Article published online: 2022-12-27

Mast Cell-Rich Pleomorphic Pineocytoma: A Rare Entity

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Indian J Neurosurg 2023;12:90–92.

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Pineal region tumors constitute a rare group of neoplasms. Primary neuronal tumor that intrinsically arise from the pineal gland are referred to as pineal parenchymal tumors (PPTs). Their neuropathological spectrum is divided into well-differentiated "pineocytoma" (World Health Organization [WHO] grade I), pineal parenchymal tumors of intermediate differentiation and papillary tumor of pineal region (WHO grades II and III), and poorly differentiated "pinealoblastoma" (WHO grade IV).[1] Pleomorphic cytological variant of pineocytoma has been rarely reported in the medical literature mainly in form of case reports. A 45-year-old man with a recent history of right-sided stroke presented to the neurosurgery department with complaint of headache for last 1 month. Multislice computed tomography (CT) imaging revealed an ill-defined hyperdense space-occupying lesion with internal calcification. The tumor was located in the region of pineal gland with extension into the adjacent lateral ventricle and resultant mild obstructive hydrocephalus. MRI scan revealed a hyperdense nodular lesion measuring 24×18 mm with scant peritumoral edema (►**Fig. 1A** and **1B**). Following placement of a ventriculo peritoneal shunt, stereotactic biopsy was performed and submitted to the pathology department. Histopathological examination revealed a pineal parenchymal neoplasm with varied morphology. In some areas, the tumor cells were small and uniform resembling pinealocytes, and arranged in sheets and at places forming pineocytomatous rosettes with abundant delicate tumor cell processes. (►**Fig. 2A**) Their nuclei were round to oval with inconspicuous nucleoli and finely dispersed chromatin. However, in other areas, the tumor was less cellular with the presence of several multinucleated giant cells and bizarre nuclei exhibiting marked nuclear pleomorphism with hyperchromasia (►**Fig. 2B**). Interstitial stroma demonstrated presence of hyaline vessels and reticulin fibrils. Mitotic figures were lacking (< 1 mitosis/10 High power field (HPF)) in the tumor. No ganglionic differentiation was observed in the present case. Immunohistochemical examination revealed neuronal tumor immunophenotype with diffuse positivity for non-specific enolase, synaptophysin, and focal positivity for S-100. The tumor cells were negative for glial fibrillary acidic protein (GFAP), placental alkaline phosphatase (PLAP), Human Chorionic Gonadotropin (HCG), and pan–cytokeratin. K_i-67 proliferation index was less than 1%. The expression of GFAP was observed only in the resident astrocytes . A striking feature observed in the present case was the presence of numerous mast cells that were concentrically arranged around larger blood vessels. Toluidine blue stain and CD117 highlighted the presence of mast cells (►**Fig. 3A–3C**).

Pleomorphic pineocytoma is a cytological variant that has been rarely reported in the medical literature.[2] The ominous nuclear appearance of tumor cells could be misleading and pose a diagnostic difficulty. A large spectrum of neoplasm

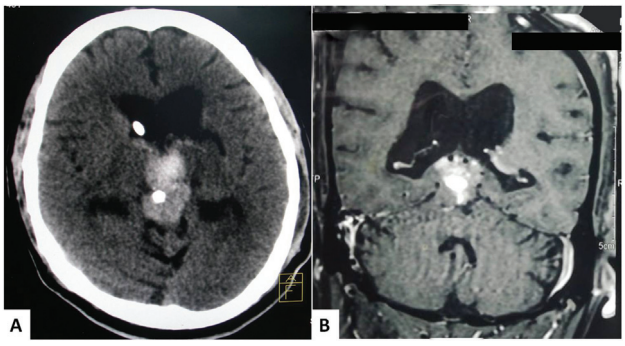

Fig. 1 (**A**) CT image showing a hyperdense space-occupying lesion in the pineal region with internal calcification. (**B**) MRI image revealing contrast-enhancing nodular mass with scant peritumoral edema.

article published online
May 25, 2021

DOI https://doi.org/10.1055/s-0042-1756489.
ISSN 2277-954X.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

