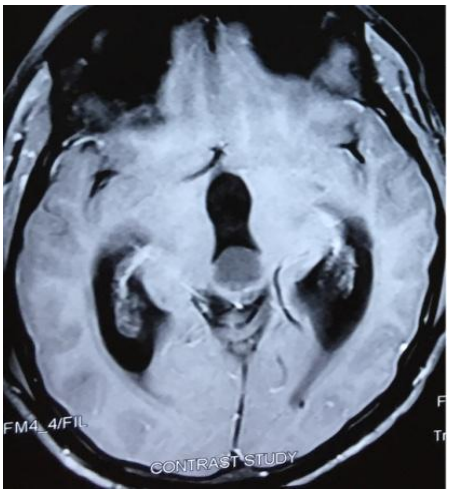

Image 1: Pineal cyst with hypo intense and non contrast enhancing lesion

Tumor markers that were tested were serum AFP and serum βHCG. βHCG was found to be weak to moderately positive in patients that were later diagnosed to have germinoma in HPE studies but AFP was negative in all the patients.

All the patients underwent preoperative VP shunting for obstructive hydrocephalus.CSF cytology for all the patients were negative for malignant cells. All the 15 patients underwent excision of the tumor by open surgery. The main approach was infratentorial supracerebeller approach whereas midline suboccipital approach was also used in few.

A histological diagnosis could be achieved in all the patients in whom biopsy was attempted (Table1). There were seven germinomas, five pinealocytomas,one astrocytomas (grades II). One had histological features suggestive of tuberculoma and one patient was diagnosed to be benign pineal cyst.

Table 2 : MRI features depending on tumor histology

Tumor type	T1 image	T2 image
Germinoma	Isointense, cystic occasionally	Slightly hyper to hyperintense, occasionally cystic
Pinealocytoma	Isointense lesion	Iso to hypointense lesion
Tuberculoma	Hypointense lesion	Hypointense lesion
Pineal cyst	Hypointense lesion	Hypointense lesion
Glioma	Iso intense	hyperintense

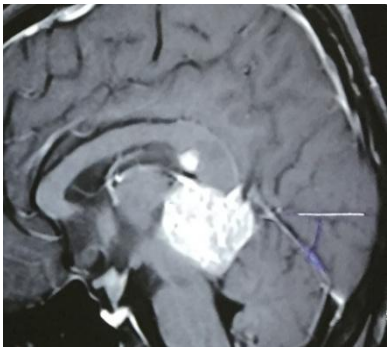

Image 2: Contrast enhancing germinoma in posterior third ventricle

Tumor histology	Number of cases	percentage
Germinoma	7	46.67%
Pinealocytoma	5	33.33%
Glioma	1	6.67%
Tuberculoma	1	6.67%
Benign cyst	1	6.67%

The patients with a histological diagnosis suggestive of germinoma received cranial irradiation and the patients with pinealocytoma were considered for craniospinal irradiation and chemotherapy. The youngest patient in this group, who was 2 years old, received only chemotherapy. Antitubercular therapy consisting of isoniazid, rifampicin for 18 months and pyrazinamide for the initial 3 months was administered to the patients with tuberculomas.

