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Case Report

Pineal parenchymal tumour of intermediate differentiation: a rare case report

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ABSTRACT

Pineal parenchymal tumours are erratic, accounting for <1% of all primary central nervous system tumours. It was first categorised by the WHO in 2000 as a pineal parenchymal tumour with an intermediate prognosis between pineocytoma and pineoblastoma. We reported a case of 38-year-old gentle man presented with chief complaints of headache and vomiting since 1 day. MRI brain showed a relatively well defined lesion epicentered at posterior aspect of third ventricle, extending and blocking Aqueduct of Sylvius leading to upstream dilatation of both lateral and third ventricles with periventricular ooze was observed. Excision of lesion was performed, histopathological diagnosis of pineal parenchymal tumours of intermediate differentiation (PPTID) was made, which was further confirmed on IHC. PPTID are enormously erratic tumour, and restricted data are available concerning their pathologic features and biologic behaviours causing interruption in making proper diagnosis and deciding an optimal treatment approach.

Keywords: Pineal parenchymal tumour of intermediate differentiation, Pineocytoma, Pineoblastoma

INTRODUCTION

Pineal parenchymal tumours are erratic, accounting for <1% of all primary central nervous system tumours.[1] Origin of pineal region neoplasm can be pineal parenchymal cells or residual stem cells and neighbouring glia. Pineal region tumours originating from pineal parenchymal cells account for about 27% and are designated as pineal parenchymal tumours.

Pineal parenchymal tumours are assorted entities, unveiling substantial morphological distinction.[2]

Revised 2007 WHO classification of the central nervous system tumours, is categorized into pineocytoma (grade I), papillary tumour of the pineal region (grade II or III), pineal parenchymal tumour of intermediate differentiation (PPTID) (grade II or III) and pineoblastoma (grade IV). PPTID was first categorised by the WHO in 2000 as a pineal parenchymal tumour with an intermediate prognosis between pineocytoma and pineoblastoma. Pathologically classification of PPTIDs as grade II or III is based on mitosis, Ki-67 proliferation index and immuno-histochemistry.[3,4]

CASE REPORT

38-year-old gentle man presented with chief complaints of headache and vomiting since 1 day. MRI brain was advised. It showed a relatively well-defined lesion

measuring approximately 6×2×2 mm epicentered at posterior aspect of third ventricle. It appears hypointense on T1 (Figure 1), hyperintense on T2/ FLAIR and show heterogeneous post contrast enhancement. It shows few cystic areas within. It is extending and blocking aqueduct of sylvius leading to upstream dilatation of both lateral and third ventricles with periventricular ooze. Beta-hCG, alpha-feto protein and CEA levels were within normal limits.

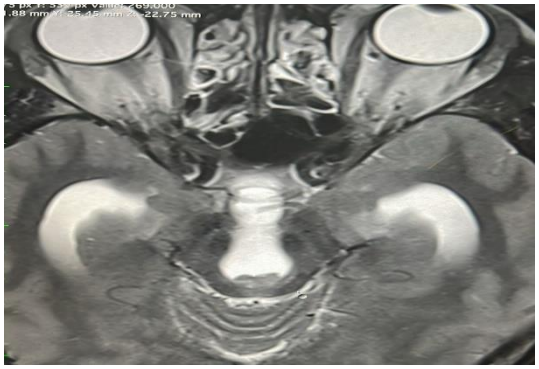

Figure 1: MRI of the brain demonstrates a well-defined lesion at posterior aspect of third ventricle which appears hypointense on T1, causing blockage of aqueduct of sylvius leading to upstream dilatation of both lateral and third ventricles.

Right sided medium pressure chhabra VP shunt followed by Neuro-navigation guided excision of space occupying lesion was performed.

Histopathological examination revealed highly cellular tumour comprising of diffuse sheets round to oval cells with small amount of cytoplasm, which was clear at places (Figure 2). Minimally pleomorphic nuclei were noted. Pleomorphic large ganglion cells or pineal rosettes were not seen. Areas of necrosis or endothelial proliferation was absent. The mitotic index was low-less than 6 mitosis per 10 higher power fields (Figure 3).

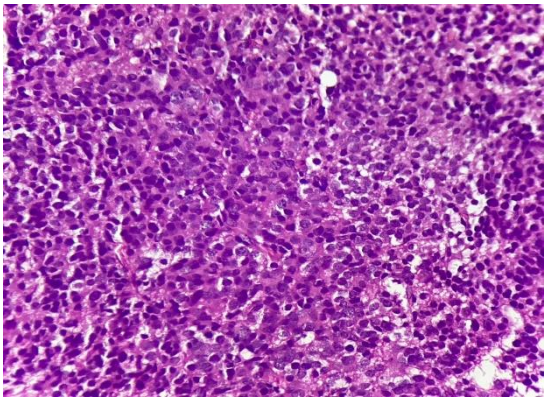

Figure 2: Haematoxylin and eosin stained slide demonstrate highly cellular tumour comprising of diffuse sheets of tumour cells with bland nuclear features (at 400X).

