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Hrushikesh Kharosekar,  
Laxmikant Bhople,  
Reshma Pujara,  
Smita Ranveer



# Supratentorial PNET in a geriatric patient. A rare differential diagnosis leading to diagnostic dilemma

Hrushikesh Kharosekar<sup>1</sup>, Laxmikant Bhople<sup>2</sup>, Reshma Pujara<sup>3</sup>, Smita Ranveer<sup>4</sup>

<sup>1</sup> Associate Professor, Dept of Neurosurgery, Sir J. J. Group of Hospitals and Grant Medical College, Mumbai, INDIA

<sup>2</sup> Assistant Professor, Dept of Neurosurgery, Sir J. J. Group of Hospitals and Grant Medical College, Mumbai, INDIA

<sup>3</sup> Consultant Anaesthesiologist, Oscar Hospital, Thane, INDIA

<sup>4</sup> Consultant Pathologist, Radiance Pathology Laboratory, Thane, INDIA

## ABSTRACT

Supratentorial PNETs are most commonly seen in children and rarely seen in adults. PNET show a proliferation of undifferentiated or poorly differentiated neuroepithelial cells and, thus, are histologically similar to medulloblastomas. They account for approximately 2.5% of brain tumours in children and only 0.4% in adults. Prognosis is poor in the pediatric age group while it shows favourable prognosis in adults. In literature, less than 100 cases of adult PNET have been reported till date with mean age of 35years. PNET in the geriatric age group is rarely been reported.

## INTRODUCTION

The term of primitive neuroectodermal tumor (PNET) was coined by Hart and Earle in 1973 for small round cell tumors with high malignant potential and affecting both the central and peripheral nervous systems. PNETs show a proliferation of undifferentiated or poorly differentiated neuroepithelial cells and, thus, are histologically similar to medulloblastomas. World Health Organization (WHO) described supratentorial PNETs as cerebral or suprasellar embryonal grade IV tumor with capacity to display differentiation along neuronal, astrocytic, ependymal, muscular or melanocytic lines. These tumors are most commonly seen in children and rarely seen in adults. They account for approximately 2.5% of brain tumors in children and only 0.4% in adults. Prognosis is poor in pediatric age group while in shows favorable prognosis in adults. <sup>(1)</sup> In literature less than 100 cases of adult PNET have been reported till date with mean age of 35years. PNET in geriatric age group is rarely been reported. <sup>(2)</sup>

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**Keywords**  
PNET,  
geriatric,  
diagnostic

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Corresponding author:  
Hrushikesh U. Kharosekar

Dept of Neurosurgery, Sir J. J. Group  
of Hospitals and Grant Medical  
College, Mumbai, India

hkharsekar@gmail.com

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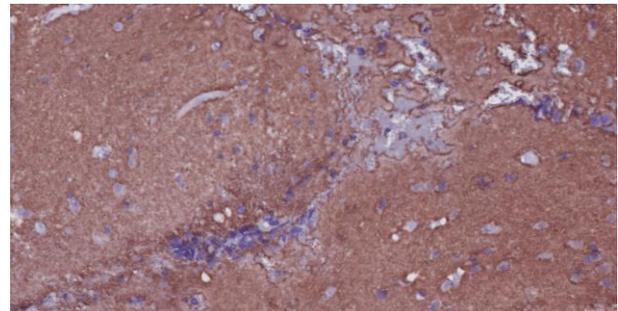
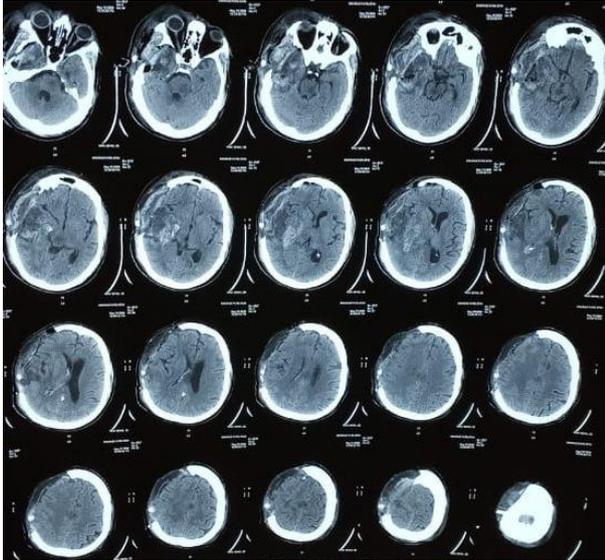
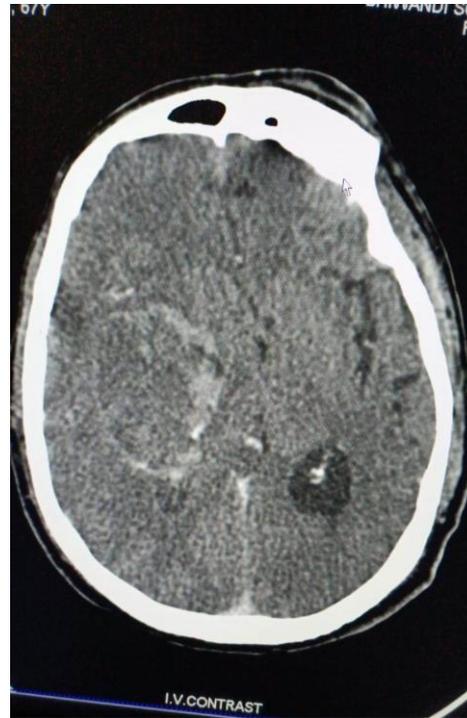


