Resection of pineal region epidermoid in semisitting position: A case report
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Abstract
Objective: A case of pineal region mass with triventricular hydrocephalus (HCP) managed by endoscopy and microsurgery is presented. Clinical presentation: A 25 yrs old women presented with headache, visual blurring and occasional vomiting. Her CT scan and MRI revealed a pineal region mass which was seemed to be a glioma. Intervention: Initially triventricular HCP was managed by endoscopic third ventriculostomy (ETV). Her headache, vomiting was relieved following ETV. Definitive surgery was done after few weeks of ETV in the form of Midline Suboccipital Craniectomy in semisitting position. Total tumour was removed and histopathology revealed an epidermoid. Conclusion: Although epidermoid is not uncommon in the pineal region, the successful total removal by proper selection of approach is crucial for avoidance of complication and recurrence free survival.

Introduction
Pineal region tumour makes up 0.4 to 1.0 percent of brain tumours in adult and 3-8 percent of brain tumours in children. While germinoma and astrocytoma accounts for 53 to 78 percent of pineal region tumours, a wide variety of histologically distant tumour types can be found in this region. Pineal region comprises pineal gland itself, tectal plate, quadrigeminal cistern and pulvinar of thalamus.

Lesions in pineal region compromise the superior colliculus, either through direct compression or through tumor invasion, results in a syndrome of vertical gaze palsy that can be associated with pupillary or oculomotor nerve paresis. This Parinaud syndrome has become virtually pathognomonic for lesions involving the pineal region.

Further compression of the Sylvian aqueduct and periaqueductal gray region causes hydrocephalus, mydriasis, convergence spasm, pupillary inequality and nystagmus.

Impairment of downgaze becomes more pronounced with tumors involving the ventral midbrain.

Torkildsen in 1948 argued for abandoning aggressive surgical resection and was in favour of cerebrospinal fluid (CSF) diversion followed by empiric radiotherapy. This became the standard of treatment until 1980s.

New Surgical approaches to the pineal region has been developed.

1. Supratentorial approaches include-
   A. Parietal-interhemispheric approach described by Dandy (1936).
   B. Occipital - transtentorial approach originally described by Horrax, later modified by Poppen.
2. Infratentorial-
   Supracerebellar infratentorial approach of Krause and modified by stein.
3. Combined (supratentorial & infratentorial approach).

Figure 1: Shows pineal region
Case Report

The algorithm of CSF diversion, radiation and observation sometimes is successful; however, patients with benign lesion may exposed to unnecessary and ineffective radiation.

Here, we report a case of pineal region epidermoid which was successfully treated with microsurgery in semisitting position.

Case report
A 25 yrs old women presented with a history of chronic headache, visual blurring and occasional vomiting for 3 months. There were no history of diplopia, ataxia and 6th nerve palsy. Neurological examination revealed normal except 2nd nerve, which were found moderate papilloedema bilateraly.

Initially CT scan was done and revealed a hypodense mass in the pineal region with triventriculomegaly.

MRI scan was done to see the details of the lesion, which was in favour of epidermoid of pineal region. It appeared hypointence in T1WI and hyperintense in T2WI. Triventricular hydrocephalus and periventricular edema were also noted.

Tumour markers (alpha feto proten and HCG) were examined from blood, which revealed normal.

The patient underwent endoscopic 3rd ventriculostomy as a part of hydrocephalus management. Following 3rd ventriculostomy her headache, visual blurring reduced.
Case Report

Subsequently she was managed by supracerebellar infratentorial approach in semisitting position to remove the pineal epidermoid. It was done successfully with microscopic total removal. Peroperative the tumour appeared pearly white in colour, soft, capsulated, suckable tumour which laid below the vein of Galen and Rosenthal vein.

After surgery patient developed pseudomeningocele in the suboccipital area which was managed by continuous lumbar drain for 5 days. Following lumbar drain her pseudomeningocele was disappeared. Before discharge she had no headache, no visual blurring, no vomiting, no ataxia and diplopia.

Discussion
The preoperative radiological diagnosis of this lesion includes arachnoid cyst, germinoma, teratoma. However tumour markers were not in favour of germinoma, seminoma or teratoma. MRI scan well evaluated the lesion and concluded that it was an epidermoid. Now the controversy is whether to shunt the patient or endoscopic third ventriculostomy as because it was associated with moderate to severe hydrocephalus due to blockage of aqueduct of Sylvius. Particularly for benign and radiosensitive tumour and if MRI suggest that obstruction of CSF pathway can be corrected by surgically- endoscopic third ventriculostomy and definitive surgery should be performed. In case of malignant tumours such as germinoma-Ventriculo-Peritoneal shunt has been implicated as a conduit for tumour cell allowing peritoneal seeding in to abdominal cavity.

Planning for the surgical resection of pineal region tumour is based on tumour location as visualized on midsagittal and coronal MR image. If the majority of the tumour located above the tentorium occipital transtentorial approach is pursued. If majority of the tumour located below the tentorium a supracerebellar infratentorial approach is favoured. Semisitting position is favoured for supracerebellar infratentorial approach.

Conclusion
We have presented the pineal region epidermoid with hydrocephalus. To the best of our knowledge it was first case which managed by endoscopic 3rd ventriculostomy and supracerebellar infratentorial approach in semisitting position in our country. We reviewed the literature on the reported cases of pineal region epidermoid with hydrocephalus.

We recommend endoscopic 3rd ventriculostomy instead of VP shunt for the management of HCP and microscopic total removal of epidermoid in proper position either by supracerebellar infratentorial or occipital transtentorial approach for decompression of Sylvian aqueduct as well as avoidance of delayed recurrence of tumour in this critical zone.

References