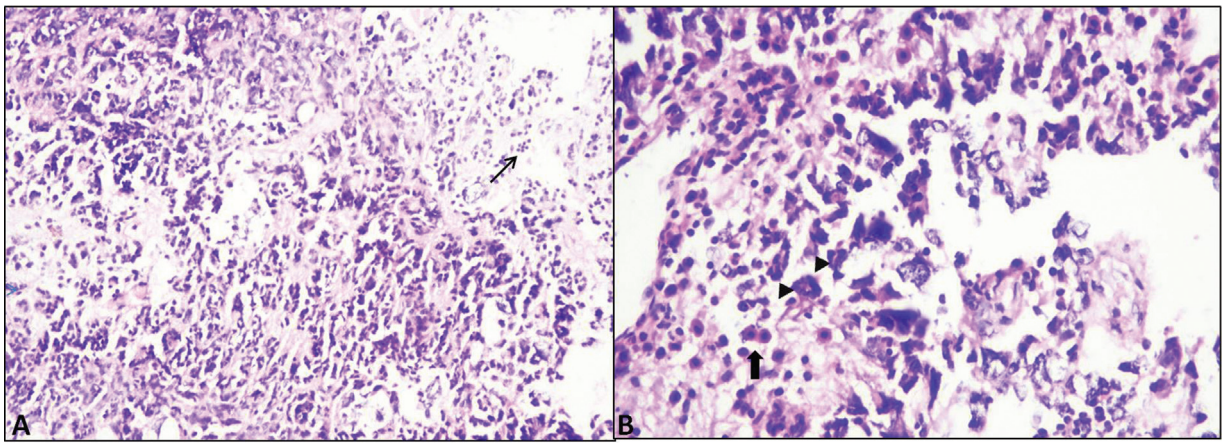

Fig. 2 (**A**) Hematoxylin and eosin-stained section showing varied morphology along with the presence of scattered pinealocytes (marked with a thin arrow; 10X magnification). (**B**) Higher power view demonstrating the presence of pineocytomatous rosettes (marked with an arrow head) interspersed with bizarre pleomorphic cells and numerous mast cells (marked with a thick arrow).

