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Pineal Region Ependymoma: A Case Report

A case of papillary ependymoma, which is a rare pathological entity in the pineal region, is presented here. Only few cases have been reported in the literature. In this case report we will discuss the clinical scenario and operative management of pineal region ependymoma

Key words: ependymoma, pineal region, infratentorial supra-cerebellar approach

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Pineal region is defined as the area of the brain bounded dorsally by the selenium of the corpus callous and the tela-choroidea, ventrally by the quadrigeminal plate and mid-brain tectum, rostrally by the posterior aspect of the 3rd ventricle, and caudally by the cerebral vermis.[9] Pineal region tumors account for less than 1% of primary brain tumors.[7] These tumors are encountered with an incidence of 0.6%–0.9% in North America and Europe, 3.0%–3.2% in Japan and Southeast Asia, and 3% in China.14 Ependymomas represent <1% of the pineal region tumors reported.

Tumors in pineal region may become symptomatic by three possible mechanisms: (1) increased intracranial pressure from hydrocephalus, (2) direct cerebellar or brain stem compression, or (3) endocrine dysfunction. Direct brainstem compression may lead to disturbances of extraocular movements, classically known as Parinaud's syndrome. a-fetoprotein and ß-hcg are markers of germ cell malignancy, which can also be useful to monitor therapeutic response and as a sensitive early sign of tumor recurrence. S antigen and melatonin have been more useful for immunohistochemical application in tissue diagnosis. The standard diagnostic workup includes magnetic resonance imaging (MRI), with and without contrast.

In the early 1900s, Krause developed the concept of an infra-tentorial supra-cerebellar approach. Stein reintroduced this approach in 1971. The posterior transcallosal approach was advocated by Dandy in 1921. The frontal approach first described by Dandy in 1921. The transcortical-transventricular approach was first

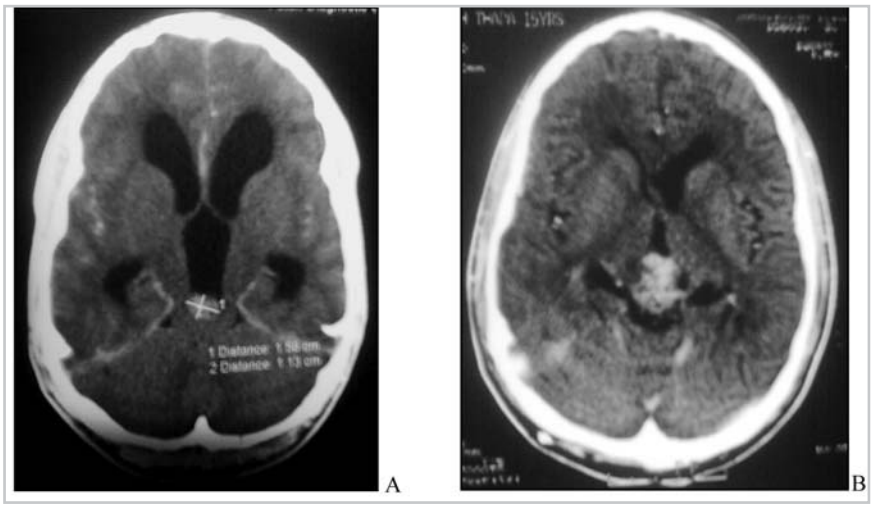

Figure1: Plain Computed tomography (CT) scans. A) An enhancing mass in pineal region and secondary dilatation of the ventricular system. B) Post V-P shunt, well defined mixed density predominately hyperdense lesion in pineal region located at the posterior aspect of the 3rd ventricle.