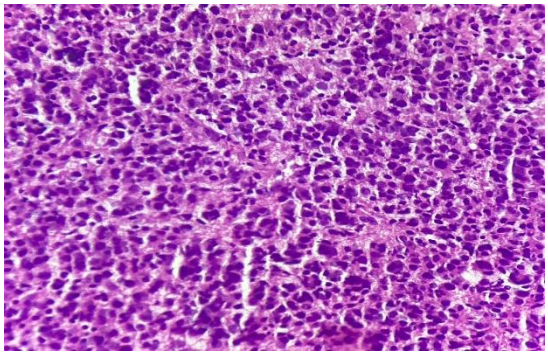

Figure 3: Haematoxylin and eosin stained slide demonstrate sheets of round to oval cells with minimally pleomorphic nuclei and small amount of eosinophilic to clear cytoplasm. Mitotic activity is low and pineocytomatous rosettes are absent (at 400X).

Immunohistochemical staining showed diffuse positivity for NSE, S-100, synptophysin (Figure 4) and focal for chromogranin while negativity for epithelial membrane, cytokeratin, glial fibrillary acidic protein, SALL 4 and placental like alkaline phosphatase. Grounded on these outcomes, a diagnosis of pineal parenchymal tumours of intermediate differentiation (PPTID) was set.

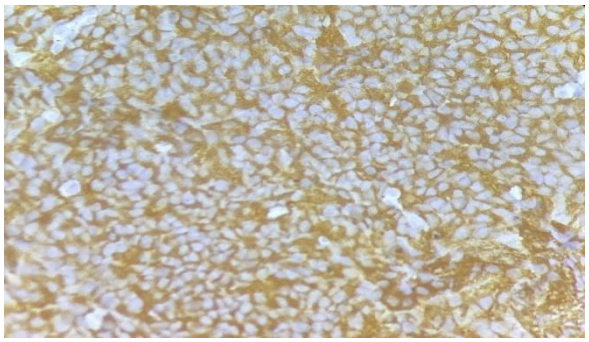

Figure 4: Immunohistochemistry shows diffuse cytoplasmic positivity of synaptphysin.

DISCUSSION

Tumours of pineal region are infrequent, accounting for over 1% of all intracranial tumours.[5] The mainstream of these tumours comprises of germ cell origin which embrace germinomas, embryonal cell tumours and choriocarcinomas.[6] Pineocytic (or their precursors) origin of pineal parenchymal tumour forms the second most common subgroup.

Previously, WHO has categorized pineal parenchymal tumours into only two subgroups: pineocytomas (WHO grade I) and pineoblastomas (WHO grade IV). However, in 2007 a new term called PPID was introduced which shows features intermediate between a pineocytoma and pineoblastoma, categorizing pineal parenchymal tumours into three histological grades.[4] PPID accounts for 45% of all pineal parenchymal tumours.

PPTID mostly presents with diplopia and headache as in our case.

Patients also presents with Parinaud's syndrome (vertical gaze disturbance due to compression of the tectal plate). Ataxia due to elevated intracranial pressure can be observed in large tumour causing hydrocephalus.[7] PPTID have a comprehensive age range between 4 to 75 years, with a mean age being 23 years. There is a trivial female preponderance (male: female=0.8:1).[8] Histopathologically, PPTID shows diffuse sheets of small uniform cells with marked cellularity, mild-to-moderate nuclear atypia and low-to-moderate mitotic activity. Pineocytomatous rosettes are usually lacking.[4]

The genetic alterations responsible for lashing PPTID is still not understood. Lee et al suggested that PPTID has persistent KBTBD4 small in-frame insertions, however it lacks DICER1 mutation or DROSHA homozygous deletion.[9] Due to the scarcity of reported cases, histopathological grading of PPTID is still contentious, even though most classify PPTID as WHO grade II or III.[3] Hasselblatt et al proposed a grading system based on mitosis, Ki-67 proliferation index and immunolabelling for neurofilament.[10] Pineal parenchymal tumours like pineocytomas and pineoblastomas, along with germ cell tumours and papillary tumours of the pineal region are differentials of PPTID.

Immunohistochemical staining, is valuable in differentiating these neoplasms as PPTID shows strong positivity for synaptophysin and neuron-specific enolase with variable positivity for neurofilament protein, chromogranin A, retinal S-antigen, S-100 protein and B-tubulinn.[3] As in our case shows diffuse positivity for synptophysin and focal for chromogranin while negativity for epithelial membrane, glial fibrillary acidic protein, SALL 4 and placental like alkaline phosphatase.

Due to rarity of these tumours, the optimal treatment protocols are yet to be established. With presenting symptom of hydrocephalus, the upmost precedence is decompression of the ventricular system. In most cases where lesion is locally limited, surgical resection is best opted. The decision of giving chemotherapy and radiation is grounded on the degree of resection and presence of spinal or cerebrospinal fluid metastases.[11] In contrast to this pineocytoma, are treated with surgical resection alone, whereas, pineoblastoma is given neoadjuvant chemotherapy and/or radiation, along with radical surgery.[12] The survival rate of PPTID varies according to grade of tumour. The overall survival of 5-year in low-grade cases is 74% whereas in high-grade it is only 39%.[13]

CONCLUSION

This is a rare case report of pineal parenchymal tumour in an adult male. PPTID are enormously erratic tumour, and restricted data are available concerning their pathologic features and biologic behaviours. Since the grading criteria of PPTID is not defined the optimal treatment protocols and prognosis of this entity is debateable. Inconsistencies of pathologic findings causes hindrance to make proper diagnosis and to decide an optimal treatment approach. WHO endorses on appropriate histopathological grading established on the proliferative index since immunoreactivity of neuronal markers did not link with the histologic grade and biologic behaviour of this tumour.

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