PAPILLARY TUMOUR OF PINEAL REGION- A RARE CASE REPORT

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PRESENTATION OF CASE

A 40-year-old female patient presented with history of diminished vision & bowel bladder incontinence for ten days. She also had a history of altered sensorium for two days. There was no history of trauma, seizure, loss of consciousness or fever. No past history of neurological tumours or tumours elsewhere.

Patient was conscious and incoherent with a Glasgow coma scale score of 14 (E4V4M6). Bilateral pupils were reactive to light, and her vitals were stable. Higher mental functions were not elicited. Motor system, reflexes, sensory system, cerebellum, meningeal signs examinations were normal. She had a wide based gait. Examination of the optic nerve showed bilateral finger counting. All other cranial nerves were normal. Fundus examination showed bilateral pallor of optic disc.

Routine blood investigations done were within normal limits and viral markers were negative. CT Brain (Plain) showed dilatation of supra tentorial ventricular system with normal 4th ventricle suggestive of obstructive hydrocephalus and an evidence of an ill-defined hyperdense lesion (2.5 x 2.1 cms) in the pineal gland region.

MRI Brain (plain) showed heterogenous mass lesion (2.7 x 2.7 x 3.7 cms) in pineal region compressing the posterior part of 3rd ventricle with dilated supra tentorial ventricular system (Obstructive hydrocephalus) suggestive of malignant pineal tumour.

A diagnosis of Pineal gland tumour was made.

DIFFERENTIAL DIAGNOSIS

1. Pineal Parenchymal Tumours
2. Papillary Tumours of Choroid Plexus
3. Ependymoma
4. Metastatic Papillary Tumours

Figure 1. MRI Brain (Contrast) Showing Moderate Heterogeneous Enhancement

Figure 2. MR Spectroscopy with Significant Increase in Choline Peak & Decrease in NAA & Creatine Peaks

PATHOLOGICAL DISCUSSION

Squash Cytology

0.5 cc of grey white tissue bits were received in the department of pathology for squash cytology which was carefully smeared and stained with toluidine blue.

The cellular cytosmear showed sheets & individually scattered small round to oval cells with vesicular nuclei & scant to moderate amount of cytoplasm. Characteristic rosettes with neuropil in lumen noticed. There is increased vascularity with collapsed blood vessels and peritheliomatous arrangement of tumour cells in papillary pattern noticed.

Possibility of papillary tumour of pineal region was considered.

Microscopy

H & E stained section from the pineal gland tumour showed tumour tissue arranged in papillary pattern with fibrovascular core. Papillae are lined by tumour epithelial cells with stratification. Individual tumour cells are round to ovoid with moderate amount of eosinophilic cytoplasm, clear cell change.

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Nuclei are ovoid with uniform chromatin and nucleoli in some. Mitotic activity of 1-2/HPF & focal necrotic area seen. Pseudo-rosettes also seen. Suggestive of papillary tumour of pineal region.

Figure 3. Toluidine Blue (10x) Low Power View Showing Tumour Cells Arranged in Papillary Pattern

Figure 4. H & E (10x) Low Power View Showing Papillary Architecture and Pseudorosettes

Figure 5. H & E (40x) High Power View Showing Tumor Cells Arranged in Papillary Pattern with a Fibrovascular Core

Immunohistochemistry

The most distinctive IHC feature is their reactivity for keratins (KL1, AE1/AE3, CAM5.2, and CK18), particularly in papillary structures. GFAP expression is less common than in ependymomas. Absence of staining for membranous KIR7.1, cytoplasmic stanniocalcin-1, cadherin-1, and claudin-2, which are positive mostly in choroid plexus tumours.^[2]

Figure 6. Showing CK 8/18 Expression in the Epithelial-Like Neoplastic Cells. Immunoexpression may be diffuse/ or may Predominate in Perivascular Areas (40X)

Figure 7. Tumor Cells with Negative Expression for GFAP (40x)

DISCUSSION OF MANAGEMENT

The patient underwent right ventriculo-peritoneal shunt after admission followed by sub occipital craniotomy (supra cerebellar infratentorial approach) and excision of the tumour. The post-operative period was uneventful.

At present, treatment involves a multispecialty approach with maximum safe surgical resection and either fractionated radiation therapy or stereotactic radiosurgery.^[6,7] Gamma knife radiosurgery, with or without temozolomide chemotherapy, should be considered as an option for the treatment of residual disease following subtotal resection of PTPR. The role of adjuvant chemotherapy is not well defined and further study is warranted.^[8,9,10]

Local recurrence is common, but because of the rarity of the lesion, the biologic behavior and histologic grading of papillary tumour of the pineal region remain to be defined. Overall survival rate is 73% at 5 years.^[2] With early diagnosis & management the patient's overall survival rate will be better.

Although PTPR is a rare tumour, it is being recognized more commonly because of an increased awareness among neuropathologists who now include it in the differential diagnoses of the tumours of the pineal region. More information is required to determine the prognosis and standardize the treatment protocol of this rare tumour.

Papillary tumour of pineal region comprising 0.4 - 1.0% of intracranial space-occupying lesions in adults is a very rare entity which has been newly described in 4th edition of WHO 2007 classification of brain tumours.^[1,5] It is a neuroepithelial tumour localized in the pineal region and characterized by a combination of papillary and solid areas, with epithelial-like cells.^[2] Pineal region tumours can be of primary or secondary origin. The name papillary tumour of pineal region (PTPR) is a neuropathologic description of the tumour manifested by papillary features, rosettes, and pseudo-rosettes. Other tumours of the pineal region which may exhibit papillary features include choroid plexus papillomas and carcinomas,

papillary ependymoma, and metastatic papillary carcinomas. Pineal parenchymal tumours, meningiomas, and germ cell tumours may rarely display papillary features. The histologic appearance of a PTPR is less papillary than choroid plexus papilloma and more epithelial than ependymoma, without the fibrillary background.

They usually present as large, well-circumscribed masses with an average age of presentation of 35 years. It shows frequent recurrence, occurring in about 58% of cases by 5 years but only occasional cases show spinal dissemination.^[2,4] In microscopy an epithelial-like growth pattern may be seen in which the vessels are covered by a layer of tumoural cells and the papillary areas show large neoplastic cells (Columnar or cuboidal) with a clear cytoplasm. Nuclei, round or infolded is found generally at the basal pole of tumoural cells.^[3] Immunohistochemical findings (i.e. cytokeratin positivity) and ultrastructural demonstration of ependymal, secretory, and neuroendocrine organelles implies that the tumour arises from the remnants of specialized ependymal cells of the subcommissural organ (SCO) along with expression of genes like ZFHX4, SPDEF, RFX3, TTR, CALCA (Normally seen in sub commissural organ). It also shows a claudin expression profile similar to that of sub commissural organ in human fetuses (Claudin 1&3 positivity and claudin 2 negativity). The biological behaviour of papillary tumour of the pineal region is variable and may correspond to WHO grades II or III.^[2]

On imaging it is seen as well-circumscribed heterogeneous masses composed of cystic and solid portions and centred by the posterior commissure or the pineal region. Aqueductal obstruction with hydrocephalus is a frequent associated finding. Intrinsic T1 hyperintensity is seen in cases of calcification, haemorrhage, melanin/fat or secretory material with high protein and glycoprotein content.^[6]

FINAL DIAGNOSIS

Papillary Tumour of Pineal Region

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