

## **Pineocytoma - Pleomorphic Grade I pineal region tumor**

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**Abstract:** 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 which can easily be overdiagnosed as a high-grade malignancy. We describe a case report of a 58 years patient who had pineal parenchymal tumor with cellular features of a pineocytoma along with pleomorphic cells.

**Keywords:** Pleomorphic, Pineocytoma, Pineal parenchymal tumors of intermediate differentiation

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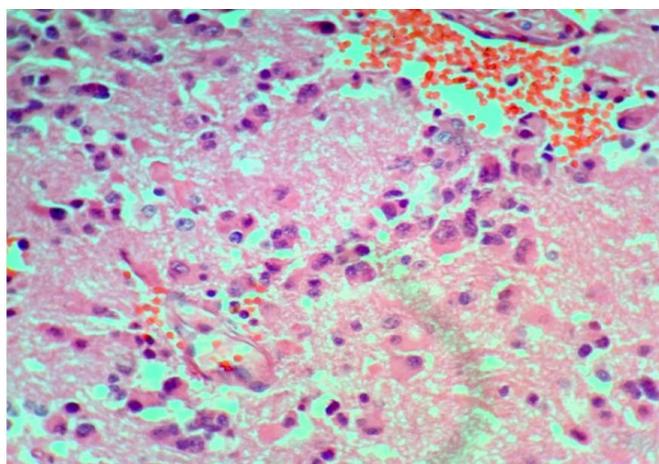
### **INTRODUCTION**

Neoplasms of the pineal region are a rare group of tumors accounting for less than 1% of all intracranial tumors, and represent a very clinically and pathologically heterogeneous group of tumors including pineal parenchymal tumors, germ cell tumors, astrocytomas, ependymomas, and papillary pineal tumors [1-3]. Many investigators have therefore stressed the importance of a tissue-based diagnosis for patient management. Tumors thought to arise from the parenchymal cells of the pineal gland, also referred to as pineal parenchymal tumors, consist of approximately one-third of all tumors of the pineal region [1, 4]. The latest World Health Organization (WHO) classification scheme, released in 2007, categorizes pineal parenchymal tumors into 3 subtypes with up to 4 different grade categories: (1) WHO Grade I

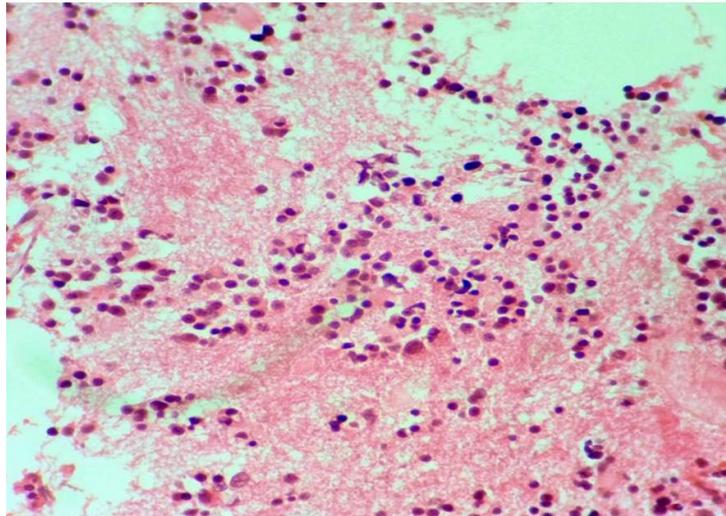
pineocytomas (PC), (2) WHO Grade II or III pineal parenchymal tumors of intermediate differentiation (PPTID), and (3) WHO Grade IV pineoblastomas (PB) [5].

### **CASE REPORT**

A 58 years old male patient presented with cognitive problems like loss of memory and control of bowel and bladder function since 4 to 5 months. MRI showed a 1.5x1 cm mass in the pineal region causing hydrocephalus. Craniotomy was done and tissue was sent for histopathological examination. The Microscopic analysis of tumor showed multiple rosettes formed by cells with small round nuclei. The other population of cells showed enlargement of nuclei with some showing multinucleation and pleomorphism and confirmed as Pineocytoma.



**Fig-1: Section studied show tumour cells arranged in sheets, some having perinuclear halo with mild pleomorphism. [H&E, x40]**



**Fig-2:** Section studied show tumour cells arranged in micro rosettes [H&E, x10]

## DISCUSSION

Pineocytomas are the lowest grade (WHO Grade I) tumors with the most favorable prognosis [6, 7]. The 5-year survival has been reported to range from 64% to 91% [1, 8-10], although the latter figure is probably more accurate when strictly defined using the most current criteria. These tumors are found most commonly in the adult population, and clinically appear to progress slowly, although symptomatic recurrences have commonly been reported even after aggressive resection [11-13]. Pineocytomas grossly are well circumscribed and cause symptoms by local growth with local compressive mass effect.