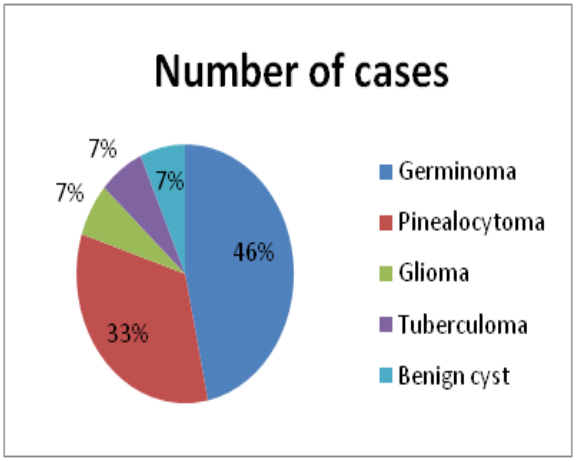

Chart 3: Histological subtypes

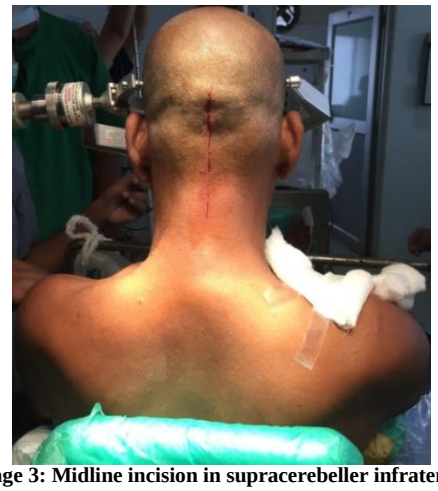

Image 3: Midline incision in supracerebeller infratentorial approach in sitting position

DISCUSSION

Pineal region tumors comprise 0.4–1% of all intracranial tumors in adults and 5–10% in pediatric population. These pose a considerable challenge even to the skilled neurosurgeon in view of its proximity to the vital neural and vascular structures. This issue is further complicated by the diversity of lesions peculiar to the pineal region and the associated hydrocephalus. The objectives of managing these tumors are to establish CSF diversion and effectively treat the primary tumor. [7,11] An accurate histological diagnosis is essential for the later. [12,13]

In our present study we found the incidence of pineal region tumors to be 0.70% which is at par with the 0.5 -1% mentioned in the literature. Out of which 80% were male and rest 20% female patients and average age was 13.13 years. Abay et al found 77.77 % cases to be male and their mean age was 13.7 years. [13] Cho et al also found 0.68% cases of brain tumor to be pineal tumors and the mean age was 13 years with a male to female ratio of 3.36:1. [14] Cho et al also stated that the most frequent presenting symptom was due to increased intracranial pressure (90%), followed by Parinaud syndrome or diplopia (50%). [14] In our present study 100% cases had signs of raised ICP and 33.33% cases had Perinaud's syndrome. It was also found that 13.33% cases presented with altered sensorium and 20% cases had quadriparesis. Visual disturbances and diplopia were found in 40% and 26.67% cases respectively. Presence of hydrocephalus in CT and/or MRI with features of raised ICP makes a CSF diversion procedure an urgent necessity in these patients and this was achieved by emergency VP shunting in our present study. In the present era MR imaging provides excellent results in locating the lesions and even predicts the histopathological variant of the tumor. This is very important in case of pineal region tumors because of the greatest difficulty in treating these tumors. MR has provided a marked improvement in the localization and characterization of tumors as a result of facile multiplaner imaging and superior tissue contrast and resolution. Edwards et al stated that they observed that pinealoblastomas tend to be large and irregular in shape, differentiating them from most of other tumors. [15]

Experiences from the SEER database reveals 50-75% cases to be germ cell tumor and 14-27% percent cases to be pineal parenchymal tumors. [16] Cho et al found the 48 cases consisted of 33 cases of germ cell tumor (69%, GCT); 9 of pineal parenchymal tumors (18.8%); 3 of anaplastic astrocytoma (6.3%); 1 of astrocytoma; 1 of glioblastoma; and 1 of ependymoma. [14] We found 46.67% cases to be germinoma, 33.33% to be pinealocytoma and 6.67% cases to be glioma, tuberculoma and benign pineal cysts.

CONCLUSION

Pineal region tumors, which for long were considered a formidable challenge to neurosurgeons, are now being increasingly managed by CSF diversion and definitive procedure to make a histopathological diagnosis. With increasing knowledge about the biology of tumors harboring in pineal region, most of the malignant tumors in the future could possibly be managed with multimodality approach using radiotherapy and chemotherapy, restricting microsurgical resection to a small subset of benign lesions.

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