

Pleomorphic Variant of Pineocytoma – A Case Report –

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We herein present a rare case of pineocytoma in a 23-year-old female exhibiting distinct histomorphological features. The tumor contained highly pleomorphic, often multinucleated giant cells in the background of otherwise benign pineocytomatous architecture, which at first led to an erroneous diagnosis of a high grade malignancy. However, the worrisome histological findings turned out to be constituents of a distinct subtype of pineocytoma previously described as pleomorphic variant of pineocytoma. Although it is rare, pathologists should be aware of this entity since the tumor takes on a benign clinical course like any other classic pineocytomas.

Key Words : Pineal gland-Pineocytoma

The histomorphological spectrum of pineal parenchymal tumors (PPT) is diverse and not as clearly defined as other brain tumors due to their rarity. At one end of this spectrum lie pineocytomas, which are well-differentiated tumors that retain the morphological and immunohistochemical features of pineal parenchymal cells. A rare, but distinct subtype of pineocytoma has been previously described as pleomorphic variant of pineocytoma with gangliocytic differentiation containing giant cells with bizarre shaped nuclei,¹ which can easily be overdiagnosed as a high-grade malignancy. We recently encountered a similar case exhibiting histomorphological features characteristic of pleomorphic variant of pineocytomas, which to our knowledge is the first reported case in Korea.

CASE REPORT

A 23-year-old female presented at our hospital complaining of headache, nausea, and vomiting. She had given birth to a live baby four months ago and developed visual disturbance one month postpartum. She had no other significant medicosurgical illness.

Brain magnetic resonance imaging (MRI) study revealed a small round mass located in the third ventricle with homogeneous enhancement on T1-weighted image.

Intraoperative examination revealed a well-demarcated, yellowish white and soft tumor, which measured 1 × 1 cm. Total removal of the mass was performed. Histologically, the tumor displayed a vaguely lobulated appearance composed of cellular areas of isomorphic small cells with round to oval nuclei (Fig. 1A), and less cellular foci with a fibrillary background and patchy areas consisting of microcysts and vascular stroma. Scattered in these less cellular foci were highly pleomorphic and often multinucleated cells with bizarre hyperchromatic nuclei (Fig. 1B). Occasional pineocytomatous rosettes, although rather ill-defined, were recognized with fine fibrillary centers. There was no discernible mitosis or necrosis. Immunohistochemistry disclosed diffuse and strong positivity for synaptophysin, neuron-specific enolase and neurofilament protein in the cytoplasm and cytoplasmic processes, suggesting neuronal differentiation of the tumor (Fig. 1C, D). The tumor showed weak immunoreactivity for glial fibrillary acidic protein and immunohistochemistry for placenta-like alkaline phosphatase and human chorionic gonadotropin were negative.