The histologic features of pineocytomas include their cellular resemblance to mature pineocytes, and they are primarily composed of well-differentiated cells. However, in contrast to the normal pineal gland's lobular architecture created by gliovascular septae, pineocytomas are arranged in sheets of round tumor cells with variable oligodendroglioma-like clear haloes. Portions of the tumor are occasionally found to have focal ganglionic and/or astrocytic differentiation, and at times cells displaying features of ganglion cells and astrocytes can be found within the same tumor. Early studies have suggested a more benign clinical course for pineocytomas with neuronal or neuronal and astrocytic differentiation, with a greater tendency to remain localized as compared with pineocytomas with astrocytic differentiation.

More recent studies, however, have failed to establish this correlation. Pineocytomatous rosettes, also referred to as pineocytic rosettes, are frequently observed, and are believed to be a distinct feature of pineocytomas. These rosettes consist of tumor cells surrounding pink neuropil (collections of neuronal processes) and are similar in appearance to Homer-Wright rosettes, but are formed by mature rather than primitive cells and tend to be somewhat larger and more

irregular. Despite the occasional resemblance to oligodendrogliomas, neuronal differentiation is evident in the form of diffuse synaptophysin immunoreactivity and often strong positivity for neurofilament protein, the latter occasionally highlighting bulbous axonal swellings typical of pineal differentiation. The level of proliferative activity is relatively low in pineocytomas, with mitoses being rare and MIB-1 (Ki-67) [14, 15] labeling indices averaging around 1.6%. In one study, the MIB-1 labeling index was significantly different for each of the 3 pineal parenchymal tumors, and the investigators suggest its use as an additional measure to differentiate between the 3 subtypes.

To conclude this case illustrates that pleomorphic pineocytomas underline the broad histomorphological spectrum of pineal parenchymal tumors and should be considered in differential diagnosis

## REFERENCES

1. Fauchon F, Jouvét A, Paquis P, Saint-Pierre G, Mottolese C, Hassel MB, Chauveinc L, Sichez JP, Philippon J, Schlienger M, Bouffet E. Parenchymal pineal tumors: a clinicopathological study of 76 cases. *International Journal of Radiation Oncology\* Biology\* Physics*. 2000 Mar 1;46(4):959-68.
2. Prahlow JA, Challa VR. Neoplasms of the pineal region. *Southern medical journal*. 1996 Nov;89(11):1081-7.
3. Schulte FJ, Herrmann HD, Müller D, Franke H, Saeger W, Spaar FW, Bartels S. Pineal region tumours of childhood. *European journal of pediatrics*. 1987 May 1;146(3):233-45.
4. Linggood RM, Chapman PH. Pineal tumors. *Journal of neuro-oncology*. 1992 Jan 1;12(1):85-91.
5. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, editors. WHO classification of tumours of the central nervous system. World Health

- Organization classification of tumours. World Health Organization; 2007.
6. Jouvet A, Saint-Pierre G, Fauchon F, Privat K, Bouffet E, Ruchoux M, Chauveinc L, Fèvre-Montange M. Pineal parenchymal tumors: a correlation of histological features with prognosis in 66 cases. *Brain Pathology*. 2000 Jan 1;10(1):49-60.
  7. Deshmukh VR, Smith KA, Rekate HL, Coons S, Spetzler RF. Diagnosis and management of pineocytomas. *Neurosurgery*. 2004 Aug 1;55(2):349-57.
  8. Dahiya S, Perry A. Pineal tumors. *Advances in anatomic pathology*. 2010 Nov 1;17(6):419-27.
  9. Schild SE, Scheithauer BW, Schomberg PJ, Hook CC, Kelly PJ, Frick L, Robinow JS, Buskirk SJ. Pineal parenchymal tumors: clinical, pathologic, and therapeutic aspects. *Cancer*. 1993 Aug 1;72(3):870-80.
  10. Clark AJ, Sughrue ME, Ivan ME, Aranda D, Rutkowski MJ, Kane AJ, Chang S, Parsa AT. Factors influencing overall survival rates for patients with pineocytoma. *Journal of neuro-oncology*. 2010 Nov 1;100(2):255-60.
  11. Borit A, Blackwood W, Mair WG. The separation of pineocytoma from pineoblastoma. *Cancer*. 1980 Mar 15;45(6):1408-18.
  12. Disclafani A, Hudgins RJ, Edwards MS, Wara W, Wilson CB, Levin VA. Pineocytomas. *Cancer*. 1989 Jan 15;63(2):302-4.
  13. Tracy PT, Hanigan WC, Kalyan-Raman UP. Radiological and pathological findings in three cases of childhood pineocytomas. *Child's Nervous System*. 1986 Dec 1;2(6):297-300.
  14. Arivazhagan A, Anandh B, Santosh V, Chandramouli BA. Pineal parenchymal tumors--utility of immunohistochemical markers in prognostication. *Clinical neuropathology*. 2008;27(5):325-33.
  15. Rickert CH, Simon R, Bergmann M, Dockhorn-Dworniczak B, Paulus W. Comparative genomic hybridization in pineal parenchymal tumors. *Genes, Chromosomes and Cancer*. 2001 Jan 1;30(1):99-104.