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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.

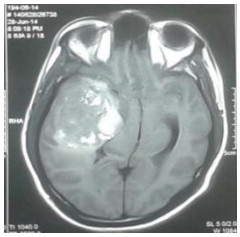

Fig.1:T1 image

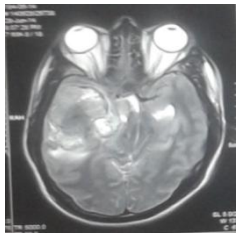

Fig.2: T2 image

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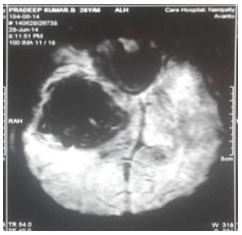


Fig.3: GRE image

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-glialorigin

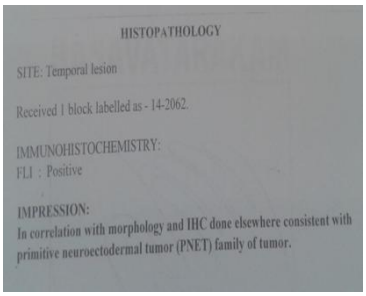
EMA: Negative for EMA -ependymal cellorigin

Ki-67index: (a cellular marker for proliferation) 30-40%

Strong Membranous Positivity with CD 99

Friend leukemia integration 1transcription gene (FLI 1): Expression of this gene is highly specific for P-PNET

Finally IHC & Histology features are diagnostic of Peripheral PNET.


HISTOPATHOLOGY

SITE: Temporal lesion

Received 1 block labelled as - 14-2062.

IMMUNOHISTOCHEMISTRY:

FLI : Positive

IMPRESSION:

In correlation with morphology and IHC done elsewhere consistent with primitive neuroectodermal tumor (PNET) family of tumor.

Fig.4: IHC Study

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.

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