A Rare Case of Peripheral PNET with A Rare Presentation

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ABSTRACT

A 28 year male without any comorbidity having fever since 2 weeks and complex partial seizures since day 1 was presented with altered sensorium with left sided hemiparesis in emergency room and evaluated with MRI Brain which s/o Right fronto-temporal bleed with mass effect. Emergency Right sided Decompression Craniotomy done, intraoperative temporal lobe having mass lesion with clot around it; total excision of mass with clot evacuated. On histopathology and IHC study show its rare case of Peripheral PNET lesion. Postop period uneventful.

Key Words: Hemiparesis, P-PNET, Craniotomy, Round Cells, Synaptophysin.

Introduction

History: A 28 year old male presented with Fever since 2 weeks on-off episodes, seizures involving right side of body since 2 weeks, on medications, altered sensorium since 1 day. Not significant family or past history.

Examination: Drowsy, arousable, febrile 100.4 F. GCS E2 V2 M5, Pupils B/L ERTL, Left side Hemiparesis, vitals stable.

Investigations: All routine blood investigations, chestxray were apparently Normal.

CT scanbrain & MRI BRAIN have been done s/o Right temporal SOL with haemorrhages inside with mass effect? Glioblastoma Multiforme? Cavernoma with bleed? AVM.

Fig.1: T1 image
Fig.2: T2 image
Management
After evaluation, patient under went right Fronto Temporo Parietal Decompressive Craniotomy and evacuation of hematoma with lesion. Procedure and post-operative course was uneventful. Patient recovered well after surgery and gain full consciousness with left hemiparesis which improved with time.

Histopathology
H & E Stain: Small round blue cells neoplasm with primitive appearing and some neuronal differentiated cells with mitotic figures.
Positivity with Synaptophysin: A marker of neuronal cells
GFAP: Negative for GFAP-glial origin
EMA: Negative for EMA -ependymal cell origin
Ki-67 index: (a cellular marker for proliferation) 30-40%
Strong Membranous Positivity with CD99
Friend leukemia integration 1 transcription gene (FLI 1): Expression of this gene is highly specific for P-PNET

Finally IHC & Histology features are diagnostic of Peripheral PNET.

Cause: Loss of the short arm of chromosome 17 (17p13.3) is the most frequent abnormality.

Conclusions
P-PNET is very rare presentation in our clinical practice & prognosis will be decided on chromosomal study & histology study.

References