Meenakshi Rattan, Madridge J Intern Emerg Med. 2019
http://dx.doi.org/10.18689/2638-1621.a2.010

AT/RT: (Atypical Teratoid Rhabdoid Tumor): Palliative Surgical Approach

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It is a rare malignant embryonal CNS tumor typically seen in young children with a peak incidence during the first 2 years of life. AT/RT represents 2–3% of all paediatric brain tumors and 10–15% of brain tumors in children less than 3 years old. It was first included in the World Health Organization classification of tumors of the CNS in 2000, although it had been recognized during the early 1980s as a rhabdoid tumor of the CNS with an unfavourable prognosis. In CNS tumors, the clinical presentation is related more to the tumor location at the time of diagnosis than to the histology. Clinical manifestations are usually non-specific and in children younger than 3 years clinical manifestations are less specific, including lethargy, vomiting or failure to thrive. Head tilt, cranial nerve palsies, headache and focal neurological deficit are more prevalent in older children. As with other malignant paediatric brain tumours, gross total resection seemed to be associated with better outcome. However, death often occurs within a year of diagnosis despite multimodality therapy. Multi-agent systemic agents, including anthracyclines, intrathecal chemotherapy and radiotherapy, demonstrated positive impact on overall survival. The overall survival ranges from 0 to 45% and salvage is extremely rare after progression or recurrence.

Biography:

Dr. Meena Rattan is general Paediatrician from Sydney working as consultant at Wyong hospital in Central Coast NSW Australia. She has special interest in paediatric endocrinology, child psychiatry, neurodevelopmental and behavioural Paediatrics. She is trainer for Rhesus 4 kids and also for neonatal resuscitation.