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Papillary tumour of pineal region.

Medical Science

KEYWORDS:

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A 50 year lady presented with altered sensorium and weakness of the both lower limbs since 6 months.Neurological examination showed bilateral papilloedema. There was restriction of upward gaze with gait ataxia. MRI brain showed a 3x3 cm well defined, heterogeneously enhancing lesion in the posterior part of the third ventricle causing obstructive hydrocephalus (Fig 1). A ventriculoperitoneal shunt was performed followed by partial excision of the lesion through a supracerebellar-infratentorial approach. The tumour was grayish, soft, suckable and vascular. Histopathology showed a cellular tumour arranged in sheets and papillae, with perivascular arrangement at places, some vessels having a hyalinized cuff (Fig 2). The tumour cells were round to oval with central nucleus, moderate nuclear pleomorphism, granular chromatin and moderate amount of eosinophilic, dense cytoplasm. A rare mitosis was seen but necrosis or endocapillary proliferation was not evident. On immunohistochemistry, pancytokeratin and vimentin were positive with a paranuclear accentuation. Chromogranin was positive in a cytoplasmic and paranuclear dot pattern. These features were consistent with a papillary tumour of the pineal region. Post operatively the patient was given radiotherapy (6140cGy in 25 fractions). Follow up at 1 and 2 years show no residual or recurrent tumour.

Papillary tumour of the pineal region (PTPR) is a rare and recently described entity with a characteristic anatomic location and histopathology.[1] The pineal region tumours constitute 1% of all primary central nervous system neoplasms. The commonest are germ cell tumours with the rest being shared by the pineal parenchymal tumours and neuroepithelial tumours. The papillary tumour of the pineal region belongs to this group of neuroepithelial tumours.

PTPR which has been introduced in the WHO classification of brain tumours in 2007[1], is a tumour unique to this region, speculated to arise from the subcommissural organ.[2,3,4]These were first descrbied by Jouvet et al in 20032 in a series of 6 patients, about 60 cases have been reported in literature since then.[2,4]

PTPR's are common in adult females and have a wide age range, from 5 to 66 years.[3,5]

Radiologically they are well circumscribed, solid masses with or without a cystic component. Histologically they are characterized by a papillary architecture admixed with solid areas as seen in our case. The tumour cells are polygonal to epithelioid with perivascular pseudorosettes and true rosettes or tubules. The papillary areas have fibrovascular cores and are lined by similar cells showing pseudostratification. The higher grade tumours show necrosis. A signet ring appearance may also be seen.[3] Mitosis and the Ki-67 index are usually not very high.[4]

Immunohistochemistry shows a dot like cytoplasmic positivitiy for cytokeratin and vimentin The vimentin positivity is accentuated in tumour cells around blood vessels. Synaptophysin and chromogranin are sometimes expressed. S-100 and NSE show a diffuse positivity[3] whereas GFAP expression has been reported focally in few cases.[5]

PTPR's need to the differentiated from other papillary neoplasms in this region, namely, papillary ependymoma, papillary meningioma, metastatic papillary carcinoma and choroid plexus tumours.

Papillary tumours of the pineal region are considered as WHO grade-II or III, though the exact criteria for differentiating into these two grades are not yet well defined. Features of increased atypia, mitosis and necrosis are reported to be seen in the higher grade tumors.

Consensus criteria for the best line of management are not available owing to the rarity And limited experience with these tumours, though maximal resection with adjuvant radiotherapy[4] has been advocated. Incomplete resection and high mitoses seem to orrelate with decreased survival and recurrence[6].

The prognosis of PTPR is uncertain. These tumors are characterized by frequent local recurrence, and thus the literature currently suggests they may be graded either II or III, but complete criteria for grading are not yetformulated. In a series of 31 cases reviewed by Fèvre-Montange et al, progression was identified in 72% of cases, with five-year estimates of overall and progression-free survival set at 73% and 27%, respectively.7 Incomplete resection and a mitotic index higher than five per 10 high power fields correlated with decreased survival and increased recurrence. Gross total resection was the only clinical factor strongly associated with overall survival and recurrence, but the results were not statistically significant.[6] Other treatments for PTPR have included adjuvant and neoadjuvant temozolomide after complete resection, adjuvant temozolomide/etoposide after complete resection, adjuvant carboplatin/etoposide following resection, and ACNU given after radiation but before surgery[8]. When the treatment was either adjuvant after surgery and/or radiation or concomitant with radiation, the independent effect of the systemic chemotherapy could not be assessed.

Figure Legends:

Figure 1 : Sagittal contrast MRI showing the pineal tumour with obstructive hydrocephalus

Figure 2 : Haematoxylin & Eosin (10 X) stain showing tumour cells arranged in papillary pattern and sheets (2). Immunohistochemistry (40X) showing Cytokeratin positivity in tumour cells (b), Chromogranin positivity with a paranuclear dot positivity (c) and Vimentin positivity (d).

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