Title : Atypical teratoid rhabdoid tumor of the central nervous system: Case series from a regional Tertiary Care Cancer Centre in South India

Author Name : L Appaji

CO Author Name : Aruna kumari B.S

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Asbstract :

Background: Atypical teratoid rhabdoid tumors (AT/RT) constitute a rare group of pediatric brain tumors.
Materials and Methods: This was a retrospective study. Demographic and clinical data were obtained from the clinical case files. Archived slides and tissue blocks were retrieved. All cases had hematoxylin and eosin stained sections. IHC was available in all the cases.
Results: There were eight cases with the mean age of presentation being 4 years (range: 4 months to 15 years) and with slight male predominance (male:female = 1.66:1). Most of the presenting complaints were due to raised intra-cranial tension. The median duration of symptoms was 0.75 months. About 62.5% of the tumors were infratentorial in location. The tumors were heterogeneous showing variable expression of cytokeratin, epithelial membrane antigen, glial fibrillary acid protein, and synaptophysin. Loss of integrase interactor-1 expression was demonstrated in seven cases in which it was done. Multimodal treatment comprising surgical resection, radiotherapy and chemotherapy was tailored based on location of tumor, resectability and patient's age. The median overall survival was 2.5 months (range: 1.5–30 months).
Conclusion: Awareness of this tumor is important as it portends a poor outcome in most patients, in spite of multi-modal treatment. Several new molecules which aim to prolong survival and improve quality of life are being developed to combat this enigmatic tumor.