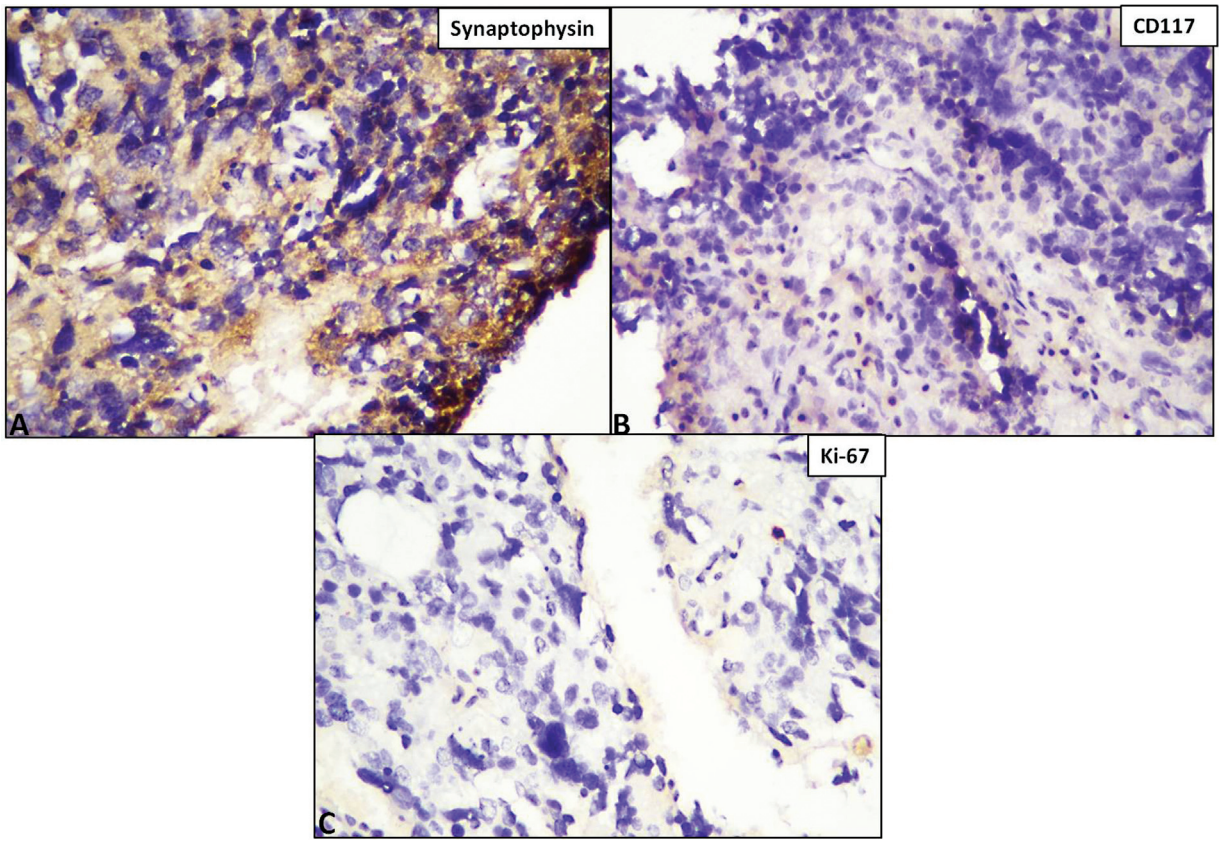

Fig. 3 Immunohistochemical stain. (**A**) Synaptophysin–tumor cells displaying strong cytoplasmic positivity (**B**): CD-117–Numerous mast cells exhibiting faint positivity are seen interspersed in the tumor. (**C**) K_i-67 labeling shows < 1% proliferation index in tumor cells.

can arise in the vicinity of pineal gland including the tumor, metastasizing to the pineal region. Differential diagnoses that were considered in the present case were atypical Astrocytoma of the pineal gland (lack of fibrillary background, tumor cells were negative for GFAP and S 100); anaplastic pleomorphic xanthoastrocytoma (lack of xanthomatous change, tumor cells were negative for GFAP, S100 and CD34), and pineoblastoma (lack of Homer–Wright rosettes, mitoses, and necrosis).[3] Pineocytomas are usually small tumors (< 3 cm) that remain localized to the pineal region, wherein they cause compression of adjacent structures. Protrusion of pineocytomas can occur in posterior one-third of the ventricle as was observed in our present case.[4] Prominence of mast cells around blood vessels was an unusual feature noted in the tumor. Various central nervous system tumors including glioblastoma multiforme, hemangioblastoma, and meningioma have demonstrated the

presence of increased mast cells.[4] To conclude, pleomorphic pineocytoma is a rare histologic entity. Bizzare appearance of tumor cells may be misleading and K_i-67 proliferative index may provide an essential clue for the diagnosis.

Conflict of Interest
None declared.

References

1 Hasselblatt M, Huang A, Jones DTW, Orr BA, Snuderl M, Vasiljevic A. Pineal Tumors. In: Cree IA, Lokuhetty D, Peferoen LAN, White VA, eds. World Health Organization Classification of Tumors of Central Nervous System, 5th ed. Lyon: International agency for research on cancer (IARC), 2021:242–255
2 Kuchelmeister K, von Borcke IM, Klein H, Bergmann M, Gullotta F. Pleomorphic pineocytoma with extensive neuronal differentiation: report of two cases. Acta Neuropathol 1994;88 (05):448–453
3 Nitta J, Tada T, Kyoshima K, et al. Atypical pleomorphic astrocytoma in the pineal gland: case report. Neurosurgery 2001;49(06):1458–1460, discussion 1460–1461
4 Polyzoidis S, Koletsa T, Panagiotidou S, Ashkan K, Theoharides TC. Mast cells in meningiomas and brain inflammation. J Neuroinflammation 2015;12:170

