Extraosseous Primary Intracranial Ewing Sarcoma Peripheral PNET- (EWS-pPNET): Series of Seven Cases and Review of Literature

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Introduction

The Ewing sarcoma peripheral PNET (ES-pPNET) is very rare small round cell tumour that involves the CNS as either a primary dural neoplasm or by direct extension from contiguous bone or soft tissue.

Methods

Biopsy proven cases of intracranial ES/pPNET with orbital involvement operated during Jan 2010-Jan 2014 were retrospectively included and their clinical data, operative and histological findings were reviewed from institutional oncology register.

Results

seven patients (4 males; 3 female) were studied with mean age at presentation of 13 years. Six patients had orbital involvement in one or other form. Surgical excision was gross total in five, near total in one, and subtotal in one patient. All patients received adjuvant therapy, only chemotherapy in 2, only Radiotherapy in four, both in one. MRI characteristics were studied in six patients. Four patients died with average survival of 33.2 months and three patients are having Progression free survival of average 23.3 months

Conclusions

The EWS-pPNET is very rare tumour and very poorly described in literature. These tumours are showing special predilection for the fronto-temporal dura and erode through the flat bone of cranium like orbital roof and lateral wall of the orbit. These tumours are aggressive, multi compartmental, vascular and very rapidly growing, so missing or overlooking the primary symptoms of dural stretching/bony involvement leads to delay in management and poor outcome.

Learning Objectives

To study this relatively rare entity and its management nuances

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