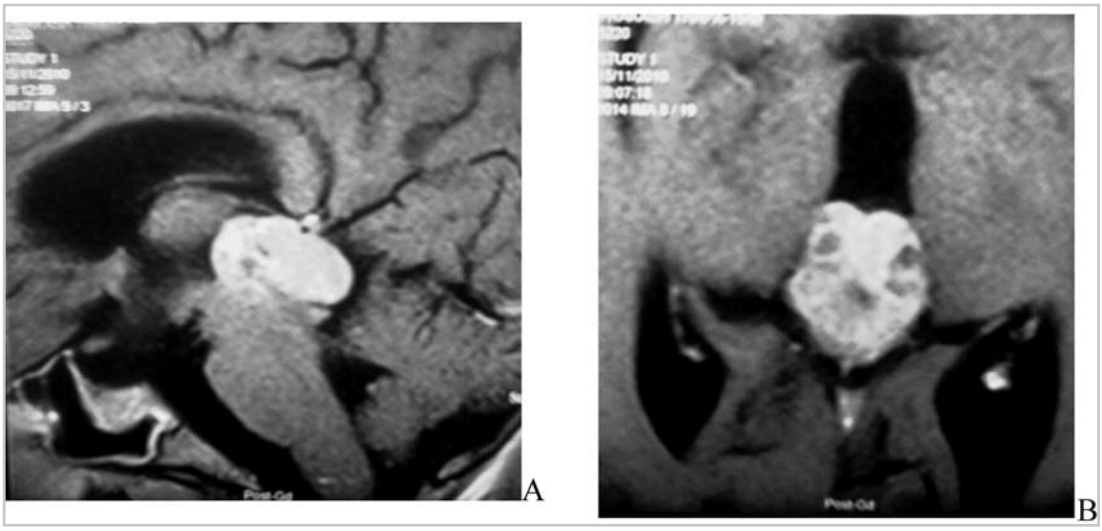

Figure2: MRI scans. Mass at the projection site of pineal gland extending to posterior 3rd ventricle and quadrigeminal plate cistern. Close-up (Post Gadolinium) views A) Sagittal view, B) Axial view.

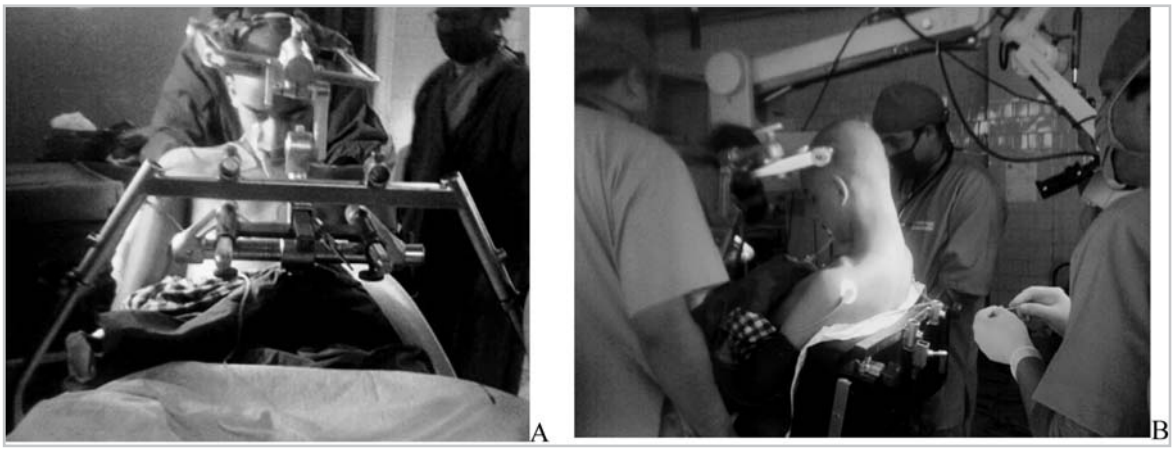

Figure3: Patient in Operation Theatre, Pre-op positioning. A) Front view, B) Left side view.