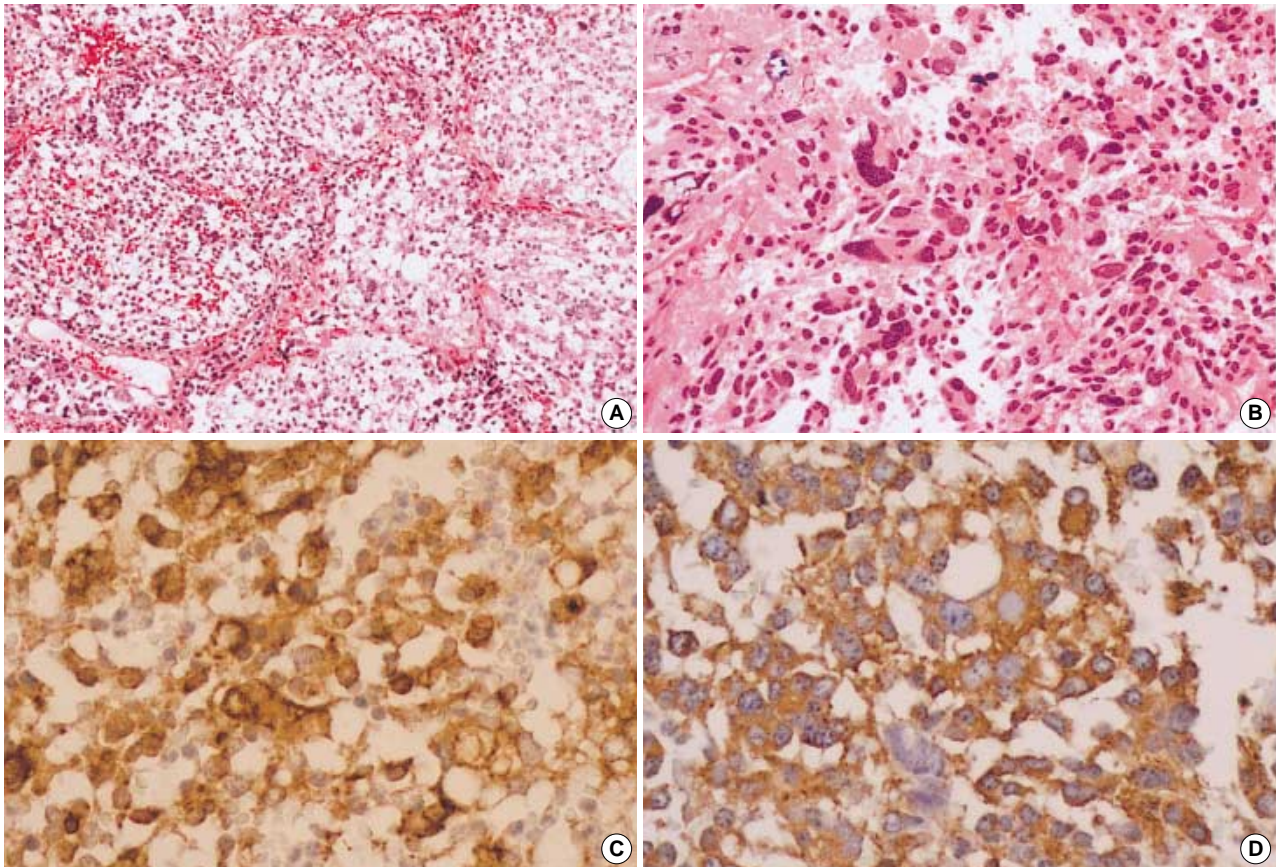


Fig. 1. (A) The tumor is vaguely lobulated and composed of cellular areas of isomorphic small cells with round to oval nuclei with a fibrillary background and vascular stroma, and (B) areas showing large, highly pleomorphic giant cells. Immunohistochemistry reveals diffuse positivity for neurofilament protein (C) and synaptophysin (D) in the tumor cells.

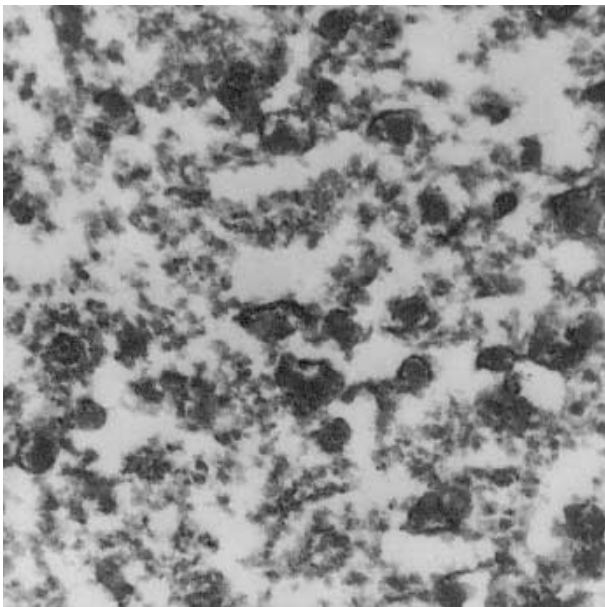


Fig. 2. Electron microscopy demonstrates numerous membrane-bound electron-dense granules in the cytoplasm ($\times 55,000$).

Ultrastructurally, membrane-bound, electron-dense granules and numerous neurosecretory granules were present in the cytoplasm and cellular processes (Fig. 2). The patient is currently well without evidence of recurrence during the past 12 months of follow-up.

DISCUSSION

PPT display a broad histomorphological spectrum comprising immature pineoblastomas, mature pineocytomas and mixed pineocytoma/pineoblastomas. They account for less than 0.1% of all brain tumors and due to their rarity, the histological features of these tumors have been reported only in limited numbers.² Nevertheless, accurate histological diagnosis is critical for proper management.

The scattered bizarre-shaped and often multinucleated giant cells can be alarming, and may mislead the pathologist into rendering a faulty diagnosis of a high-grade malignancy, as in our

case. However, these features have been previously observed in PPT and do not necessarily indicate malignancy.³ In fact, in a collective review of PPT by Jouvét et al., five cases of pleomorphic variants of pineocytoma were included and none were found to demonstrate recurrence or any other signs of malignant behavior.¹ According to Herrick and Rubinstein, pineal parenchymal cells bear the multipotential to differentiate into diverse morphological patterns during tumorigenesis.⁴ The diffuse immunoreactivity to neuronal markers of the tumor cells and the ultrastructural findings of numerous neurosecretory granules in our case also confirm the previous findings that a neuronal immunophenotype is common in pineocytoma.⁵

The differential diagnoses may include gangliocytomas, gangliogliomas, and atypical pleomorphic astrocytomas of the pineal gland. However, the isomorphic, rosette-bearing areas characteristic of pineocytomas are not seen in these tumors,^{6,7} and pleomorphic astrocytomas demonstrate exclusively astrocytic differentiation.⁸ Central neurocytoma may also be considered in the diagnosis as they frequently demonstrate rosettes, however, they are localized in the supratentorial ventricles and do not show the cellular pleomorphism seen in this case.

We believe our case is consistent with pleomorphic variant of pineocytoma, both histomorphologically and immunohistochemically. Pathologists should be aware of this variant of pineocytoma in order not to overdiagnose it as a pineoblastoma or some other malignancy especially in stereotaxic biopsies, as an overdiagnosis may call for unnecessary aggressive therapeutic management. Despite the worrisome presence of highly pleomorphic, multi-

nucleated giant cells with bizarre nuclei, the biological behavior of pleomorphic variant of pineocytoma is essentially benign without cerebrospinal seeding or distant metastasis.¹

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