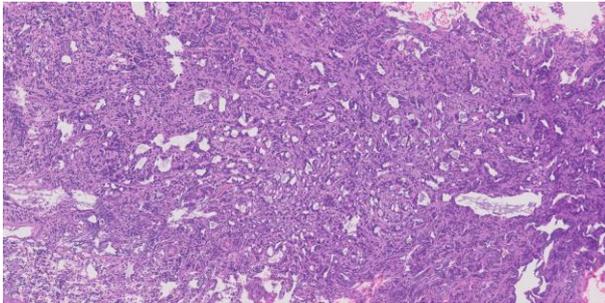
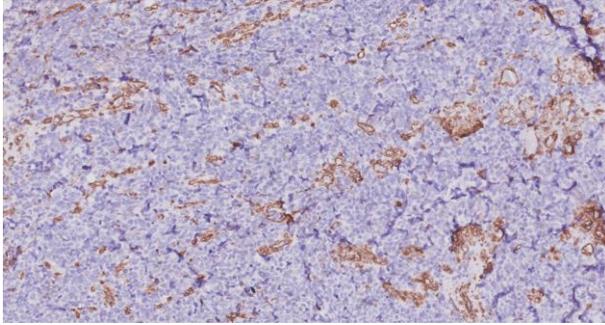
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**CASE**

We had a 67-year-old male patient presented to us with sudden onset severe headache and vomiting for 1 day, patient was drowsy on examination. His CT scan of brain with contrast was suggestive of a large, 5\*4\*3 cm, well circumscribed lesion in right temporal lobe with perilesional edema. Lesion was minimally enhancing on contrast with isodense centre. As patient was drowsy, he underwent emergency craniotomy with near total excision of lesion. Small part of lesion which was invading the basal ganglia was left behind as seen on post-operative CT scan. Post operatively patient improved in consciousness and was discharged on 7th post-operative day without any deficits.

Histopathology was suggestive of highly cellular lesion with nests, cords and sheets of tumor cells with high N/C ratio. Hyperchromatic nuclei, inconspicuous nucleoli and brisk mitoses was seen. On IHC, tumor cells were positive for CD99, CD 56 and vimentin, GFAP negative, Ki-67 ratio was high (60-70%), suggestive of PNET (Intracranial central). Patient was advised post-operative radiotherapy and chemotherapy.





## DISCUSSION

Primitive neuroectodermal tumors (PNETs) include a group of tumors, thought to originate from undifferentiated neuroepithelial cells, that are commonly seen in pediatric patients and rare in adults. Hart and Earle described the term of primitive neuroectodermal tumor – PNET – in 1973 originally for cerebral high-grade undifferentiated neuroepithelial tumor of childhood, rarely demonstrating focal differentiation along glial and neuronal lines. This term was soon generalised for undifferentiated embryonal tumors of all CNS sites and all ages by Rorke in 1983. PNETs may occur in almost any location within or outside the central nervous system. PNETs seen outside the CNS are termed as peripheral PNETs (pPNET). CNS PNET and pPNET are two different entities with different immunohistochemical profiles and genetics. Clinically both are aggressive tumors, but show different local manifestation and metastatic spread. CNS PNETs can be further divided into 2 types: infratentorial tumors (medulloblastoma) and supratentorial tumors (sPNETs). All these tumor types are rare in adults.<sup>(1,2)</sup>

Intracranial PNETs are uniformly distributed in the frontal lobe, temporal lobe, and parietal lobe and they are more than 6cm in size. Intracranial PNETs

are further divided into central and peripheral according to their location with peripheral variety carrying a better prognosis. Accordingly, it is essential to differentiate between the two types. Chromosomal translocation of chromosome (11;22) is unique to central and not seen in peripheral PNETs. Immuno-histochemical assay for CD99 and fluorescence in situ hybridization (FISH) assay for the (11;22) translocation are specific for central PNET.<sup>(2,3)</sup> The exact protocol for treatment of PNET is not clearly defined yet. So as per patient's clinical status, complete tumor excision, chemotherapy, and radiotherapy are performed as standard procedures like any other intracranial tumors. Prognosis of patients with PNET differs according to age. Pediatric age group carries poor prognosis with older age appears to be have favorable prognosis. Unfortunately, the 5-year survival in case of CNS PNET remains to be less than 50% in all age groups.<sup>(1,2)</sup>

## CONCLUSION

The diagnostics and treatment protocol of primitive neuroectodermal tumors does not differ from other types of the central nervous system tumors. Complete tumor excision, chemotherapy, and radiotherapy are performed as a treatment standard with better survival prognosis in old age. So even though very rare in older age group PNET should be kept as one of differential diagnosis as it carries better prognosis in this age group. A CD99 and FISH assay for the (11;22) translocation (unique to central and not peripheral PNETs) should be conducted in order to distinguish between intracranial peripheral and intracranial central PNETs, as peripheral PNET carries better prognosis.

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