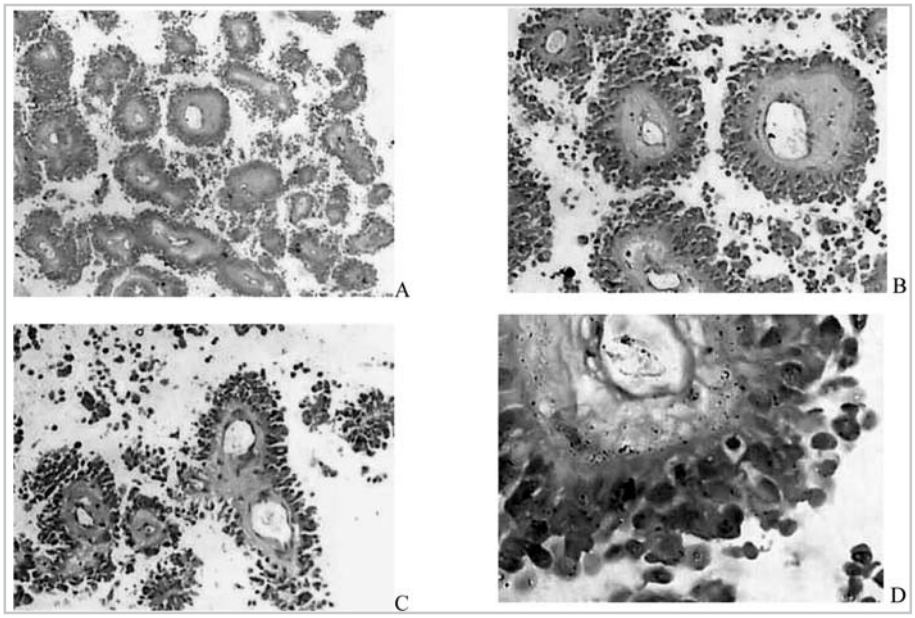

Figure4: Histopathological microscopic photographs of the tumor. A)4X, B) 10X, C) 40X, D) 100X

described by Von Wagenen in 1931. The occipital transtentorial approach was initially described by Poppen in 1966 and then modified by Jamieson.

Germ cell tumors, ependymomas and pineal cell tumors metastasize easily through the CSF ("drop metastases"). Thus, clinical and radiological follow-up of the entire cerebrospinal axis is mandatory. We present here a patient with pineal region ependymoma.

Case report

Clinical presentation

A 15-year-old boy was admitted via emergency room in semi-comatous state (GCS 7/15). Headaches, nausea and vomiting proceeded before he lapsed into unconsciousness. Both of his pupils were sluggishly reacting 3mm in diameter with papilledema. Non-enhanced brain computed tomography (CT) scans showed small well defined slightly hyperdense enhancing mass measuring approx 15 x 12 mm in pineal region with dilatation of bilateral lateral and 3rd ventricle **(Figure1)**. Emergency vetriculo-peritoneal shunt was done. After the procedure his general condition improved with GCS 15/15. However, the parinaud sign was positive and there was gait ataxia.

Post VP shunt MRI of Brain **(Figure 2)** showed ~ 3.2 x 2.2 x 2.2 cm size mass at the projection site of pineal gland which was heterogeneously enhanced after gadolinium administration. High signal intensity area was noted in posterior aspect of the mass in T1 weighted images. The mass displays heterogeneous iso to low signal in T1 and mixed iso and high signal intensity in T2 weighted images. Post contrast there was marked heterogeneous enhancement in the mass. There was compression of the tectal plate with obstruction of Foramen of Munro. Cerebrospinal fluid cytology and serologic examination of a-fetoprotein (AFP) and human chorionic gonadotrophins (ß-hcg) were all negative.

Operative procedure and pathologic findings Infratentorial supra-cerebellar approach was performed with patient in sitting position **(Figure 3)**. There was solid suckable grayish white capsulated mass in the posterior part of the third ventricle originating from tectal plate of mid brain. A total microscopic excision of the tumor was achieved.

On the fifth post operative day the patient had an episode of absence seizure for which sodium valproate was started. Following which he made an excellent recovery. The patient was discharged without any neurological deficit. The MRI spine during discharge confirmed absence of drop metastasis.

Histopathologically **(Figure 4)**, there is increased cellularity with papillary and diffuse pattern of tumor cells. There are perivascular pseudo-rosettes and ependymal rosettes as well. The features were compatible with ependymoma (WHO grade II).

Post operative MRI **(Figure 5)**, showed no residual tumor iin the pineal region. Post contrast study showed minimal heterogeneously enhancing area in postero-medial aspect of the thalamus. There was no abnormal parenchymal and meningeal enhancement, except for minimal enhancement at tumor excision site.

Discussion

Although pineal region tumors were first described in 1717, the earliest attempts at pineal region surgery did not

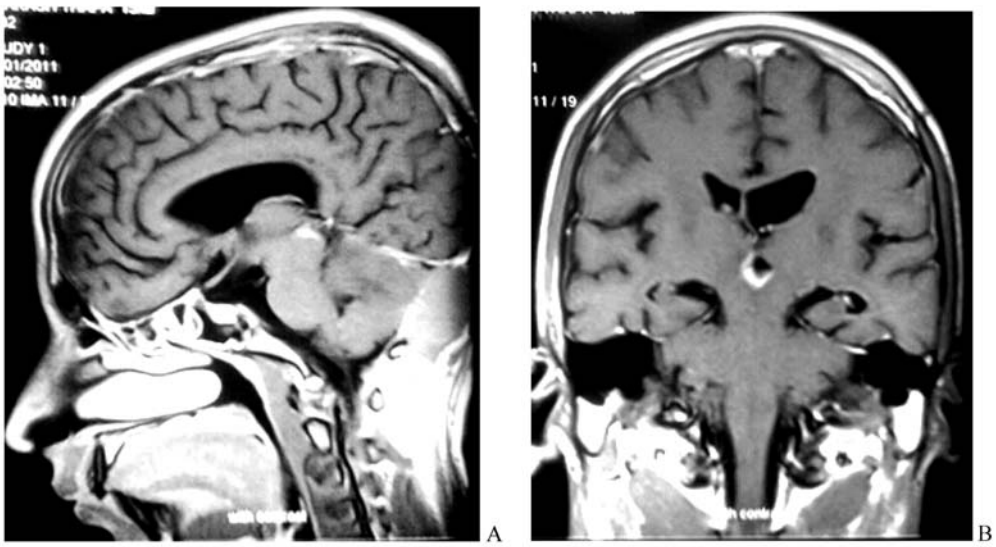

Figure5: Magnetic resonance imaging (MRI) scans 11 days after surgery . A) Sagittal view, B) Coronal view

occur until 200 yrs later. In 1926 Krause described modest success without operative mortality in three patients using infratentorial approach.[4] In 1971 Stein ushered in the microsurgical era for pineal region surgery with his successful modification of Krause's infratentorial approach.[11]

Ependymomas are infrequently seen tumors that have been defined as neoplasms arising from ependymal cells lining the ventricles and the central canal of the spinal cord. The four most prevalent locations are supra- and infratentorial, apical, and conus-cauda-filum. The occurrence of supratentorial ependymoma in the pineal region was well recognized as early as in 1954.9 In 1998, Cho et al. reported pineal region tumors treated at Seoul National University Hospital. Among the pineal region tumors, there was one pineal ependymoma. Surgery is the treatment of choice for the rarest pineal tumor, ependymoma. Despite the advances in high-resolution MRI, tumor histology cannot be reliably predicted based on radiographic features alone. Computed tomography scans can complement MRI by providing details of calcification, blood-brain barrier breakdown, and degree of vascularity.[2]

The classic dilemma facing the surgeon whose patient has an ependymoma has been described concisely: attempt a complete resection and risk serious neurological complications, or leave tumor behind to spare morbidity and await the inevitable recurrence.[12] The literature reported approximately 90% mortality before 1943 and 30% to 79% mortality and 65% morbidity after that.5 Nonsurgical methods, including chemotherapy and radiotherapy, have been predominantly used to treat pineal lesions, although these strategies are mostly recommended for patients who are elderly, in poor medical condition, or affected by recurrent disease.[8]

The choice of approach depends on the location, extent of the lesion, and surgeon preference. More recently, stereotactic and endoscopic technology has added a new dimension to management of this tumor.10 Burr hole endoscopic techniques may be performed in a non-emergency situation and after correction of the hydrocephalus by endoscopic third ventriculostomy, when a direct microsurgical approach is indicated.

In infratentorial supracerebellar approach (Krause approach), patient is operated in a sitting position with the neck well flexed such that the tentorium is parallel to the floor. A suboccipital craniectomy is done and the dural opening is based on the transverse sinus. Gravity aids in the retraction of the cerebellum. The corridor between the tentorium and the superior surface of the cerebellum is opened up by dropping the culmen after arachnoid dissection. The tentorium may be incised to access the supratentorial region where necessary.[6]

Generally, the infratentorial supracerebellar approach in preferred for several reasons:

1. The approach is to the center of the tumor, which begins at the midline and grows eccentrically.
2. The approach is ventral to the velum interpositum and the deep venous system to which the tumor is often adherent. This minimizes the risk of damage to the vascular drainage of this critical region.
3. The exposure in the sitting position is comparable with that of other routes.
4. No normal tissue is violated on route to the tumor.

The disadvantages include inability to access tumors with supratentorial extension, vision being restricted to the midline, and sacrifice of the draining veins of the cerebellum.3 Utilization of the sitting position also carries the risk of an air embolism.

The main surgical complications in the pineal surgery are (1) intra- and/or postoperative hemorrhage, (2) damage to the midbrain, thalamus, and hypothalamus, (3) visual field defect. After removal of the tumor the Valsalva maneuver should be applied to confirm complete hemostasis. On tumor removal every effort should be made to preserve the medial posterior choroidal artery, which is always attached to the lateral side of the tumor. Obstruction of the artery may cause ischemia in the medial thalamus. Damage to the major veins in the pineal region is well documented by many authors.

Conclusion

The supracerebellar infratentorial approach is the ideal approach to the pineal region tumor.

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